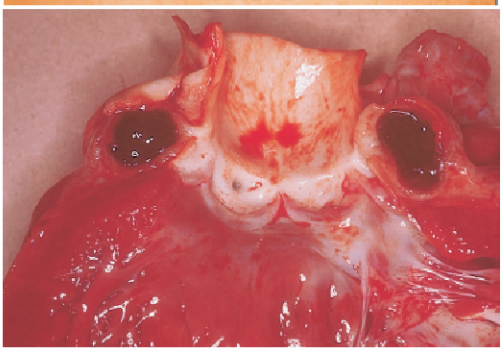
3rd Annual Conference of

Indian Society of Kawasaki Disease



Executive Board:

President: Dr. Saji Philip

President Elect: Dr. Surjit Singh

Secretary: Dr. Priyankar Pal

Venue: Zoom platform
Dates: 31.10.2020 and 01.11.2020
Time: 16:00 pm (GMT+5.5 hours)

Book of Abstracts

Sponsors: INTAS PlasmaGen Biosciences

Program

Day 1: **31st OCT 2020** **(Saturday)**	**Topic**	**Speaker/panelist**	**Chairpersons**
16:00-16:10	**Introduction and welcome**	Dr. Saji Philip	
16:10-16:25	**Remembering Dr. Tomisaku Kawasaki**	Dr. Surjit Singh	
16:30-17:15	**KD epidemiology in India- a viewpoint** **(10 minutes each)**	North: Dr. Vignesh Pandiarajan West: Dr. Vijay Viswanathan South: Dr. Sagar Bhattad East: Dr. Priyankar Pal	Dr. Nutan Kamath, Dr. Snehal Kulkarni
17:15-17:45	**Diagnosis of Kawasaki Disease** The 2017 AHA criteria- *a critique* The enigma of incomplete and atypical KD	 Dr. Avinash Sharma Dr. Manjari Agarwal	Dr. Amitabha Chattopadhyay Dr. Sikha Agarwal
17:45-18:15	*My most memorable case of Kawasaki Disease*	Dr. Anand P. Rao Dr. Ankur Jindal Dr. Dhrubajyoti Sharma	Dr. Jyothi Raghuram / Dr. Mushtaq Bhat
18:15-19:00	***Imaging in Kawasaki Disease*** *Role of 2-D Echocardiography* *Controversies regarding coronary artery 'Z' scores* *Newer imaging modalities in KD*	 Dr. Saji Phillip Dr. Nageswara Rao Dr. Manphool Singhal	Dr. Debdatta Mukherjee / Dr. Sanjeev Naganur
Day 2: **1st NOV 2020** **(Sunday)** 16:00-16:50	**Management issues** Aspirin: Where does it stand? Steroids Infliximab Statins	 Dr. Tapas Kumar Sabui Dr. Sujata Sawhney Dr. Deepti Suri Dr. Sathish Kumar	Dr. Narendra Bagri / Dr. Rashna Dass
16:50-17:20	***Management of IVIG resistant Kawasaki Disease: Taiwan Experience***	Dr. Ming -Tai National University Hospital Taipei ,Taiwan	Dr Bhaskar Shenoy / Dr. Sharath Kumar
17:30-18:30	**3rd Dr. Tomisaku Kawasaki Oration** *Pathology of cardiovascular lesions of Kawasaki Disease*	Dr. Kei Takahashi Chairperson, Japanese Society of Kawasaki Disease	Dr. Saji Philip / Dr. Priyankar Pal
18:30-19:00	***Free paper Presentations*** **(5 minutes x 5)**		Dr. R. Ravichandran / Dr. Latika Gupta
19:00-19:30	***Symposium on PIMS / MISC***	Dr. Chandrika Bhat Dr. Rakesh Mondal Dr. Aruna Bhat Dr. Mahesh Janarthanan Dr. Suparna Guha	Dr. Suma Balan
19:30-19:45	**Handing over the baton** Message from incoming President Announcement of venue of 4th Annual Conference of ISKD 2021 *Vote of Thanks*	Dr. Saji Philip / Dr. Surjit Singh	 **Organizing Secretaries:** Dr. Ankur Jindal Dr. Nageswara Rao
19:45	**Adjourn**		

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Message

We started our journey in 2010 by founding Kawasaki Disease foundation in India for the purpose of better awareness of this life-threatening disease affecting coronary arteries leading to myocardial infarction.

Our vision is to help our desperate parents with children on life term treatment for coronary artery aneurysms by counselling and health education and also to provide better coronary care in India. As the incidence of KD and non-responders to IVIG are increasing, a Society for KD was needed. This idea sprouted in the mind of Prof Surjit Singh and further supported by Dr Sagar Bhattad during 5th KD summit held at Bangalore in 2017 to take care of major issues such as our own criteria to diagnose KD in India. First ISKD meeting was then held at Chandigarh in 2018 and got registered finally at Kolkata in 2019along with the 2nd ISKD organised by Prof. Priyankar Pal, Prof Tapas Kumar Sabui and Prof. Rakesh Mondal.

I am glad that this 3rd Virtual IKDS symposium will enlighten the diagnosis and management of KD including epidemiology and other diagnostic criteria issues to update newer developments in the field of research in Kawasaki syndrome. On behalf of ISKD, I welcome all delegates for this symposium. I appreciate Dr. Nageswara Rao and Dr Ankur Jindal for their hard work to formulate this International congress on KD.

Dr Saji Philip

Clinical coordinator & Pediatric Cardiologist

President of Indian Society of Kawasaki Disease

Dr K M Cherian Heart Foundation, Parumala, Kerala

Message

Dear Friends and Teachers,

On behalf of the Organizing committee a very warm welcome to the 3rd annual conference of the Indian Society for Kawasaki disease. Although the ongoing pandemic has restricted physical meetings, it has taught us to explore the hitherto unexplored possibility of meeting virtually.

'The old order changeth yielding place to new'.And thus the e conference was born and till date we have about 650 registrations (including several international delegates) and 33 abstracts for free papers. That is a huge jump in terms of numbers from the last 2 conferences.

Even after 53 years since the first publication and making it's way to be the commonest childhood vasculitis and number one cause of acquired heart disease, the disease continues to be missed, and regarded with some cynicism.I sincerely hope that the conference would create further awareness about the enigmatic disease.

Warm regards

Dr Priyankar Pal

Secretary

Message

Dear Colleagues:

I hope you and your family are doing well as we continue to navigate around COVID-19.

It is my great pleasure of inviting you to participate in 3rd Annual conference of Kawasaki Disease on 31st October and 1st November. This year conference is unique due to unfortunate pandemic. Many of us suffered directly or indirectly in several ways. However, the enthusiasm towards learning and sharing knowledge is utmost important in our profession.

I assure you that this annual meeting is going to be memorable to all of us.

We look forward to meeting you on a virtual platform soon.

Best Regards

Dr. Nageswara Rao K
Organising Secretary

ATYPICAL KAWASAKI DISEASE

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3rd Year post graduate resident (MD Pediatrics)
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Introduction:

Atypical Kawasaki disease usually affects infants and toddlers and, due to delayed diagnosis and treatment, is strongly associated with an increased risk of permanent heart damage. Among the possible clinical presentations of atypical KD, isolated lung involvement is considered a very uncommon feature and most cases reported in literature involve children in the first year of life.

Case:

3.5 year old male child was admitted with complaints of high grade fever since 10 days associated with a generalized erythematous rash. Examination revealed fissured lips, perianal excoriation, cervical lymphadenopathy and oedema over hands and feet with decreased right sided air entry.Radiography showed right middle and lower lobe consolidation with hemogram showing neutrophilic leucocytosis for which IV antibiotics were given. Not responding, he was further worked up,lab parameters showed elevated Pro-BNP, CRP and ESR, hyponatremia and hypoalbuminemia.2D ECHO showed coronary artery dilatation (LAD , Z score 3.7).Witha working diagnosis of Atypical Kawasaki disease, he was given IVIG @2g/kg and started on high dose Aspirin @80mg/kg/day. Fever and symptoms subsequently resolved, and the patient was discharged after 8-days hospitalization.

Conclusion:

A high index of suspicion is required in children with refractory pneumonia, fever and persistently high inflammatory markers. Pediatricians should consider atypical Kawasaki disease as a possible alternative diagnosis, because early clinical suspicion and prompt treatment with IVIG and aspirin dramatically abate the high risk of permanent coronary damage. Increasing awareness about this condition is of utmost importance as it continues to be a challenge.

AN UNCOMMON OVERLAP OF KAWASAKI DISEASE AND HENOCH-SCHONLEIN PURPURA

Akshi Sharma, MBBS; **Sandesh Guleria**, DM; **Parveen Bhardwaj**, MD; **Surinder Singh**, MD
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Introduction

Kawasaki disease (KD) and Henoch-Schonlein purpura(HSP) are themost common childhood vasculitic disorders. Overlap of 2 rheumatological disorders has been described but overlap of KD and HSP is extremely unusual.

Case report

A 7-year-old girl presented with fever for 10 days, swelling multiple joints and erythematous rash on lower limbs for 5 days. On day 2 of fever she developed conjunctival injection, which subsided after 3 days. She also had history of strawberry tongue. There was no history of pain abdomen and hematuria. On examination she was irritable, febrile. She had palpable purpura on lower limbs (Fig. 1A and B), non-pitting edema on hands and feet (Fig. 1D), red cracked lips (Fig. 1E), left posterior cervical lymphadenopathy (largest lymph node 2x1 cm), swelling bilateral ankle, knee (Fig. 1C) and wrist joints with tenderness and restriction of movements. On day 5 of admission she developed periungual desquamation (Fig. 1F).

Laboratory investigations showed neutrophilic leucocytosis, elevated erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) and normal platelet counts. Skin biopsy revealed leucocytoclastic vasculitis. Two-D echocardiography showed medium aneurysm in right coronary artery (5 mm, 6.5z) and left main coronary artery (5 mm, 6.1 z).

Result:

A clinical possibility of KD and HSP overlap was considered and she was initiated on oral aspirin (50 mg/kg/day) and was given intravenous immunoglobulin (2 gm/kg). Her fever and irritability subsided in next 24 hours and arthritis and rash in next 4 days. She was discharged on low dose aspirin (3 mg/kg/day) and at 2 weeks of follow up she was well with no recurrence of rash and coronary artery aneurysms with similar dimensions as on discharge.

Conclusion

Two or more rheumatological disorders may coexist at the same time but an overlap of KD and HSP is extremely rare and one must always be vigilant.

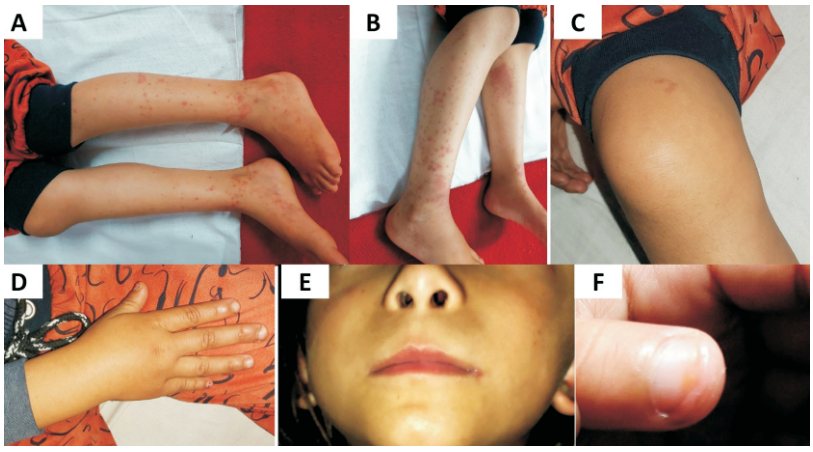

Fig. 1A and B, palpable purpura on lower limbs in the index child; C, swelling in left knee joint; D, non-pitting edema on hands; E, red cracked lips; F, periungual desquamation

AGE IS NO BARRIER TO KAWASAKI DISEASE

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Introduction

Kawasaki disease (KD) is a common childhood vasculitis, mainly affecting children between 2-5 years of age. It has rarely been reported in infants less than 6 months of age. Infants with KD have higher incidence of incomplete form of KD as well as coronary artery abnormalities (CAAs).

Case Report

Two-month-old male infant presented with high grade fever for 7 days. On examination, he was irritable, had fever (temprature 102° F), conjuctival injection, generalized maculopapular rash and bilateral cervical lymhadenopathy. On day 2 of admission, he developed bilateral periungual desquamation. Investigations revealed anaemia (hemoglobin 9.6 g/dl), neutrophilic lucocytosis (total leucocytes 25200/dl; P_{82} $L_{16}M_2$), thrombocytosis (9,00,000/dl), elevated erythrocyte sedimentation rate (100 mm in 1st hour) and C-reactive protein (200 mg/L). He also had elevated transaminases and sterile pyuria. His blood culture was sterile. Two-dimensional echocardiography showed giant aneurysms in proximal left anterior descending coronary artery (5 mm; 14.1z) and ecstatic left main and right coronary artery.

Result

Kawasaki disease with giant coronary artery aneurysm was considered and he was given intravenous infliximab (8 mg/kg), intravenous immunoglobulin (2 gm/kg) and oral aspirin (30 mg/kg/day). His irritability and fever subsided in next 24 hours and inflammatory parameters decreased over next 7-10 days. He was discharged on subcutaneous enoxaparin (1 mg/kg 12 hourly), low dose aspirin (5 mg/kg/day) and was kept on follow up.

Conclusion

No age is barrier to KD. Younger age (infancy) is a risk factor for CAAs and may also pose diagnostic challenge due to rarity of KD in children < 6 months of age.

ATYPICAL KAWASAKI DISEASE WITH DENGUE CO-INFECTION – A DIAGNOSTIC DILEMMA.

Arun Kumar N, Jyothi Raghuram
Columbia Asia Hospital, Whitefield, Bangalore

Introduction

Kawasaki disease (KD) is an acute inflammatory vasculitis and a leading cause of paediatric acquired heart disease.Atypical or incomplete forms of KD are common (15–20%).We report a case which initially presented as serologically confirmed dengue fever who subsequently developed atypical KD.

Case report

A 7 years old female child presented with history of fever , rash ,vomiting and pain abdomen since 6days,with serologically confirmed dengue fever showed expected improvement initially only to develop high grade fever spikes with oral ulcers and mucositis which also subsided in 36 hours without specific treatment. Secondary bacterial infection, macrophage activation syndrome, Kawasaki disease and COVID-19 co-infection/MISC were considered and child was evaluated for the same. Criteria for incomplete KD were met once during the course of illness but in view of clinical improvement, parents did not consent for IVIg infusion. Close follow-up revealed increasing coronary dilation 7 days later and IVIg was given.

Key message

The diagnosis of KD with dengue co-infection is challenging as clinical features of both the diseases overlap. We wish to emphasise the very short duration of acute phase of KD in this child with dengue fever and incomplete KD.

Key words

Atypical Kawasaki disease; dengue fever; children.

A STUDY TO EVALUATE THE E.C.G CHANGES IN CHILDREN WITH KAWASAKI DISEASE

Dipsikha Jana*, Tapas Kumar Sabui, ;BiswajitMajumdar*****
*3rd yr PGT, H.O.D.,**Department of Pediatric Medicine, ***Associate Professor
Department of Cardiology.R.G.Kar Medical College and Hospital.

Objective

1. Any E.C.G. abnormalities in the acute phase of Kawasaki Disease.
2. Comparison of the E.C.G. changes between the complete and incomplete Kawasaki .
3. Relationship between different acute inflammatory markers of blood with E.C.G. changes in acute phase.
4. Effects of Coronary changes on echocardiography upon E.C.G.changes in Acute phase.

Study Design

Hospital based Case Control Observational Study.

Subjects

Case :All patient admitted in Pediatric Department diagnosed to have Kawasaki disease.

Control

Same no of age and sex matched healthy control.

Sample size:39 cases and 39 controls.

Time Line

January 2019 to June 2020.

Results

The mean of heart rate,P wave voltage,QTc interval,QT dispersion,Tpeak-Tend dispersion ,the percentage of right axis deviation and left axis deviation were significantly higher but PR interval in case group was significantly shorter in case group . ECG abnormalities are insignificantly more evident in Incomplete Kawasaki Disease case group as compared to Complete Kawasaki Disease. Most significant correlations were with Aneurysm, Platelet count, ESR, CPK-MB and LDH .In the follow ups the decrement of the mean heart rate and PR interval ,QTc interval ,QT dispersion,Tpeak-Tend dispersion were notsignificant until 2nd follow up .

Conclusion

Most of the E.C.G. findings obtained in acute phase are significantly abnormal showed insignificant improvement in the follow ups .

KAWASAKI DISEASE IN COVID PANDEMIC ERA!!

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Introduction

Kawasaki disease is an inflammation of medium vessel seen in usually seen in pediatric population under 5 years of age. The usual peak onset is from 18 to 24 months with long term complications such as corona artery dilatation[1]. Several reports have emerged in 2020, a inflammatory syndrome similar to Kawasaki diseaseduring the COVID-19 pandemic, challenging previous observationsthat children have mostly mild disease[2]. Currently two guidelines are used worldwide for the diagnosis of complete Kawasaki that is AHA(American heart association) and JMH (Japanese Ministry for health). As per the current guidelines IVIg @2gm/kg and also oral aspirin are used for the treatment of Kawasaki diseasei[1]. Despite timely initiation of treatment KD children have increased risk of developing coronary artery aneurysm[3] and also risk of developing ivig resistance[4].

Case

2 years old female child was presented to us with complaints of moderate to high grade fever spikes for9 days along with maculopapular,erythematous rash which appeared first over bilateral limbs and gradually progressed tobilateral upper limbs. Later she also developed 4, Peri-anal and peri-genitalexcoriation with fissured lips and strawberry tongue. On Day6 of illness mother also noticed redness of both eyes with no discharge which subsided with unilateral preauricular lymphadenopathy.CRP, ESR and D dimerlevels were elevated. COVID rt-PCR documented was negative. She received IVIG @2gm/kg and was started on oral high dose Aspirin. She responded well to the above line of management and gradually feverspikes and rash subsided. 2D Echo done showed no coronary abnormalities.

Conclusion

The above description typically suggests a classic case of Kawasaki disease but with the advancement of laboratorial tools the major question arise that why there is a lack of clinical trials on inflammatory markers and association with KD. Why there is a association of coronary artery with the cytokinin storm?More work remains to be done, especially in long-term follow-up of pediatric COVID-19 survivors.

Reference

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2. Kam KQ, Ong JS, Lee JH. Kawasaki disease in the COVID-19 era: a distinct clinical phenotype?. The Lancet Child & Adolescent Health. 2020 Sep 1;4(9):642-3.
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4. Baek JY, Song MS. Meta-analysis of factors predicting resistance to intravenous immunoglobulin treatment in patients with Kawasaki disease. Korean journal of pediatrics. 2016 Feb;59(2):80.

SETTING THE STAGE FOR GENETIC BIOMARKERS OF KD: FIRST STUDY ON MIRNA PROFILE OF CHILDREN WITH KAWASAKI DISEASE FROM INDIA

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Postgraduate Institute of Medical Education and Research, Chandigarh

Introduction

Genetic biomarkers have been increasingly explored in association with KD. MicroRNAs are a group of small noncoding RNAs that regulate gene expression. In KD, differential expression of miRNAs has been studied for diagnosis and predicting severity and response to therapy.

Objectives

To assess miRNA profile in patients with Kawasaki disease in comparison with febrile controls using microRNA-145-5p, miRNA-320a and miRNA-499

Method

We enrolled patients fulfilling the American Heart Association 2017 criteria for the diagnosis of KD. These children were diagnosed and later followed up in Pediatric rheumatology clinic at Advanced Pediatrics Centre,PGIMER, Chandigarh between January 2019 to June 2020. On the basis of available literature two miRNA were chosen (microRNA-145-5p, miRNA-320a). These miRNAs have been shown to have higher expression during acute phase of KD in Chinese and Japanese population. Further, one miRNA which has been studied in association with viral myocarditis (miRNA-499) was included in the study to evaluate its utility as potential biomarkers in acute KD. miRNA isolation was done from the plasma during acute state followed by RNA quantification and cDNA synthesis. This was followed by Real time PCR for quantification and analysis of miRNA expression.

Results

Twenty children with a diagnosis of KD were recruited for the study; 2 had coronary artery abnormalitiesand compared with 20 age matched febrile (non KD illness) controls. Comparison between febrile controls and patients showed significant decrease in levels of miR145 in our cohort of patient with KD (p=0.005) as compared to febrile controls, significantly reduced levels of miR-320a during acute stage of the disease (p= 0.05) as compared to febrile controls and no difference of expression in febrile and disease group with regard to miRNA 499 (p= 0.54).

Conclusion

This was the first study in the Indian population to explore the miRNA profile in patients with KD. There was significantly decreased expression of miR-145 and miR-320-a in our cohort of patients with KD in comparison to febrile patients with other illnesses. There was no significant difference in expression of miR-499 in patients with KD in comparison to febrile patients with other illnesses. This may be due to different genetic characteristic of Indian population as compared to Japanese, Chinese and American populations. Further studies are needed to establish these as potential biomarkers for diagnosis and prediction of complication and response to therapy in patients with KD.

HYPERINFLAMMATORY SYNDROME IN CHILDREN IN COVID ERA
A Case Series of Ten Children From Tertiary Care Centre of Sub Himalyan Region

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Hyper-inflammatory syndromes are life-threatening disorders caused by overwhelming host immune response often resulting from defects in negative feedback mechanisms. These include Classical Kawasaki disease (KD), Incomplete KD, KD shock syndrome, toxic shock syndrome (TSS), macrophage activation syndromes (MAS) and multisystem inflammatory syndrome in children (MIS-C).We are describing a case series of 10 patients with hyper-inflammatory Syndrome highlighting clinical presentations,laboratory profiles with management and outcome during this COVID-19 pandemic to raise awareness among paediatrician.

Case Series

Ten children between 1 month to <19 years, admitted to pediatric intensive care unit with features of hyper inflammation and multiorgan failure were included in study. The detail history , physical examination findings, chest x ray or ECHO results and laboratory investigations including complete blood count with differential count, ESR, CRP, coagulation profile, ferritin, liver enzyme, lactate dehydrogenase, triglycerides, D dimer were recorded. Nose and throat swab viral PCR (polymerase chain reaction) for SARS-CoV-2 of all children were sent.

The median age was 13.5 years . Seven out of 10 were male. All these patients were critically sick with muco-cutaneous manifestations like non purulent conjunctivitis,cracked lips or edema of hands or feet,fulfilling the partial criteria for KD according to 2017, AHA guidelines. All had hypotension which was consistent with Kawasaki disease shock syndrome.Nine met the MAS criteria by Paediatric Rheumatology International Trials Organisation. In clinical profile 5 had respiratory distress,2 had pericarditis and 1 showed aortic valve regurgitation .Two had acute motor sensory axonal neuropathy (AMSAN) . Only two (20%) out of 10 had GIT manifestation in form of loose stools and vomiting. All children were treated as per protocol. Seven children died and 3 were discharged home.

The importance of suspecting hyper inflammatory disease in children during this pandemic cannot be overemphasized.These patients can deteriorate very rapidly.Hence, Early recognition and prompt treatment are essential for better outcome.

NEUTROPHIL: LYMPHOCYTE RATIO PREDICTS RESISTANT KAWASAKI DISEASE- A PROMISING BIOMARKER FOR DEVELOPING COUNTRY SETTING

Jaikumar Govindaswamy Ramamoorthy[1], Avinash Anantharaj[2], Dhandapany Gunasekaran[1], Prakash Duraisamy[1],Saragondlu Lakshminarasappa Darshith[1], Pediredla Karunakar[1], Venkataraman Ranjith Kumar[1]

Department of [1]Pediatrics, Jawaharlal Institute of Postgraduate Medical Education and Research (JIPMER), Puducherry, India
Department of [2] Cardiology, Jawaharlal Institute of Postgraduate Medical Education and Research (JIPMER), Puducherry, India

Introduction

Persistence of fever in Kawasaki Disease (KD) for minimum 36 hours post completion of Intravenous Immunoglobulin (IVIG) infusion is termed as resistant Kawasaki Disease (rKD)[1]. It can be predicted using multiparametric scoring systems like Kobayashi, Sano and Egami with 75-85% sensitivity and specificity. Accuracy of these models outside Japanese population is debatable[1]. Neutrophil: Lymphocyte ratio(NLR) has evolved as a promising biomarker in predicting rKD in East-Asian population [2] . We share 2 cases with rKD and our experience with NLR performed 24 hours post-IVIG completion in rKD prediction.

Case Details

Case 1: A 3 years old well thriving boy presented with fever without foci for 12 days. Examination was unremarkable with normal anthropometry. Evaluation for tropical infection, bacterial sepsis, tuberculosis and novel corona virus were non-contributory. Investigation findings are depicted in table-1. Echocardiogram showed small aneurysm of left anterior descending artery. There was persistent fever 36 hours after completion of IVIG infusion. There was progressive aneurysm development in left main coronary artery (LMCA). Child was treated with second dose of IVIG and achieved deferevescence in 12 hours. Repeat echocardiogram 2 weeks after second IVIG dose showed normalisation of coronary dimensions.

Case 2: A 2 months infant presented with fever for 2 days, poor feeding and lethargy for 1 day. Systemic examination was unremarkable except diffuse erythematous maculopapular rash and Bacillus-Calmette-Guerin vaccine site erythema. The details of investigation are provided in table-1. Echocardiogram showed small aneurysm of LMCA. There was persistent fever 36 hours after completion of IVIG infusion. There was progressive development small aneurysm of right coronary artery. The infant achieved defervescence in 18 hours after second IVIG infusion. Repeat echocardiogram 2 weeks after second IVIG dose showed normalisation of coronary dimensions.

Discussion

NLR depicts the balance between ongoing inflammation and counter-regulatory immune response. During acute/active phase of KD there is intense infiltration of neutrophil in coronary arteries, hence persistently higher NLR despite IVIG indicates ongoing inflammation and/or lack of counter regulatory immune response[3,4]. The cut-off point for NLR post-IVIG infusion to predict rKD ranged from 1.26-1.45 with a specificity ranging from 62%-87% and the area under curve(AUC) was 73%-86%[2]. In a meta-analysis of 7 studies NLR was found to be 77% specific in predicting the development of rKD with AUC of 84%[2]. NLR >1 post-IVIG infusion was also predictive of development and progression of coronary artery abnormality (CAA) in children[5]. Persistently high NLR 24 hours post-IVIG infusion flagged impending rKD in both the cases.

Conclusion

NLR is very simple and non-cumbersome than many complex scoring systems. Prospective studies in our population are needed to validate its predictive capacity.

Table 1: Laboratory investigation and echocardiogram findings

Case No.	Status	NLR	ESR (mm/hr)	CRP (mg/dl)	Platelet count ($\times 10^3/\mu l$)	Total leucocyte count ($\times 10^3/\mu l$)	AST (U/L)	Echocardiogram (Z score)
Case 1	Pre-IVIG	4.2	140	12	872	27.3	26	LMCA-2.43 LAD-2.53 LCX-1.83 RCA-1.8
	Post-first IVIG	4	-	11.6	864	27.9	24	LMCA-3.4 LAD-2.48 LCX-1.9 RCA-1.76
Case 2	Pre-IVIG	2.9	47	4.8	858	28.6	17	LMCA-3.1 LAD-2.37 LCX-1.61 RCA-1.93
	Post-first IVIG	2.5	-	4.6	891	27.6	20	LMCA-2.6 LAD-2.1 LCX-1.6 RCA-3.1

NLR- Neutrophil: Lymphocyte Ratio; ESR- Erythrocyte sedimentation rate; CRP- C-Reactive protein; AST- Aspartate Transaminase; IVIG- Intravenous Immunoglobulin; LMCA- Left main coronary artery; LAD- Left Anterior Descending artery; LCX- left circumflex artery; RCA- Right coronary artery.

ORAI1 GENE POLYMORPHISMS IN CHILDREN WITH KAWASAKI DISEASE: A STUDY FROM NORTH-WEST INDIA

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Introduction

Pathogenesis of Kawasaki disease (KD) remains an enigma and new insights have suggested genetic predisposition of KD. ORAI1 is a calcium (Ca2+) channel protein involved instore-operated Ca^{2+} entry pathway required of activation of lymphocytes. Association of ORAI1 gene with risk of KDwas reported in Japanese children. We evaluated polymorphisms in ORAI1 gene in Indian children with KD.

Methodology

This is an observational study conducted from June 2018 to December 2019 enrolling children with KD. As baseline prevalence data of ORAI1 polymorphism in north Indian population is not available, 50 healthy adult controls were also enrolled for the study. Patients with KD weredivided into KD with coronary artery abnormalities (KD with CAA) and KD without CAA for sub-analysis. DNA was extracted and amplified by conventional polymerase chain reaction technique using primer of exon 2 of ORAI1. Sequencing of DNA was, then, done using Sanger's chain termination method.

Results

Fifty children with KD and healthy adult controls each were enrolled with 25 children having CAA.Demographic data are depicted in Table 1. Three different polymorphisms in ORAI1 gene were identified viz. rs3741596 at c.652A>G p.Ser218Gly, rs3825175 at c.798T>C p.Thr266Thr and rs3741595 at c.546 C>T p.Ile182Ile. The AA and AG genotype of rs3741596 were found in 45 (90%) and 5 (10%) respectively in KD group. Whereas, all the 50 controls had AA genotype. There was statistical significant difference ofvariants of rs3741596 between the two groups (p-0.022). Whereas, no statistically significant difference was observed forpolymorphisms of rs3825175 and rs3741595between the two groups. Polymorphisms of ORAI1was also not significantly differentbetween KD with CAA and without CAA.

Conclusions

This is the first study in Indian population to explore the association of KD with ORAI1 gene. The study provides evidence to supportassociation of polymorphism of ORAI1with KD susceptibility. Further studies with a larger sample size are required to confirm this finding.

Table 1: Demographic profiles and investigation findings of KD cases

Parameters	**Results of patients with KD (n=50) Mean (SD)**
Mean age	3.9 (2.53) years
Sex	36 (72%) male, 14 (28%) female
Hemoglobin (g/L)	103 (18.9)
Total leucocytes (x 10^9 cells per L)	13.5 (7.24)
Neutrophils (%)	57 (17.8)
Lymphocytes (%)	36.3 (20.6)
Platelets (x 10^9 cells/L)	474 (189)
ESR (mm in the first hour)	50.9(30.5)
CRP (mg/L)	47.98 (41.78)
Serum albumin (g/dl)	3.24 (1.1)
NT-proBNP (pg/ml)	895.8 (1286)
Coronaries diameter Z score	Left main coronary artery: 1.28 (1.70) Left anterior descending artery: 0.81 (2.09) Left circumflex artery: -0.29 (1.78) Right coronary artery: 0.36 (2.12)

INCOMPLETE KAWASAKI DISEASE CAMOUFLAGE AS INTESTINAL PSEUDO-OBSTRUCTION

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A 3-month-old female infant presented with fever and bilious vomiting for 2 days. She was febrile with a distended abdomen and decreased bowel sounds. X-ray abdomen revealed dilated bowel loops and serum potassium(4.9 mEq/L) was normal. Surgical causes were ruled out, a provisional diagnosis of septicemia with intestinal pseudo-obstruction was considered and empirical antibiotics were administered. BCG vaccination site erythema (BCG-itis) was noted on Day 3 which prompted us to suspect Kawasaki Disease (KD). Investigations revealed elevated C-Reactive Protein(4.8mg/dL), erythrocyte sedimentation rate(100mm/1st hour), normocytic anemia(7.9g/dL), leucocytosis(27x10^{9}/L), thrombocytosis (604x10^{9}/L), sterile pyuria(300 WBC/HPF), hypoalbuminemia(2.8g/dL). Blood and urine cultures were sterile. Echocardiogram on day 4 revealed dilation of left main coronary artery(2.5mm,+3.08Z) & distal segment of the right coronary artery(1.7mm,+3.00Z) confirming incomplete KD. She was treated with intravenous immunoglobulin (IVIG) and aspirin. As fever persisted even after 36 hours, resistant KD was diagnosed; IVIG was re-administered following which defervescence occurred. Echocardiogram on follow up revealed no residual coronary artery abnormalities(CAA). Our case highlights the point that KD can present as acute intestinal pseudo-obstruction and thrombocytosis and CAA can appear within the first week(as in our case) which usually occur after 7 days of fever. The axiom of a young infant with fever> 7 days without focus should be investigated for KD, may not be entirely valid as it could delay the diagnosis, impeding the goal of preventing CAA. BCG-itis is an early and important marker of KD.

CLASSIC KAWASAKI DISEASE

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Introduction

Kawasaki disease is an acute systemic vasculitis and is the most common cause of acquired heart disease in children in the developed world. The disease is believed to result from a genetically susceptible individual's exposure to an environmental trigger. Incidence is rising worldwide, and varies widely across countries and within different ethnic groups. It has surpassed rheumatic heart disease as the leading cause of acquired cardiovascular disease in children in the developed world.

Case

4.5 year old male child was admitted with complaints of high grade fever since 6 days associated with a generalized erythematous rash. Examination revealed fissured lips, perianal excoriation, cervical lymphadenopathy and oedema over hands and feet.. Not responding to antibiotics, he was further worked up,lab parameters showed elevated CRP and ESR, hyponatremia and hypoalbuminemia.2D ECHO showed coronary artery dilatation (Z score LAD 4.31, RCA3.55).Witha working diagnosis of Classic Kawasaki disease, he was given IVIG @2g/kg and started on high dose Aspirin @80mg/kg/day. Fever and symptoms subsequently resolved, and the patient was discharged after a 10 day hospital stay.

Conclusion

Given its severe morbidity and potential mortality, Kawasaki disease should be considered as a potential diagnosis in cases of prolonged paediatric fever.Early clinical suspicion and prompt treatment with IVIG and aspirin dramatically abate the high risk of permanent coronary damage. Increasing awareness about this condition is of utmost importance and it continues to be a challenge for pediatricians to this day.

KAWASAKI DISEASE WITH CONCOMITANT CHICKEN POX: AN UNUSUAL OCCURRENCE

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Introduction

Kawasaki disease (KD) is a common childhood vasculitis. The etiology of KD is still unknown but various infections have been implicated as trigger for it.

Case report

A 7-year-old girl presented with high grade fever and rash for 5 days and redness of eyes for 2 days. On day 1 of illness she developed erythematous papules and some fluid filled vesicles on abdomen, which progressed to involve trunk, face and limbs in next 48 hours and later had evolved into scabs. She also developed conjunctival injection on day 3 of illness. There was H/O chicken pox in one of sibling 10 days prior to her illness.

On examination,She had generalized papulo-vesicular rash with few lesions with scabs (Fig. 1A), cracked red lips, strawberry tongue (Fig. 1B), conjunctival injection (Fig. 1C) and bilateral cervical lymphadenpathy (multiple nodes, firm, painless and largest lymph node 2x1 cm). On day 3 of admission, she developed perianal and periungual desquamation (Fig. 1D).Investigations revealed hemoglobin 103 g/L, total leucocyte counts 15.57 x 10^9/L (P_{92} L_6 M_2), platelets 114 x 10^9/L, C-reactive protein 168.5 mg/L and erythrocyte sedimentation rate 19 mm in 1st hour. Serological test for varicella immunoglobulin M (IgM) was also positive.

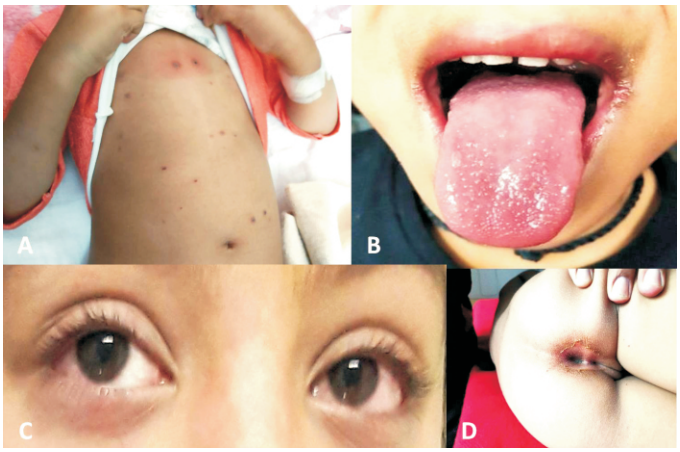

Figure-1: A, Papulovesicular rash with scabs in the index child; B, red cracked lips with strawberry tongue; C, conjunctival injection; D, characteristic perianal desquamation.

Result

Based on clinical features and laboratory investigations, diagnosis of chicken pox with concomitant KD was considered and the child was administered intravenous immunoglobulin (2 gm/kg) over 24 hours. Her fever subsided in next 24 hours. Two-D echocardiography of coronary arteries was also normal. She was discharged on low dose aspirin (4 mg/kg/day) and kept on follow up.

Conclusion

KD triggered by varicella infection has been reported earlier but concomitant chicken pox and KD has rarely been reported.The etiology of KD is still elusive but concomitant occurrence of various infections can help in elucidating it.

A COMPARATIVE STUDY OF IL6 AND CRP LEVELS IN POST COVID PEDIATRIC HYPERINFLAMMATORY MULTISYSTEM SYNDROME (PIMS) AND KAWASAKI DISEASE PATIENTS.

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Interleukin 6 (IL 6) is a pleotropic cytokine produced in response to tissue damage and infections. It helps regulate immune responses and has been shown to be elevated in certain diseases, like Kawasaki disease(KD) and the recent pandemic of COVID 19. PIMS is a recently reported hyperinflammatory syndrome seen in the paediatric age group affected with COVID 19 (SARS CoV 2). A single center analytical study was conducted comparing IL 6 & C- Reactive Protein(CRP) levels between 72 patients of KD with Coronary artery lesions(CALs) collected over a period of 18 months (1) in pre COVID era(Jan 2017 to June 2018) and 25 patients of PIMS (since July 2020) admitted at the Institute of Child Health Kolkata.

Among 72 cases of KD admitted during the study period,22 cases had CALs. The mean IL 6 level observedwas 143.6 pg/ml (normal< 7pg/ml), which was significantly lower than the mean IL 6 levels in PIMS (n = 25) which was 184.1 pg/ml. The difference was found to be statistically significant (p value<0.05) using the t-test for comparing means, computed using the scipy library in Python. The mean CRP values in KD patients with CALs was 135.17 mg/L (normal < 5mg/L) where the difference was statistically significant when compared to the mean CRP value in PIMS patients which was 181.37 mg/L., using the same test. However, considering the less than appropriate sample sizeand assumptions of the test used, there remains a margin of error. (For the tests, a significance level of 5% was used)

In conclusion,it can be said that both IL 6 and CRP levels are significantly higher in PIMS patients as compared to KD and can be used as a predictor for distinguishing KD from the KD like new entity PIMS.

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EVALUATION OF PIMS-TS VS MIS-C DIAGNOSTIC CRITERIA IN COVID 19 ASSOCIATED HYPERINFLAMMATION – DEVELOPING COUNTRY PERSPECTIVES

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Introduction

Pediatric Inflammatory Multisystem Syndrome Temporally associated with SARSCoV2 (PIMS-TS) and Multisystem Inflammatory Syndrome in Children (MISC) are the two terminologies for COVID 19 associated hyperinflammation given by Royal College of Pediatrics and Child Health (RCPCH) and World Health Organization (WHO) respectively.

Objective

To compare the performance of PIMS-TS andMISC as diagnostic tool in evaluation of childrenadmitted with COVID 19 associated hyper inflammation.

Methods

We conducted a multicentred prospective observational study recruiting children with COVID 19 related hyper inflammation between age group of 1 month to 12 years from three tertiary care hospitals of Kolkata from May 2020 to August 2020. PIMS -TS and MIS- C criteria were applied in those patients. Comparison was done based on the identification of patients, clinical &laboratory parameters and outcome differences between Group A (fulfilling both PIMS -TS & MIS-C criteria) and Group B (those fulfilled only PIMS-TS criteria).

Results

Out of total 35 patients with features of COVID19 associated hyperinflammation, 32 patients satisfied criteria for PIMS -TS and 21 patients fulfilled criteria for MIS C(p value 0.001). COVID 19 RT-PCR was positive in 50% of patients while IgG antibody against COVID 19 was found in 62.5%. Shock, Ejection fraction <55% and inotropic support showed significant association (p value <0.05) with Group A whereas the milder cases were not picked up by MIS-C criteria.Only death of PIMS-TS also fulfilled MIS-C criteria.

Conclusion

PIMS-TS is better diagnostic tool as it detectsgreater number of patients and milder cases of COVID19 associated hyperinflammation thus helping early referral from resource poor peripheral centre in developing country setting.

RISK FACTORS IN IVIG RESISTANT KAWASAKI DISEASE AND CORRELATION WITH JAPANESE SCORING SYSTEMS

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Introduction

The purpose of this study was to assess the risk factors of IVIG resistant KD and to evaluate the performance of the three Japanese risk scoring systems, namely the Kobayashi, Egami and Sano scores in predicting IVIG resistance.

Methodology

This is a prospective observational study of children admitted with KD at Institute of Child Health,Kolkata for a period of 16 months, from January 2019 to April 2020. This study included 70 children with KD all of whom were treated with IVIG. Important clinical parameters, laboratory variables and risk scores were compared between the IVIG responsive group and the IVIG resistant group.

Results

I. 31.4% (n=22) were non-responders.

II. Skin rash was found to be significantly associated with IVIG resistant KD (p value = 0.0431).

III. The IVIG resistant group had higher total bilirubin (p value=0.045), lower albumin (p value=0.002), higher CRP levels (p value=0.0020) and higher ALT and AST levels (p=0.0379, p=0.0029 respectively).

IV. High Kobayashi score (p value = 0.0009), high Egami score (p value=0.0106) and high Sano score (p value=0.0262) were significantly associated with IVIG resistance, individually.

V. Sano score had the highest sensitivity (81.8%). Kobayashi score had the highest specificity (77.1 %).

Conclusion

I. The presence of skin rash, high total bilirubin, high CRP, high AST, high ALT and low albumin are important predictors of IVIG resistance in our population.

II. Amongst the three scores, Sano score is the most reliable in identifying potential non-responders to IVIG. But Sano score lacked good specificity. Therefore Indian KD patients may need an exclusive scoring system to predict non responsiveness to IVIG so that a more aggressive therapy can be instituted at the earliest.

KAWASAKI DISEASE WITH DENGUE (IgM) FEVER

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Introduction

It is important to recognize that' incomplete 'Kawasaki Disease as it can be fatal. On the contrary, these children often have devastating coronary sequelae as the diagnosis and treatment often gets delayed. Kawasaki Disease has a much higher mortality in India than in developed countries. In the Chandigarh cohort, the mortality rate over the last 20 years is 0.8%, as compared to 0.01-0.08% in children in developed countries[1].'Incomplete' and 'atypical' forms of Kawasaki Disease are now being increasingly diagnosed and reported[2,3]. The acute phase of illness as a result of inflammation and scarring of the heart and blood vessels. This scarring takes years, sometimes decades, to develop and is often clinically silent. Early intervention with IVIG can benefit in terms of prognosis.

Case

3 years old Rivaan, 2nd issue of a non-consanguinous marriage was admittedt with complaintsof mild to moderate grade, intermittent, fever since 11 days with no afebrile interval and reddish rash that appeared all over the body since 6 days associated with swelling of lips and both sides of the neck 4 days back. He was admitted in an outside hospital where he received symptomatic treatment and Inj Ceftriaxone for four days where he was diagnosed of having dengue fever. Child was continued with IV Ceftriaxone and IV fluids with other symptomatic treatment. With clinical suspicion of rickettsial fever, weilfelix test was sent and started on Doxycycline, which came to be negative. Dengue IgM ELISA was sent, which was positive. Since high grade fever spikes persisted even after 12 days of illness, along with lymphadenopathy, erythema and cracking of lips, erythema and swelling of hands and feet, clinical diagnosis of Kawasaki Disease was made. ESR and CRP were raised (38 and 121 respectively with Haemogram showing raising trend of platelets).IV Ceftriaxone and Doxycycline were omitted. Serum Ferritin and LDH were sent, which were 151 and 988 respectively with normal LFT ruling out HLH secondary to dengue. 2D Echo done was suggestive of mild pericardial effusion with Z score < 2 of coronary arteries. Dr Vijay Vishwanathan's (Pediatric Rheumatologist) opinion was taken who suggested sending NT Pro BNP (sensitive marker for Kawasaki disease) and consider starting IVIG, which was raised (1662 pg/ml). Child was given IVIG @2g/kg over 3 days and low dose Aspirin (3mg/kg) was started. Gradually fever spikes and oedema reduced and oral intake improved. Presently, as child is afebrile, orally accepting well, hemodynamically stable he is being discharged with plan to do 2D ECHO after 4 weeks on OPD basis.

Conclusion

Development of coronary arteryabnormalities (CAA) is the hall mark of Kawasaki Disease and accounts for most of the morbidity and mortality as sociated with the disease. Prompt recognition of the disease and early initiation of treatment with intravenous immunoglobulin (IVIG) results in significant reduction in the occurrence of CAA. It is, therefore, imperative for the pediatrician to diagnose and treat Kawasaki Disease expeditiously. Alotmore needs to be done as far as increasing awareness about this condition is concerned by means of .Information and education campaigns. Kawasaki Disease remains a challenge for all pediatricians in the country.

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A COMPARATIVE STUDY OF NT-proBNP LEVELS IN PATIENTS OF ACUTE PHASE OF KAWASAKI DISEASE, SEPSIS AND OTHER FEBRILE ILLNESSES

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Introduction

The diagnosis of Kawasaki disease (KD) is based on clinical signs and symptoms. There is no definite bio-marker to differentiate Kawasaki disease from sepsis and other acute febrile illnesses. The purpose of our study was to assess the NT-proBNP value in acute phase of Kawasaki disease, sepsis and other acute febrile illnesses and to compare the respective values and see if it specifically increases in acute phase of Kawasaki disease.

Methodology

Cross-sectional study of serum NT-proBNP levels in 40 Kawasaki disease patients and 40 age and sex matched sepsis and other febrile illness patients admitted in a tertiary hospital from January 2019 to April 2020 and comparatively studied the levels in both the groups.

Results

The NT-proBNP level was significantly higher in acute phase of Kawasaki disease than sepsis and other febrile illnesses. The value of NT-proBNP level in acute phase of KD and sepsis and other febrile illnesses were 914.9168n ± 1269.8089 (mean ± SD) and 219.0390 ± 306.3808 (mean ± SD) respectively. The p-value was 0.0012. Considering 225 pg/ml as cut-off value, there were significantly higher number of KD patients (p = 0.0007) whose serum NT-proBNP levels were above 225 pg/ml when compared to sepsis and other febrile illnesses.

Conclusion

The serum NT-proBNP level may be regarded as an important bio-marker in the diagnosis of Kawasaki disease in acute phase.

LYMPHOCYTE ACTIVATION MARKERS IN CHILDREN WITH KAWASAKI DISEASE- A PRELIMINARY STUDY FROM NORTH-WEST INDIA

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Introduction

Kawasaki disease (KD) is one of the commonest vasculitis of childhood and a common cause of acquired heart disease in children in developed countries. Lymphocytes plays a major role in the pathogenesis of KD as evidenced by coronary artery autopsy studies. Previous studies in T cell activation in KD have shown conflicting results.

Objective: To assess flow-cytometry based T-cell activation markers and serum soluble CD25 levels in children with KD

Methods

Study was conducted in Allergy Immunology Unit and Paediatric Rheumatology Clinic of the Advanced Paediatrics Centre, PGIMER Chandigarh during the period of January 2019 to July 2020. We enrolled 10 childrenwith KD, 10 age and sex-matched healthy controls, and 9 febrile controls in our study after obtaining written informed consent from parents. Diagnosis of KD was made according to the AHA criteria for diagnosis of KD.Febrile and healthy controls were enrolled from in-patient ward and out-patient services, respectively.We studied both early and late activation markers of T cell activation by flow cytometry (CD69 and HLA-DR, respectively) in children with KD in acute phase and compared them with healthy and febrile controls. Longitudinal follow-up of KD patients was done and T cell activation profiles were assayed after 24 hrs and 3 months of intravenous immunoglobulin (IVIg) administration. Sandwich ELISA was used for soluble CD25 estimation in serum. The study was approved by the Institute Thesis Committee and the Institute Ethics Committee.

Results

Median age group (inter-quartile range (IQR)) in cases, febrile and healthy controls was 3.5 years (2.12, 4.75), 3 years (2, 5), and 3.5 years (3, 5), respectively. Among patients with KD, 4 had complete KD, 6 had incomplete presentation, and none had coronary artery aneurysms or IVIg resistant forms of KD. Median duration (IQR) of fever in cases and febrile controls was 12 days (9.25, 15) and 6 days (3, 7), respectively.Compared to healthy controls [median(IQR):1.74% (0.78-2.29)], children with KD [median (IQR): 2.65% (1.81-4.74)] and febrile controls[median(IQR): 3.57% (2.68-4.48)] showed increased expression of CD69 on CD3+CD4+ T cells (p-0.04). CD69 expression on CD3+CD8+ T cells was also increased in children with KD [median (IQR):4.84% (3.07-6.7)] and febrile controls [median(IQR): 6.33% (3.32-7.93)] as compared to healthy controls [median(IQR): 2.7% (1.86-3.27)], however, this difference was not statistically significant(p-0.056). No difference was found in HLA-DR expression between KD, febrile, and healthy controls.On longitudinal follow-up of cases, no change in HLA-DR expression was noted. Soluble CD25 was significantly elevated in KD [median (IQR):3681.5pg/ml (2026.75-4542)] as compared to febrile [median (IQR) 2755 pg/ml (2026-4098)] and healthy controls[median(IQR): 514 pg/ml (503.75-534)]. Levels of soluble CD25 in children with KD decreased on follow up after IVIg administration, however, levels were higher even 3 months after diagnosis when compared to healthy controls.

Conclusions

To conclude, we document early but not late activation of T lymphocytes in milder forms of KD.Markers of lymphocyte activation do decrease with subsidence of systemic inflammationfollowing IVIg therapy in KD. Future studies in a larger cohort are needed to establish the role of markers of lymphocyte activation as diagnostic or prognostic biomarkers in KD.

CD40 GENE POLYMORPHISM AND D40 EXPRESSION IN CHILDREN WITH KAWASAKI DISEASE

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Introduction

Kawasaki disease (KD)is a medium vessel vasculitis of unknown etiology.High prevalence of KD in the North-East Asian nations led to a speculation that KD hasgenetic predisposition.Polymorphism of CD40 gene has been documented for susceptibility and development of CAAs in KD in Japanese, Chinese and Taiwanese population. There ispaucity of literature pertaining to CD40 gene polymorphism in KD from the Indian subcontinent. We planned to ascertain the role of CD40 gene polymorphism and CD40 expression in children with KD from North India.

Methods

This hospital-based case-controlled study including 37 KD during acute phase and 37 febrile control was carried out during the year 2019-20. CD40 expression on B cell was assessed in both groups.Toidentify single nucleotide polymorphism (rs4810485, rs1535045) of CD40 gene,Sanger sequencing was carried out in KD and controls.

Results

TheCD40expression(stimulation index (SI), Δ mean fluorescence and CD40 percentage expression) between two groups were not statistically significant (p=0.46; p=0.44; p =0.65) respectively. There was no statistically significant difference in genotypes and allele frequencies of rs153045, rs4810485 and between KD and controls. A metanalysis including our results and previous studies also failed to reveal any association of thesepolymorphisms with KD.

Conclusion

The CD40 expression between KD and controls was not statistically different. There was no association of CD40 polymorphism SNP(rs153045,rs4810485) with susceptibility to KD or KD with coronary abnormalities in our cohort. Further study with a large sample size required to reach a definite conclusion.

A PRELIMINARY STUDY OF CLINICOPATHOLOGICAL CHARACTERISTICS OF KAWASAKI LIKE PAEDIATRIC INFLAMMATORY MULTISYSTEM SYNDROME (PIMS) DURING COVID ERA

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Introduction:

With the ongoing COVID pandemic, children presenting with PIMS are being reported increasingly. We hereby present an observational study of the clinicopathological characteristics and therapeutic responses of 20 such patients from 3 tertiary centres of Eastern India.

Method

Data of patients presenting with KD like manifestations in the last 3 months, associated with the detection of evidence of SARS-COV2 infection were compiled from 3 tertiary hospitals of Kolkata.

Results

20 patients were included (Median age 7years, range 1 - 15 years).65% had history of COVID exposure,and presented with fever (102-103F) and irritability/drowsiness within 4-6 weeks of exposure,associated with an erythematous rash,red lips and bilateral nonpurulent conjunctivitis in 80% patients.50% of the patients had disproportionate tachycardia,hypotension with low ejection fraction(40-45%) on echocardiography. 40% had gastro-intestinal complaints like pain abdomen,loose stools and vomiting.Neutrophilic leucocytosis with lymphopenia, normal platelet count,hyperferritinemia(mean 762.7 ng/ml) and very high CRP (mean 192.7 mg/L), LDH (mean 348.6 U/L) were predominant in all ; associated with raised D-dimer (mean 2797 ng/ml) and high NT-PRO BNP (mean 9449 pg/ml) in 70% and high IL6 (56.5 pg/ml) in 60%.All patients received IVIG @2gm/kg and antiplatelet dose of aspirin.Fever, elevated CRP (with ongoing myocarditis) persisted in 25%, and they were treated with intravenous methyl prednisolone 2mg/kg/day for 3 to 5 days . 70% needed PICU admission, 10% having cardiogenic shock needed inotropes and 1 patient with refractory hypotension required mechanical ventilation. All patients with myocarditis had rapid improvement of ejection fraction after 2 to 3 days of therapy and 3 had transient episodes of bradyarrhythmia that subsided spontaneously. There was no mortality and majority could be discharged after 7-8 days of hospital stay on low dose aspirin and advised follow up echocardiography after 2 and 6 weeks.

Conclusion

Majority of the patients had atypical KD like or toxic shock like presentation. 70% were positive for COVID IgG antibody,and presented after a 4-6 weeks lag. Unlike KD,PIMS involves an older age group, have normal platelet counts and a very high NT-PRO BNP(sometimes>10000), CRP, D-dimer and IL-6 are characteristic. There was a higher incidence of myocarditis and a lower coronary artery dilatation as compared to KD. Timely diagnosis and immediate institution of therapy rapidly reverts the hyperinflammation without significant residual sequelae.

KAWASAKI DISEASE INCIDENCE AT CHANDIGARH, NORTH INDIA: 2015 – 2019

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Introduction

Kawasaki disease (KD) is a common childhood vasculitis. Epidemiological studies have shown an increasing incidence of KD in Japan, Korea and Taiwan. However, there is paucity of nationwide data on the epidemiology of KD from India.

Patients and Methods:

This study was carried out to calculate annual incidence of KD at Chandigarh, India. The population of Chandigarh was taken from the National Census data, 2011. Population estimation for remaining intervening years was obtained from decadal growth rate (17.19%). Children <5 were calculated by subtracting from under 6 group on pro-rata basis.

Results and Discussion:

Eighty three patients (66 boys, 17 girls) were diagnosed with KD who resided in Chandigarh. Annual incidence was 5.64, 9.25, 9.11, 9.87 and 10.6 per 100,000 children <5 (Mean: 8.89/100,000), while for children <15 it was 2.65, 4.44, 3.86, 5.32 and 4.99 per 100,000 (Mean: 4.25/100,000) in 2015, 2016, 2017, 2018 and 2019 respectively. Male: female ratio was 3.9:1. Median age for boys and girls was 48 and 54 months respectively. Peak incidence was in the fourth year of life. Peak incidence was noted in the months of April and September; while a nadir was seen in February. Coronary artery abnormalities were seen in 14 patients. Cardiac complications were reported in 17.7% patients.

Conclusion

We demonstrate a 53% increase in annual incidence of KD in children <5 and a 53.7% increase in children <15 in last 5 years as compared to 2009-2014. This may reflect a true increase in the incidence of KD or may be due to increased ascertainment of disease as a result of increased awareness.

MACROPHAGE ACTIVATION SYNDROME IN KAWASAKI DISEASE: OUR EXPERIENCE AT CHANDIGARH

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Objectives

To carry out a review of clinical characteristics, laboratory profile, management and outcomes of patients with KD and macrophage activation syndrome (MAS).

Methods

Medical records of patients treated for KD and MAS between January 1994 - December 2019 were reviewed. Patient demographics, clinical signs, laboratory values, coronary artery abnormalities (CAAs), treatment and outcome of the patients having KD and MAS were noted. We have also performed review of literature of published studies on the subject.

Results

Of the 950 cases of KD, 12 (1.3%; 10 boys; 2 girls) were diagnosed to have MAS. Median age at diagnosis was 4 years (range:9 months - 7.5 years). Median interval between onset of fever and diagnosis of KD was 11 days (range:6-30 days). Median duration of fever was 7 days (range:5 - 35 days). Thrombocytopenia was seen in 11 patients. Median proBNP value was 2101 pg/ml (range:164-75911 pg/ml). CAAswere seen in 5(41.7%) patients– 2 had dilatation and ectasia of LMCA; 1 had ectasia of RCA; 1 had dilatation of LMCA and RCA both, and 1 had bright coronaries. All patients received intravenous immunoglobulin (IVIg) – 2g/Kg as first line therapy. All patients required additional therapy (methylprednisolone - 11; second dose IVIg - 1; intravenous infliximab – 4; oral cyclosporine - 1).

Conclusion

MAS is an unusual and under-recognised complication of KD. In our cohort of 950 patients of KD, 1.3% had developed MAS. KD with MAS is associated with significantly high levels of proBNP and increased incidence of CAAs.

DENSE CALCIFICATIONS OF A GIANT CORONARY ANEURYSM IN AN 11-YEAR-OLD GIRL WITH KAWASAKI DISEASE

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Introduction

Patients with Kawasaki disease (KD) with residual coronary abnormalities show evidence of coronary artery calcifications. Detection of coronary artery calcifications may be useful for risk stratification in the long-term management of patients with KD.

Case details

3-year-old girl, was diagnosed to have KD on Day 15 of fever. 2D-transthoracic echocardiography showed diffuse right coronary artery (RCA) dilatation (6mm in diameter), fusiform aneurysms in left main coronary artery (LCA) (3.7mm in diameter) and left anterior descending (LAD) artery (proximal LAD 5.2mm; mid LAD diameter 8.6mm), pericardial effusion and mild mitral regurgitation. She received intravenous immunoglobulin (2 g/kg) and intravenous infliximab (5 mg/kg). In view of giant aneurysm in LAD, she was initiated on aspirin and warfarin. Follow-up echocardiography showed persistence of coronary artery abnormalities.

At 11 years Computed Tomography Coronary Angiography was performed which revealed mural calcifications in distal LCA. LAD in its proximal segment (thick arrow in a and c) shows densely calcified giant fusiform aneurysm (8.4mm in diameter and 20mm in length) and a densely calcified fusiform aneurysm measuring (17.5mm length, 5.5mm diameter) in RCA in proximal segment (arrow in b and c) with diffusely dilated mid and distal RCA (4.2mm diameter) with lack of normal distal tapering. A focus of mural calcification was noted in dilated mid RCA (arrowhead in b). Left circumflex was dilated in its proximal and mid segment (4mm in diameter).

Conclusion: Dense calcifications (Agaston score-2522) of this severity are extremely unusual in young children and occur due to dystrophic calcification in coronary artery abnormality of KD.

Acknowledgement: None

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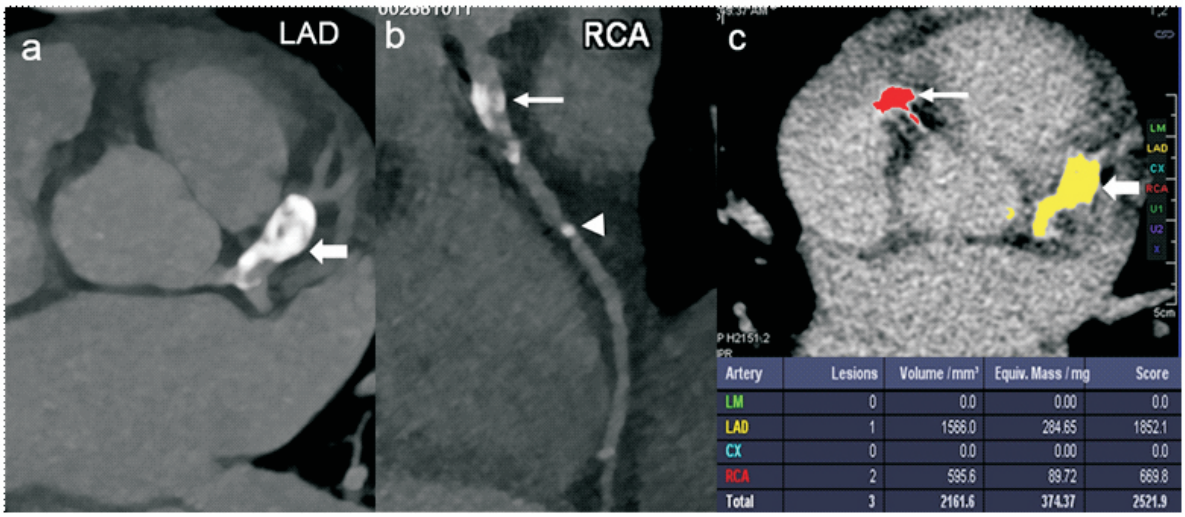

Artery	Lesions	Volume /mm³	Equiv. Mass / mg	Score
LM	0	0.0	0.00	0.0
LAD	1	1566.0	284.65	1852.1
CX	0	0.0	0.00	0.0
RCA	2	595.6	89.72	669.8
Total	3	2161.6	374.37	2521.9

CASE SERIES OF KAWASAKI MIMICS- A DIAGNOSTIC CHALLENGE

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Introduction

Kawasaki disease(KD) is diagnosed clinically when 4 out of 5 criteria are present. In incomplete KD,the diagnosis can be made with two criteria. Infections, rheumatological disorders may mimic KD and need to be ruled out before the diagnosis. We present 2 cases which closely mimicked KD and posed a diagnostic challenge.

Case 1

3yr/male presented to Pediatric institute with two weeks fever, unilateral cervical nodes, erythematous tongue. He wasinactiveand confined to bed, attributed to prolonged fever. On workup, he had anemia, leucocytosis with mature cells, thrombocytosis, elevated ESR, CRP and normal liver, renal function.USG abdomen showed mild hepatomegaly and minimal right pleural effusion. Workup for common infections including tuberculosis, blood andurine cultureswere negative. Echo done as a part of completion showed coronary hyperechogenicity with normal z scores(fig1).His ferritin was 1300ng/mL and repeat blood counts, ESR were decreasing to low normal levels. He was suspected as incomplete KDand referred for rheumatology opinion. On examination he had tenderness,synovial thickening in bilateral knee and elbow. Hehad increased triglycerides and normal fibrinogen. We advised bone marrow and USG of above joints. USG showed synovitis in bilateral knee, ankle and bone marrow revealed hemophagocytosis. He was diagnosed as systemic arthritis with macrophage activation syndrome and managed with pulse steroids, toclizumab, cyclosporine with remarkable improvement.

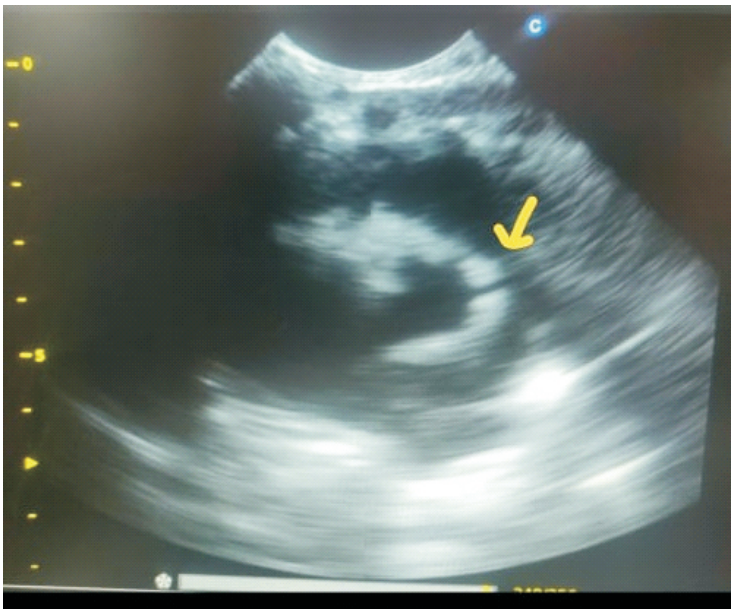

Fig. 1 Echo showing coronary hyperechogenicity(arrow) in short axis view

Case 2

5yr/malepresented with three weeks fever, bilateral cervical nodes, hepatomegaly and desquamation of skin of extremities.CBC showed leucocytosis with mature cells, thromboctyosis, elevated ESR, normal renal, liver function, triglycerides. His workup for common infections, cultures were negative. Echo showed coronary wallhyperecho-genicity(fig 2). Child had history of similiar episodes, twice in past 18 months. He was diagnosed as incomplete KD and treated with IVIG for the past, current episodes. As child had persistent fever after IVIG, he wasreferred for rheumatology opinion regarding second course of IVIG. On examination patient had tenderness, synovial thickening,mild FFD in bilateral elbow. USG elbow was suggestive of synovitis. He was treated as systemic arthritis with steroids, methotrexate with resolution of symptoms.

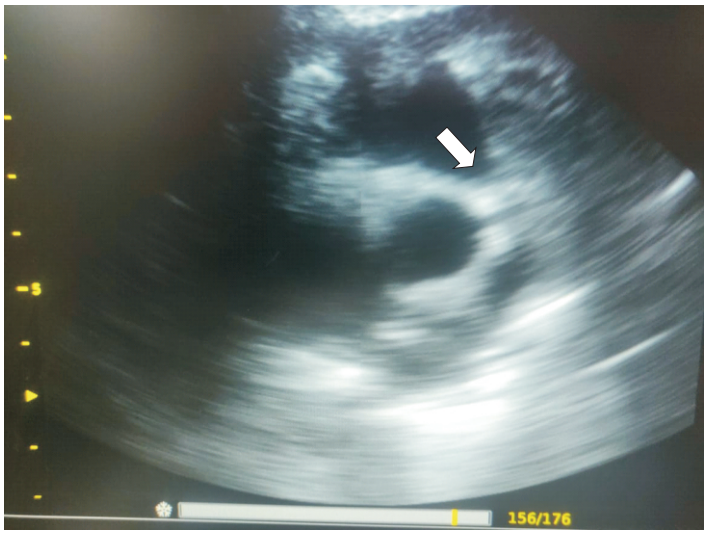

Fig. 2 Echo showing right coronary wall hyperechoes(arrow) in short axis view.

Conclusion

Coronary hyperechogenicity and dilatation has been reported in systemic arthritis[1-3].The diagnosis of incomplete KD has to be done after ruling out all the illness mimicking KD. Vigilant examination for synovitis may be fruitful.

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COMPLETE VERSUS INCOMPLETE KAWASAKI DISEASE IN PAEDIATRIC PATIENTS : AN EMPIRICAL ANALYSIS

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Introduction:

Kawasaki disease, an acute self-limiting systemic vasculitis involving medium and small-sized arteries, has been a challenge for paediatricians for several decades. Approximately 15-20% of patients with Kawasaki disease (KD) is known to have incomplete presentation, but the incidence over the years is gradually rising.

Objectives

The study aimedto determine the differences in clinical and laboratory features and coronary artery outcomesbetween complete and incomplete Kawasaki disease.

Methodology

This hospital-based observational study was conducted over a 15-month period(1st June, 2019 to 31st August, 2020) among children between 1 month to 12 years of age in Paediatric ward of Burdwan Medical College, West Bengal.Clinical and laboratory findings and echocardiographic features of all the patients who were admitted during that time period with the diagnosis of complete or incomplete KD were noted in the study.

Results

:We identified 41 patients with amean age of 30 months (±24). 21 patients had incomplete and 20 patients had complete KD. Among the incomplete KD group 11 patients fulfilled 2 principal criteria and 10 fulfilled 3 principal criteria. Median age at presentation in Incomplete and complete KD group was 20.5 and 26 months respectively. The group with incomplete KD had significantly lower incidence of changes in extremities, conjunctival injection, exanthem and a higher incidence of Coronary artery abnormalities (42.8% vs 30% in the complete KD group). No significant differences were found regarding sex, blood cell counts and laboratory markers for inflammation. One patient with incomplete KD had BCG inoculation site inflammation one had anterior uveitis.

Conclusion

The proportion of incomplete KD in our study is higher compared to other recent studies. There was higher incidence of coronary artery abnormalities in the incomplete KD group in our study, which may be due to delay in diagnosis. These make us question whether we should wait for a suspected KD case to fulfill all the clinical and laboratory criteria or should we start treatment immediately in those patients to reduce complications, and also whether it is time to add some additional criteria or redefine Kawasaki disease.

INCOMPLETE KAWASAKI DISEASE

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Introduction

It is important to recognize that 'incomplete' Kawasaki Disease as it can be fatal. On the contrary, these children often have devastating coronary sequelae as the diagnosis and treatment often gets delayed. Kawasaki Disease has a much higher mortality in India than in developed countries[1]. In the Chandigarh cohort, the mortality rate over the last 20 years is 0.8%, as compared to 0.01-0.08% in children in developed countries. 'Incomplete' and 'atypical' forms of Kawasaki Disease are now being increasingly diagnosed and reported[2,3],. The acute phase of illness as a result of inflammation and scarring of the heart and blood vessels. This scarring takes years, sometimes decades, to develop and is often clinically silent. Early intervention with IVIG can benefit in terms of prognosis.

Case

1 ½ year male child was admitted with fever since 5 days after which he started developing maculopapular rash all over body followed by cracking of the lips and tongue. Laboratory investigations revealed normal CBC with elevated ESR and CRP while the urine routine showed sterile pyuria. With clinical suspicion of incomplete Kawasaki disease, 2D ECHO showed impaired systolic function EF- 24% with left coronary artery dilatation (Z score >4). Child was started on IVIG @2g/kg and on Aspirin (@75 mg/kg/day). Gradually his fever and rash subsided and follow up 2D Echo was normal with ejection fraction 65-70%. Currently he is on antiplatelet dose of Aspirin of 5mg/kg and was advised to repeat 2D ECHO after 3 months.

Conclusion:

Development of coronary artery abnormalities (CAA) is the hallmark of Kawasaki Disease and accounts for most of the morbidity and mortality associated with the disease. Prompt recognition of the disease and early initiation of treatment with intravenous immunoglobulin (IVIG) results in significant reduction in the occurrence of CAA. It is, therefore, imperative for the pediatrician to diagnose and treat Kawasaki Disease expeditiously. A lot more needs to be done as far as increasing awareness about this condition is concerned by means of information and educational campaigns. Incomplete Kawasaki Disease still remains a challenge for all pediatricians in the country.

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MIDTERM OUTCOMES OF KAWASAKI DISEASE WITH GIANT CORONARY ANEURYSMS IN THE INDIAN SETTING

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Introduction

The cohort of children with Kawasaki Disease (KD) and giant coronary aneurysms (GCA) are at an increased risk of major adverse cardiovascular events (MACE). However very little data is available regarding GCA in the Indian population.

Methodology

We conducted a retrospective study of children with Kawasaki Disease referred to our department between January 2015 and December 2019.

Results

During the study period, 31 children were identified presenting at a median age of 27 months (2-110 months), of which 21 (67.7%) were male. 6 patients were diagnosed as incomplete Kawasaki disease. 16 children (51.6%) were diagnosed after 10 days of fever. There was no cardiac involvement on echocardiogram in 7(22.6%) while 24 (77.4%) had evidence of coronary artery abnormalities (CAA) including 11 (35.5%) with GCA. Intravenous immunoglobulin(IVIG) therapy was administered in 30 children. IVIG resistance was noted in 16.6% (n=5) including 2 patients who were diagnosed late.

Children with GCA were more likely to be diagnosed after 10 days of fever (p=0.01). All children with GCA were treated with oral anticoagulants in addition to Aspirin. On a median follow up of 35 months, 2 of these children had MACE. One child developed an aneurysm of the LV apex secondary to infarction involving the LAD while another child presented with recurrent myocardial infarction 30 days after discharge from the hospital and underwent coronary artery bypass grafting. Both the children remained clinically well at the last follow up.

Conclusions

Children with GCA in KD had a low incidence of MACE on medium term follow up.

A PRELIMINARY OBSERVATIONAL STUDY ON PREDICTING TREATMENT INTENSIFICATION IN PEDIATRIC MULTISYSTEM INFLAMMATORY SYNDROME (PIMS)BASED ON CRP, NT-PROBNP AND INITIAL ECHOCARDIOGRAPHY

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With the emergence of COVID 19 pandemic, there has been a rise in an elusive entity namedas Pediatric Multisystem Inflammatory Syndrometemporally associated with COVID 19.

Being quite in the nascent stage, a consensus guideline is yet to be achieved on the treatment protocol. Presently we are using intravenous immunoglobulin2g/kg as initial therapy, methyl prednisoloneis being added in patients with inadequate clinical/biochemical response.

CRP and N-terminal brain natriuretic peptides(NT-proBNP) are 2 commonly used markers in KD as well as PIMS.Wehave attempted to observe the predictive values of these two molecules along with the initial ECHO report in determining the need for treatment intensification in these patients.

Methods

Retrospective analysis of data of PIMS patients admitted from July 2020 till mid September 2020.

Results

The study had 19 patients of PIMS with data available on their CRP, NTproBNP and initial echocardiography report. 9 of them responded to IVIG alone, while the other 10 required IVIG and Methyl Prednisolone. A logistic regression model was implemented to find out the dependence of Methyl Prednisolone on CRP, NTproBNP levels and Echocardiography findings (EF levels), and essentially attempt to predict Steroid requirements in patients from the 3 factors. The computation was done using the Statsmodels library in Python. The coefficients for CRP, NTproBNP and EF levels did not show any significant difference from 0 (p value>0.05), meaning that they have no significant dependence on Steroid requirement criteria, or the log odds (probability) of a Methyl Prednisolone requirement does not change significantly on changes in any of those 3 variables.

A separate T-test was run to compare mean values of CRP levels in patients requiring only IVIG and patients requiring both IVIG and Methyl Prednisolone. There was no significant difference in the mean levels of CRP in both types of patients. The same test was run for NTproBNP levels in both groups of patients, and there was no significant difference in the levels here either.

Conclusion

With the present data, it was not possible to statistically predict the requirement of steroids alongwith the use of IVIg based on initial CRP, NT-pro BNP and initial echocardiographic values. However, drawback of the present study is the inadequate patient number.

NEONATAL MIS-C – HARNESSING THE CYTOKINE STORM

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A term baby girl with normal ante-natal history started having fever from day 8with an erythematous generalised rash. She was admitted on D10 at a local hospital and treated as late onset sepsis and given broad spectrum antibiotics along with 1 gm/kg of IVIG for thrombocytopenia. Referred to our hospital on D24 due to recurrence of symptom.

She had tachycardia, mild respiratory distress, severe anaemia, thrombocytopaenia, oedema, hypoalbumanaemia and macular rash with central necrosis in some lesions.

GeneXpert for COVID-19 was positive. Within twelve hours, baby developed fulminant cardiogenic shock with significant pulmonary oedema and got ventilated with 100% Oxygen and high pressures. Echocardiography revealed ejection fraction of 40% and mild pericardial effusion. NT-Pro BNP was 33000gm/L,D-dimer 16500 µg/L, Ferritin 16000µg/L.Dexamethasone, IVIG and Enoxaparin was started keeping COVID myocarditis with cytokine storm as diagnosis.

Next day, she developed pulmonary haemorrhage, a seizure and cardiac arrest needing CPR. She subsequently developed acute kidney injury. She was extubated to HFNC after 5 days. Steroid was stopped after 5 days as she developed hypertension and Haematuria.

However, she developed respiratory distress again 4 days after extubation and was ventilated again the following day. Echo showed moderate LV dysfunction. NT-proBNP and Ferritin, which initially decreased showed a secondary rise. Diuretics was increased and Milrinone infusion given for 72 hours. Methyl-prednisolone was restarted and given for 5 days followed by tapering dose of oral Prednisolone. All inflammatory markers improved. Baby was discharged at the age 50 days.

TRANSFUSION RELATED ACUTE LUNG INJURY (TRALI) POST IVIG THERAPY IN KAWASAKI DISEASE WITH PULMONARY INVOLVEMENT:

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Background

Kawasaki disease (KD) presenting with isolated lung involvement in its early stages is uncommon.1 Transfusion related acute lung injury (TRALI) is a rare complication following IVIG therapy.

Case characteristics

11 months old male infant presented with fever, respiratory distress (RD) and irritability. Chest X-ray (CXR) showed right lower zone reticulonodular infiltrates (RNI). For a provisional diagnosis of severe pneumonia, oxygen and antibiotics were started. On day-4 of hospital stay, child had high grade fever, oral mucosal redness and perianal desquamation with persisting RD. Erythrocyte sedimentation rate was 60 mm/hr. Echocardiogram showed aneurysm of left main coronary. The diagnosis of incomplete KD was confirmed and IVIG (2 g/kg) given. Twelve hours post IVIG infusion, the child developed worsening of RD with Spo2 <90%, without hepatomegaly or cardiac gallop.

Observation

Repeat CXR which showed diffuse RNI. PaO2/FiO2 ratio was 226. The child was initiated on Non-invasive ventilation. Over the next 36 hours, weaned to oxygen prongs. Based on the clinical and radiological features, it was understood that respiratory distress was due to IVIG induced delayed TRALI (dTRALI).2 Child continued to have minimal RD requiring oxygen, suggestive of resistant KD. Hence, child was started on oral prednisolone.3 Oxygen was weaned and stopped in the next three days.

Message

Incomplete KD should be considered in a case of pneumonia not responding to appropriate management. Since KD is associated with significant inflammation it can predispose the child to develop TRALI following IVIG. Steroids are indicated for IVIG refractory KD.

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PAEDIATRIC INFLAMMATORY MULTISYSTEM SYNDROME TEMPORALLY ASSOCIATED WITH SARS-COV-2 IN A CHILD WHO HAD KAWASAKI DISEASE FIVE YEARS BACK

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Introduction

Kawasaki disease (KD) like illness in context of SARS-CoV-2 was first described from Bergamo province, an Italian epicenter for SARS-CoV-2 epidemic (Verdoni et al.). This syndrome referred as Paediatric inflammatory multisystem syndrome temporally associated with SARS-CoV-2 (PMIS-C) has now been described world over. It is characterized by high grade fever, mucocutaneous involvement, and multisystemic involvement along with marked elevation in inflammatory markers. It usually follows 2-6 weeks after SARS-CoV-2 infection and RT-PCR for SARS-CoV-2 may be negative, but antibodies can be identified. We here in report a child who KD 5 years back but developed PIMS 4 weeks after SARS-CoV-2 infection.

Case

K, 8 years old female child, second born to a non consanguineously married couple presented with continuous high-gradefever (103-degree F) for 5 days. She received antimicrobials at a local hospital without any response.On day4 of illness fever, she developed reddish discoloration of tongue and conjunctival injection. Due to persistent pyrexia, mucocutaneous involvement parents suspected KD and reached our centre. She was diagnosed and treated for classical KD 5 years back with intravenousimmunoglobulin (IVIg) and aspirin. No coronary involvement was noted then. There was recent history of exposure to SARS-CoV-2 virus. Grandfather and mother had developed COVID-19 prior to onset of symptoms. The index patient had remained asymptomatic. On examination she was febrile, had strawberry tongue and mild conjunctival injection. Systemic examination was unremarkable. Laboratory investigations revealed: Haemoglobin (Hb): 12.8 g/dl, Total Leukocyte count (TLC): $7.9*10^{9}$/L, Differential Leukocyte count (DLC): $N_{65}L_{29}M_{2.3}E_{2.1}$, Platelets: $320*10^{3}$/L. Inflammatory parameters were markedly elevated Erythrocyte Sedimentation Rate (ESR): 53 mm in first hour, C - reactive protein: 80 mg/dl, Pro–B-Type NatriureticPeptide (Pro BNP): 840pg/ml and serum ferritin 550 ng/mL.

RT-PCR from nasopharyngeal swab for SARS-CoV-2 was negative but IgG antibody titres were elevated (115 units; Normal < 1.0 AU/mL). Echocardiography was normal. A possibility of PIMS-C was considered and she was treated with IVIg 2 gram/Kg. Fever responded dramatically and the laboratory parameters normalized.

Discussion and conclusion: Etiopathogenesis and risk factors for development of PIMS-C are not well understood. It is unclear if children with prior KD are at higher risk for SARS-Co-V-2 triggered inflammatory syndrome. Reoccurrence of KD like illness temporally following SARS-CoV-2 infection in the index child suggests a genetic predilection for inflammatory syndrome following viral triggers.

There is a need for more information about similar patients and their follow up as it may provide valuable insights to recurrent KD.

INCOMPLETE KAWASAKI DISEASE

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Introduction

Kawasaki disease (KD) is a medium vessel vasculitis of young children with a special predilection for the coronary arteries[1]. In Japan, J Fukushige and his team have found that almost 10% of all the cases admitted as classic Kawasaki disease were finally diagnosed as incomplete Kawasaki[2]. These children go unnoticed and further develop coronary artery involvement which can be fatal. In India alone as per a cohort study in Chandigarh, the mortality rate over the last 20 years is 0.8%, as compared to 0.01-0.08% in children in developed countries[3]. 'Incomplete' and 'atypical' forms of Kawasaki Disease are now being increasingly diagnosed and reported[4.5]. Early recognition with high suspicion of index and prompt treatment is the key to avoid such fatalities,

Case

3 year male child was admitted with fever since 11 days. On day 6, he started developing an erythematous maculopapular rash all over body followed by swelling and cracking of lips with palpable masses over both sides of his neck and swelling of hands and feet. Laboratory investigations revealed normal CBC with elevated ESR and CRP. Hence suspecting Kawasaki disease, 2D ECHO was done showed mild pericardial effusion with z score of <2. NT Pro BNP raised (1662 pg/ml). With a working diagnosis of incomplete Kawasaki's disease he was started on IVIG @2gm/kg and on oral low dose Aspirin (@3 mg/kg/day). Gradually his fever spikes, edema and rash subsided and follow up ESR and CRP showed a falling trend.

Conclusion:

Incomplete Kawasaki disease is an underdiagnosed critical illness which can cause fatal coronary outcomes. Prompt recognition and early treatment with intravenous immunoglobulin (IVIG) helps in reducing complications. A low threshold of suspicion is mandatory and the need for widespread awareness is a must to overcome the disease burden.

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