An Unusual Cause of Superior Mesenteric Artery Syndrome in a Young Boy

Superior Mesenteric Artery (SMA) syndrome is an uncommon clinical entity characterized by compression of third part of duodenum against aorta by SMA resulting in high intestinal obstruction and often posing a diagnostic dilemma. It is usually seen in middle aged frail people who are bedridden and have had recent weight loss. We present a case of SMA syndrome in a 14 year young boy caused by dual pathology of SMA compression and malrotation of caecum and ascending colon aggravating the compression over duodenum.

A 14 year young boy reported with history of recurrent pain abdomen of ten years duration gradually worsening for last one year. Pain was colicky in nature following meal and relieved with vomiting. Initially he used to get relief of pain on lying in prone position but subsequently change in position was not of much help. Vomitus contained undigested food mixed with bile. He had lost 7 kg of body weight in the last one year.

Clinically, he was thin, 119 cm tall, and weighted 30 kg. Abdomen was scaphoid with mild epigastric tenderness. Succussion splash was present. There was no organomegaly.

Haematological investigations were normal except haemoglobin of 8 gm/dL. Plain radiograph abdomen showed distended fluid gas filled stomach and duodenum. USS abdomen suggested distended stomach. Upper gastro intestinal (GI) contrast study showed dilated stomach, first and second part of duodenum with abrupt vertical cut-off at the level of third part of duodenum (Fig.1). CECT scan demonstrated distended stomach and duodenum with positive whorl sign suggestive of midgut volvulus (Fig. 2).

Exploratory laparotomy revealed extrinsic obstruction of third part of duodenum. Caecum and ascending colon were intraperitoneal and were rotated to left from middle of the transverse colon. Duodenojejunostomy was performed to bypass the obstruction. The child had an uneventful post operative recovery and gained 6 kg of weight in next six months.

SMA syndrome was first described by Rokitsky in 1861. In 1921, Wilkie [1] provided a pathophysiological description and renamed it 'Chronic Duodenal Ileus'. About 400 cases have been described since then. It can also occur due to peritoneal bands and short ligament of Trietz [2]. Diagnosis is often made by exclusion of other causes of upper GI obstruction. The incidence varies from 0.013 to 0.3% [3]. The compression of duodenum is due to sharp decrease in angle between Aorta and SMA (6 to 16°), the normal angle being 35 to 65°. Rapid and severe weight loss causes diminution of fatty cushion between Aorta and SMA resulting in diminished angle and compression of duodenum. Other causes implicated include high insertion of duodenum at the ligament of Treitz, congenital low origin of SMA, a short ligament of Treitz and peritoneal adhesions.

Anatomical anomalies very rarely cause precipitation of narrowing of aortomesenteric angle resulting in mechanical obstruction of duodenum. In this reported case, anatomical anomalies of caecum and ascending colon resulted in reducing the angle between aorta and SMA, which is a very rare combination and has not been previously reported.

Fig. 1 : Upper GI contrast study showing dilated duodenum with abrupt vertical cut off at third part of duodenum.

Fig. 2 : CECT showing positive whorl sign suggestive of midgut volvulus.

References

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Contributed by

Col Niranjana Dash*

*Senior Advisor (Surgery), Command Hospital (NC), C/o 56 APO.