Kawasaki Disease: Is Atypical More Common than Typical

Kawasaki Disease (KD) is a vasculitic disorder and has replaced rheumatic fever as the commonest cause of acquired heart disease in children(1,2). In India, KD perhaps is more common than it has been diagnosed and reported.

The profile of KD patients admitted in the Institute of Child Health, Kolkata in the period from May 2003 to June 2004 was taken up for this study. A total of six patients of either typical or atypical KD were included in the study. Detailed clinical evaluation and echocardiography was done in all the patients. Age varied between 3 and 6 years. Two of them (Case II & V) presented with fever of 10 and 11 days respectively. The remaining had fever of less than 10 days duration. Three of the cases (Case I, II, III) had less than 4 of the classical clinical features of typical KD and were diagnosed only after echocardiography showing characteristic coronary artery findings. These three cases were labeled as Atypical KD. One case (Case V) fulfilled all the clinical criteria of KD, but did not develop any coronary artery abnormalities. The echocardiographic findings are described in *Table I*. LMCA diameter above 2.5 mm is considered dilated, any value above 2 mm for RCA is considered dilated and any value above 2.3 mm for LAD is considered dilated(3). Repeat echocardioghraphy at 2 months of cases I, II and V showed normal coronary arteries with minimal luminal irregularity. Case VI at 3 months showed still dilated LMCA (3.1 mm).

Atypical or incomplete KD cases are those who develop fever with fewer than 4 of the classical clinical features of the disease, but develop coronary artery disease. In this study 3 of the 6 cases were atypical KD patients.

The diagnosis of KD or atypical KD is largely clinical and a high degree of suspicion needs to be borne in mind. Since KD is not being diagnosed frequently in our country(4,5) majority of the affected patients are at present being left untreated rendering them liable to coronary complications, contributing to the total load of coronary artery disease in young adults who have no other risk factor. Routine use of echocardiography, in patients with features suggestive of KD, though not fulfilling all the criteria to be labeled KD, might go a long way in early detection of this potentially devastating cardiac complication

TABLE 1– *Echocardiographic Findings in the Cases*

Cases	LMCA (mm)	RCA (mm)	LAD (mm)	Ejection fraction (%)
I	4	2 (N)	2 (N)	63%
II	2 (N)	1.5 (N)	4.5	67%
III	3.5	1.2 (N)	2.2 (N)	65%
IV	2.2 (N)	4	2 (N)	65%
V	2 (N)	1.7 (N)	1.9 (N)	70%
VI	3.2	2 (N)	2.6	61%

LMCA = Left Main Coronary Artery, RCA = Right Coronary Artery, LAD = Left Anterior Descending Artery, N = Normal

and initiation of early prophylaxis might prevent some of the unexplained myocardial infarctions of early life.

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Gelastic Epilepsy and Hypothalamic Hamartoma

Gelastic epilepsy is characterized by episodes of loud, hollow, mirthless, stereotyped, forced laughter(1). The patient may stare and giggle briefly without any other motor manifestations. Hypothalamic hamartoma is most often the cause of gelastic seizures(1,2). Lesions in ventro-medial nucleus of hypothalamus cause hyperphagia and obesity, not usually described with hypothalamic hamartoma. We report a case of gelastic seizures with precocious puberty and obesity in a child with hypothalamic hamartoma.

A 1½-year-old male child born out of non consanguineous marriage presented with progressively increasing episodes of unprovoked stereotyped laughter lasting for few seconds since the age of 5 months. Along with this, child was having excessive weight gain and signs of premature development of secondary sexual characters as hair growth in face, axilla and pubic area, with enlarged penis and large testis for his age. He was also having concomitant squint. There was no other neurological deficit. A clinical diagnosis of gelastic seizures with obesity and precocious puberty was made. MRI brain showed a lobulated isointense solid lesion on TIW and relatively hyper intense on T2W and FLAIR images noted in the region of hypothalamus between infundibular stalk and mamillary bodies with splaying of cerebral peduncle posteriorly measuring about 2.3 × 2.3 × 2.8 cm. The lesion did not enhance on post contrast study (*Fig.1*). The morphology is suggestive of hypothalamic hamartoma. Patient was started on sodium valproate and dose increased upto 1000 mgs per day and his seizure frequency was not reduced signifi-