KAWASAKI DISEASE (SYNDROME)

- OUR EXPERIENCE -

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Abstract : Kawasaki disease also known as mucocutaneous lymph node syndrome is a generalised vasculitis of unknown etiology. Presenting a review of the literature and using our cases for illustration we discuss some of the pertinent features in the evaluation and management of patients with this condition. 2 cases of Kawasaki disease have been evaluated and treated in the department of ENT and Paediatrics who responded satisfactorily to treatment with I.V. Immunoglobulin and Aspirin. The case reports have been presented in view of the rarity of reporting of similar cases in India and the need for early diagnosis and prompt treatment to prevent coronary artery aneurysm and other serious complications.

Keywords : Kawasaki Disease, Generalised Vasculitis, I.V Immunoglobulin, Aspirin, Coronary Artery Aneurysm

INTRODUCTION:

Kawasaki disease, also known as mucocutaneous lymph node syndrome or infantile periarteritis nodosa / vasculitis is a generalized vasculitis of unknown etiology. Tomisaku Kawasaki in Japan first described it in 1976. We report 2 Cases of Kawasaki disease which have been sparingly reported In India and such similar cases could be misdiagnosed as routine upper respiratory tract infection and treated by routine Conservative Methods. Kawasaki disease is a non-specific disease of unknown etiology reported mainly from Japan and U.S.A.

Principal diagnostic features include the following: High grade fever $>102^{\circ}\text{F}$ presenting for more than 5 days and not responding to antipyretics / antibiotics, non purulent [aseptic] conjunctivitis, bright red chapped / cracked lips, reddish oral mucous membrane, strawberry tongue, reddish palms of the hands and soles of the feet, swollen hands and feet, beau's lines (transverse grooves in the nails), rashes resembling erythema multiforme, lymphadenopathy particularly in neck area, joint pain (Arthralgia) and swelling usually symmetrical, peeling of palms and soles, occurring later in the illness.

LAB FINDINGS INCLUDE:

Elevated WBC count with predominance of immature and mature granulocytes, normocytic Anaemia, thrombocytosis usually occurring during second or third week of illness, elevated ESR, C-reactive protein levels and alpha one anti trypsin, mild to moderate pyuria and proteinuria.

LAB FINDINGS IN COMPLICATED KAWASAKI DISEASE INCLUDE:

Elevated cardiac enzyme levels in myocardial infarction, ECG changes - Tachycardia, prolonged PR interval, ST-T wave changes and reduced R wave voltage in myocarditis. Q wave or ST-T wave changes may indicate myocardial infarction. Chest X-Ray findings : Cardiomegaly, subclinical pneumonitis.

Echocardiography: May show features of coronary artery aneurysm, myocarditis, valvulitis and pericardial effusion.

TREATMENT:

Treatment must be prompt and should be started within 10 days of onset of the fever to prevent serious cardiac complications. **I.V. Immunoglobulin** is given in a dose of 2gms/kg over 10-12 hrs. **Aspirin** is given orally in the acute phase as 80- 100 mg/kg in tid or qid dose. After 36hrs, 3-5 mg/kg/day is given every morning for 3 months. Follow up includes clinical examination after 1 week of discharge from hospital and repeat 2D ECHO CARDIOGRAM after 3 wks, 3 months, 6 months and 1 year.

CASE REPORT [1]:

A female child aged 11 years was evaluated in the department of E.N.T and paediatrics in March 1998 with the following symptoms: fever and sore throat not responding to conservative treatment since 7 days. Right sided neck pain and torticollis since 5 days. Clinical examination revealed right sided tender cervical adenopathy with Right tonsil pushed medially. The next day, patient developed bilateral aseptic conjunctivitis, red straw berry tongue and erythema of hands and feet. Body temperature ranged from 99°F to 102°F which was not responding to antipyretics.

INVESTIGATIONS REVEALED :

C.T scan of Neck with contrast showed lymphadenopathy involving the Nasopharynx, oropharynx, Para pharyngeal space and deep parotid space. C - reactive protein was positive (96 mg/lit). Total WBC was 15,500/ μl . ESR was 66mm/hour. Platelet count was 4.10 lakhs. Neutrophils - 82%. Urine protein - 1+. ECG and 2D cardiac ECHO were within normal limits.

A diagnosis of KAWASAKI DISEASE was made and the patient was given I.V. Injection of immunoglobulin, 1 gram/day X 5 days and Tab. Aspirin, 375 mg 2 tab, tid for 7 days. The child made an uneventful recovery and a 2D cardiac ECHO done after 3 weeks showed no coronary artery abnormalities.

CASE REPORT [2]:

A male child aged 5 years presented with the following features: High grade pyrexia of 9 days duration not responding to conservative treatment including antibiotics, generalised oedema and swelling of hands and feet, erythematous maculopapular rash : starting over the trunk and extending all over the body : 4 days duration. Dysentery of 1 day duration.

CLINICAL EXAMINATION SHOWED :

Non purulent conjunctivitis, Oral fissures, cracked lips, Oedema and redness of hands and feet, Red tongue, Neck swelling (lymph nodal swelling) and Peeling of skin: 12 days later.

INVESTIGATIONS REVEALED:

Total count :13,036/ μ l, mild shift to left and moderate atypical lymphocytes, Platelets : 2.1 lakhs in the 2nd week and 5.2 lakhs in the 3rd week, ESR :45 mm/hr, RBC : Anisocytosis, hypochromia , microcytosis, ECG and Echo Cardiogram : Normal limits and Urine showed plenty of pus cells and urinary protein : 114 mg/ day.

TREATMENT:

A provisional diagnosis of Kawasaki disease was made and patient was given I.V immunoglobulin 400 mg /kg/4 days and aspirin 325 mg /4th hourly for 1 wk followed by twice daily for 2 wks later, once daily for 4 wks. The child became completely afebrile after 4 days of starting the treatment. At the time of discharge, repeat echocardiogram was done which was within normal limits. On follow up visits, it was noticed that the child did not have any cardiac complications.

DISCUSSION

Kawasaki disease is an acute self limited vasculitis that occurs in children of all ages. There is no specific diagnostic test (Burns JC.Glode MP, 2004). A ubiquitous virus of the gamma herpes virus family is a likely etiological agent for KD in genetically susceptible individuals (Shingadia D; Bore et al 2002). Many hypotheses regarding the etiology of KD exist. The debate continues as to whether the inflammatory response of KD results from a super antigen or a conventional antigen (Guzman - Cottrill JA 2004).

Kawasaki disease is a febrile systemic vasculitis complicated by coronary and peripheral arterial aneurysm in 20-35 % of untreated patients. There is still no diagnostic test available for Kawasaki disease, and the diagnosis is based on clinical criteria after the exclusion of other diseases presenting with high persistent fever. (Cimaz R. Falcini.F) If there is no response to two standard doses of I.V Immunoglobulin treatment, pulse methyl prednisolone therapy should be considered (Hung JJ.Chiu CH.2004).

When there was no response to I.V Immunoglobulin and Methyl prednisolone, Cyclosporine administration was tried. The study endorses the role of specific cellular and humoral immunity in Kawasaki Disease (Kuijpers J.W, et al 2003). Coronary bypass operation utilizing the pedicled internal thoracic artery has been suggested as a safe and reliable surgical modality for coronary

artery sequelae in children due to Kawasaki Disease. (Kitamura. S).

A case of Kawasaki Disease with acute renal failure which had recovered without the need for dialysis was reported by Banany P J et al in 2002.

COMPLICATIONS :

Coronary artery aneurysms develop in up to 25% of untreated patients. These complications are seen in less than 5% of patients treated with I.V Gamma Globulin. Other complications like myocardial infarction in children, congestive Heart failure, myocarditis, pericardial effusion have also been reported. 80% of cases are younger than 4 years, whereas one of our cases was aged 11 years. In both of our cases, the clinical criteria of KD described in the literature have been satisfied. These two cases remind us the need to have a suspicion of Kawasaki disease when the patients presenting with the above clinical features are not responding to routine conservative treatment. Presently we do not have a definitive diagnostic test for detecting Kawasaki disease.

CONCLUSION:

The case reports have been presented in view of the rarity of reporting of similar cases in India and the need for early diagnosis and prompt treatment with I.V.Immunoglobulin and Aspirin to prevent serious cardiac complications in children. Also there is a need to create awareness among the medical fraternity about this disease so that the children affected with this condition may get early appropriate treatment by prompt diagnosis.

Other illnesses with Fever and Rash resembling Kawasaki disease should be excluded by proper history and Physical examination because of lack of a specific and sensitive diagnostic test. There is a need for devising a proper diagnostic test in order to avoid misdiagnosis leading to suffering from a preventable morbidity and mortality. Adequate research is to be done to obtain a proper diagnostic test which can identify children who need treatment. There is also a need to identify the etiology of this condition.

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