Case Report

Life Threatening Vasculitis in a Child- Kawasaki Disease – Case Report & Literature Review

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Abstract

Kawasaki disease(KD) though not very common , is a clinical condition which needs a high degree of suspicion for diagnosis. The child presented to our hospital with history of fever of 12days duration and a laboratory evidence of thrombocytosis. Child was afebrile with minimal peeling of skin of both soles. Based on the retrospective history and clinical presence of unilateral cervical lymphadenopathy with a lab evidence of raised ESR , platelet count and CRP child was diagnosed to have KD and started on immunoglobulin. Echo revealed giant coronary aneurysms . Child is on follow up with aspirin, clopidogrel and warfarin.

Key Words: Kawasaki disease, coronary aneurysm, vasculitis.

Introduction

Kawasaki disease is a mucocutaneous lymph node syndrome that affects infants and children. High index of suspicion supported by laboratory investigations, leads to early diagnosis and treatment that can prevent coronary aneurysms and long term morbidity and mortality.

Case report

This is the case report of a 5 year old male child who was referred to our institute with 12 days fever and thrombocytosis. There was no other complaint . On examination the child was conscious, oriented, febrile and hemodynamically stable. Child had right cervical lymphadenopathy (2cm). systemic examination was not contributory. Examination of the extremities revealed peeling of the skin over the soles (Fg-1). Retrospective history revealed that the child had fever with erythematous rashes and a red tongue during the first week of the illness. A clinical diagnosis of kawasaki disease was suspected. Laboratory evaluation revealed total count of 6000 cells/cumm, differential count of P64 L33 E3, Platelet count: was 8 lakhs/cu.mm. CRP was 24 mg/dl, ESR was 40mm in 30 minutes and 70 mm at 1 hour. Repeat platelet counts were 10 lakhs and 12 lakhs over the next 3-4 days. Blood widal test, non enteric culture, urine routine, urine culture, serology for scrub typhus and MSAT were negative. Echo revealed giant aneurysm of Right Coronary artery, Left main coronary and Left anterior descending artery. (Fig 2) Right coronary artery was dilated with a measurement of 5.8mm. Left Main Coronary Artery measurements at the origin and at the distal ends were 3.8 mm and 11*16 mm respectively - aneurysm. Left Anterior Descending artery was hugely dilated with a measurement of 10 mm. X-ray chest and ECG were normal. Child received 2g /kg IVIG as an infusion over 12 hours. Child was also treated with T. Aspirin 100 mg/kg/day until ESR was normal, T. Clopidogrel and T. warfarin . Repeat ECHO after 8 weeks revealed no regression. Child is now on follow up with oral medications.

Discussion

Kawasaki disease also known as mucocutaneous lymph node syndrome is a vasculitis in infants and children KD predominantly involves the small and medium sized arteries. Coronaries are the commonest to be involved. The exact cause of this disease is not known but is a consequence of exaggerated immunological events following a pathogen in a genetically predetermined individual[1]. The first week is characterized by neutrophilic arteritis which forms saccular aneurysm followed by fusiform aneurysm in months. Over the years stenosis and thrombi may occur. Diagnosis of kawasaki disease is based on clinical and laboratory criteria. Fever for 5 days with 4 of the following criteria

1. Bilateral nonexudative conjunctival injection with limbal sparing;
2. Erythema of the oral and pharyngeal mucosa with strawberry tongue and red, cracked lips;
3. Edema and erythema of the hands and feet;
4. Rash of various forms and
5. Nonsuppurative cervical lymphadenopathy, usually unilateral, with node size >1.5 cm.

Alternate criteria for diagnosis includes Fever for at least five days and two or three principal features; coronary artery abnormalities on transthoracic echocardiography[2]. Incomplete (atypical) Kawasaki disease occurs in children with fever lasting five or more days and with two or three of these findings[1]. KD is a systemic illness with involvement of gastro intestinal, musculoskeletal, central nervous system , genito urinary and other systems. The acute phase presents with fever and other manifestations of the illness for the first 2 weeks. This is followed by desquamation and thrombocytosis with development of coronary aneurysms for 3 weeks. Sudden death has been reported in this phase of illness. Convalescent phase shows resolution of the signs and continue till the ESR becomes normal. The laboratory findings of KD include, neutrophilic leucocytosis, anemia,

thrombocytosis in the second week, elevated liver transaminases, sterile pyuria, pleocytosis of CSF. Common viral ,bacterial infections, rheumatologic illness, Echo features include lack of tapering, perivascular brightness, ectasia decreased ventricular function, mild valvular regurgitation, pericardial effusion and aneurismal changes . Management is aimed at prevention of coronary involvement. In untreated the occurrence of coronary aneurysm varies from 15- 25 %. In children with treatment, transient changes are reduced by 5 % and giant aneurysm by 1% .

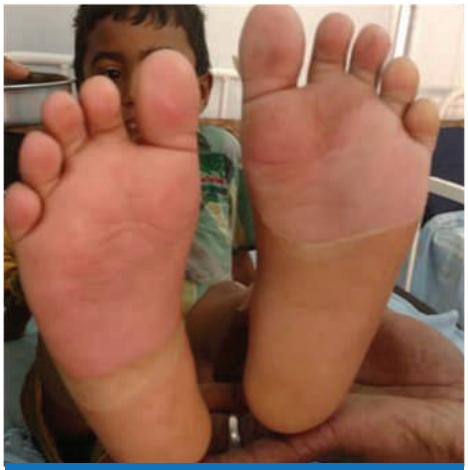

Fig 1 - peeling of skin over the soles

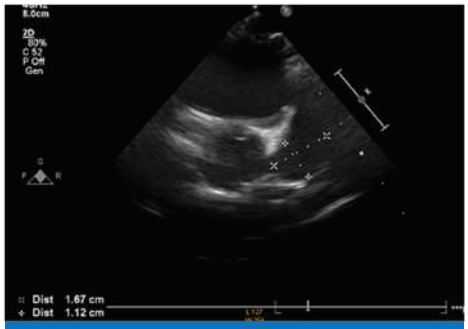

Fig 2 - Left main coronary artery dilatation

Intravenous (IVIG) at a dose of 2g/kg given over 10-12 hours is the treatment of choice to prevent the coronary complications. Recent meta analysis has revealed that addition of corticosteroids to IVIG help to prevent coronary abnormalities[3]. If the child continues to have persistent symptoms or persistent fever after 36hours of IVIG and high dose aspirin then a repeat dose of IVIG may need to be given. Therapy is most useful in the first week of illness. However the therapy with IVIG can still be tried if the child presents with fever and raised ESR and CRP. Aspirin at 80-100 mg/kg /day in 4 divided doses till afebrile for 48-72 hours and then at a dose of 3-5 mg/kg/day for 6-8 weeks. If coronary abnormalities are seen then aspirin is continued along with oral clopidogrel and warfarin. Child needs to be followed up with repeat echo at diagnosis, 2 weeks and 6-8 weeks if initial echo is normal[4]. Children on long term aspirin should receive the influenza vaccine, and varicella vaccination. The long term follow up and intervention are similar to adults with coronary artery disease. KD may recur in 1- 23% of children. The aneurysms regress in 50% by 1-2 years. However giant aneurysm are less likely to regress and may lead to stenosis and thrombosis. Coronary artery aneurysms more than 8mm size are unlikely to regress[5]. Options for second-line treatment include additional IVIG, Intra Venous methylprednisolone pulse (IVMP), prednisolone (PSL), IFX, ulinastatin (UTI), Cyclosporine A, Methotrexate and plasma exchange (PE).But for IVIG , none has been recommended with strong evidence[5] . All children with KD need to follow heart healthy diet, exercises,limit injury prone activity, monitor lipid levels and avoid narcotics.

The child described had giant aneurysm and was a delayed referral to the Institute. The child had elevated ESR and CRP and hence was given IVIG. Literature evidence shows that IVIG can be given up to 10 days of illness or later if a patient has persistent fever, aneurysms, or inflammation[1]. Since this child is at high risk for stenosis and thrombosis leading to myocardial infarction the child is on long term aspirin, clopidogrel and warfarin.

Prevention of coronary aneurysm in KD needs high degree of suspicion for an earlier diagnosis .

Acknowledgement

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Conflict of interest : Nil

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