

IMAGES IN PAEDIATRICS

Superior mesenteric artery syndrome: a rare complication in a child with Marfan syndrome

A 14-year-old girl presented with sudden onset abdominal pain and distension with bilious vomiting. She had no significant prior history or weight loss. On examination, bowel sounds were normal. She was noted to have a typical marfanoid habitus with severe kyphoscoliosis (figure 1), arachnodactyly, positive thumb and wrist sign. Echocardiogram showed mitral valve prolapse, tricuspid regurgitation and normal-sized aortic root. Laboratory investigations were normal. Barium study showed dilated proximal duodenum and an abrupt cutoff near the spine (figure 2). On endoscopy, the third part of the duodenum could not be negotiated, where pulsations of superior mesenteric artery (SMA) were seen anteriorly. A diagnosis of SMA syndrome (SMAS) was confirmed. A trial of conservative management failed and a retrocolic side-to-side duodenojejunostomy was performed. Unlike classical SMAS, the duodenum was trapped between the spine and SMA. Postoperatively, the child remained asymptomatic at 1-year follow-up.

SMAS is a rare cause of duodenal obstruction secondary to compression by the SMA.¹ It has not been reported in Marfan syndrome (MFS) earlier, where gastrointestinal complications are noted to be rare.² SMAS can present with acute symptoms, as in the present case, or with chronic nonspecific postprandial abdominal pain or fullness, vomiting or early satiety, leading to a diagnostic dilemma.^{3 4} Asthenic build, rapid weight loss and spinal deformities are major risk factors for SMAS.^{1 5} In



Figure 2 Barium study showing grossly distended stomach and abrupt cutoff in duodenum near the spine without distal flow of contrast. Note severe kyphoscoliosis.

children with scoliosis coupled with adolescent skeletal growth, the aortomesenteric angle may become narrow, leading to a risk of SMAS.⁵ Moreover, MFS patients are underweight, and thus, may have less periduodenal fat to cushion and protect the duodenum in the SMA angle. Thus, the present case elicits the unusual causal association between these two syndromes.

Figure 1 Clinical photograph showing severe kyphoscoliosis in a patient with marfanoid habitus.



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