Dissecting Aortic Aneurysm with Marfan Syndrome

Aortic dissection is a rare entity in childhood and a high index of suspicion is required for prompt management. It should be considered in children with Marfan syndrome complaining of chest pain.

A 15 years male was hospitalized with a month long history of recurring episodes of severe left sided chest pain, five days history of cough and fever. The pain was piercing in nature, radiating to the neck, left shoulder and back, associated with profuse sweating and occasionally with vomiting. His father had expired two months ago after a similar brief illness at the age of 38 years.

Examination revealed an asthenic built child with height of 162 cm, arm span 174 cm, arm span-to-height ratio 1.074 and US:LS ratio 0.82. He had high arched palate, arachnodactly, positive wrist and thumb signs. There were signs of consolidation over left base. Cardiovascular system examination revealed a short diastolic murmur in aortic space. Skiagram revealed a massive consolidation of left lower zone. Clinical and radiological features of consolidation resolved following antibiotic therapy. However repeat radiological evaluation revealed an underlying mediastinal widening. High resolution computerized tomography revealed a large aortic aneurysm **(*Fig.* 1)**. Color Doppler revealed aortic dilatation just after the origin of left subclavian artery and a large intimal flap just before the origin of superior mesenteric artery.

The catastrophic cardiovascular events in Marfan syndrome, most often a result of aortic aneurysm and dissection, result in an average age expectancy of 32 years [1]. Several studies have validated the role of beta-blockers in retarding aortic root dilatation and preventing aortic dissection/rupture [2]. Therapy with calcium channel blockers, angiotensin inhibitors and angiotensin II receptor antagonists have also shown similar effects [3]. Successful surgical intervention has revolutionized the management of these patients, resulting in median survival of 61 years [4].

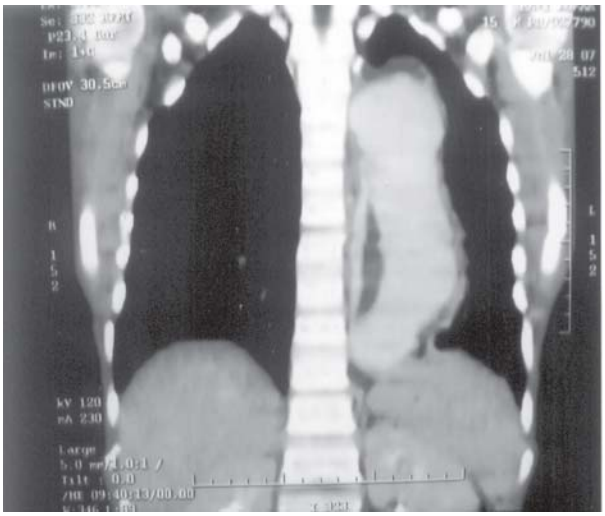

FIG. 1 *High resolution CT revealing a large aortic aneurysm arising from aortic knuckle and extending till the celiac axis with a large posterolateral dissecting part in the left paraspinal gutter.*

The patient showed symptomatic improvement following beta blockers and was referred for surgical correction, which was however denied on grounds of extremely high operative risk. The patient is surviving after two years of follow up.

Rekha Harish and Ashu Jamwal,
Department of Pediatrics,
Govt. Medical College, Jammu, India.
kkrhdang@gmail.com

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