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Atypical and incomplete Kawasaki: A silent spectrum with significant coronary changes - experience in an Indian tertiary care hospital

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The diagnosis of Kawasaki disease still remains elusive due to the spectrum of atypical and incomplete presentations. Here we describe some atypical and incomplete presentations of the disease, all accompanied by significant coronary changes in the form of aneurysm, ectasia, lack of tapering and increased perivascular hyperintensity. All except one responded to IVIg with much improvement in the coronary artery size on follow up echocardiography. There were 17 patients diagnosed with atypical and incomplete features over a span of two years (Of them, incomplete Kawasaki disease present in 11 and atypical presentation in 6). There were eight infants, five in age group 1-5 years and four >5 years). The youngest was only 42 days old and the oldest 9 years 7 months. The incidence of incomplete presentation with fever and one or two other diagnostic criteria was seen more commonly in the small infants. This group also had more extensive coronary involvement like multiple coronary artery involvement, moderate to large aneurysms and tortuous, dilated coronaries. Red lips and oral mucosa was the commonest finding (present in 9 out of 11 cases), followed by nonpurulent conjunctivitis (6 of 11) and cervical lymphadenopathy (5 of 11). Platelet count was high or high normal at presentation in all infants. One infant developed macrophage activation syndrome and despite treatment with Dexamethasone, Etoposide and Cyclosporine developed multiorgan failure and died. This child also had severe ventricular dysfunction. The atypical presentations like pericardial effusion, joint swelling and staphylococcal septicemia, coexistence of dengue infection (positive serology and few clinical features) and shock occurred more common in the older age group. This group had a delayed rise in platelets increasing on the second or third weeks in five cases). Aneurysms were also less common in the older children with dilatation, lack of tapering and marked perivascular hyperintensity dominating the picture. They also had more of single artery involvement; predominantly left anterior descending (LAD) followed by right coronary artery (RCA) and left main coronary artery (LMCA). Three cases with pericardial effusion had ST elevation in ECG. There was no regional wall motion abnormality in any of the echoes at presentation or on follow up. Though the number of cases was small, there was an unequivocal extensive and severe degree of coronary involvement in the infants presenting with incomplete Kawasaki disease. Also, in the older age group, due to atypical presentation and the fact that laboratory parameters like platelets and inflammatory markers were inconclusive, there was a diagnostic dilemma. Echocardiography was decisive for the diagnosis and treatment.

Biography

Debadatta Mukhopadhyay has completed her MBBS with Honors from Kolkata Medical College, India. After completing her DCH from Institute of Child Health, Kolkata, she has completed her MD and then MRCPCH. She has trained in Pediatric Intensive Care and is also a BPICC Instructor. She has trained in Pediatric Cardiology in RN Tagore International Institute of Cardiac Sciences, Kolkata and as Fellow Pediatric Cardiology at University Hospital, Southampton, UK. Currently, she is working as an Assistant Professor of Pediatrics in, Kolkata Medical College, India where she also runs pediatric cardiology services. Her special interests are complex congenital heart diseases and fetal echo.

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