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Clinical Vignette

Cecal volvulus in a child with Cornelia de Lange syndrome

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CASE PRESENTATION

A 14-year-old boy, known case of Cornelia de Lange syndrome (characteristic dysmorphism, and developmental delay) was seen in the pediatric outpatient department with a history of non-bilious vomiting for 4 weeks, several times a day. He had not passed stools for several days and was managing to take fluids. He was managed conservatively for sub-acute intestinal obstruction elsewhere before coming to our center.

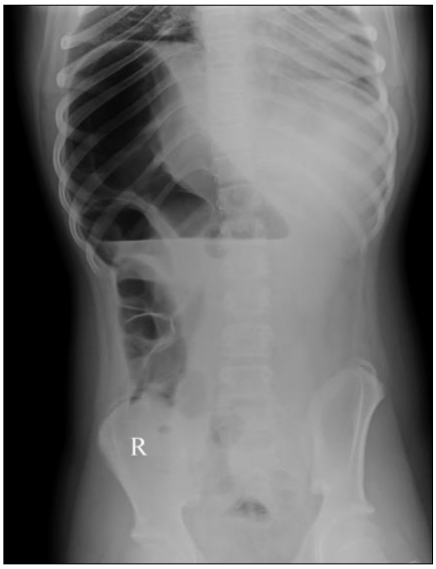

Figure 1: X-ray abdomen erect, showing multiple air-fluid levels and a huge air shadow in the right upper abdomen.

He had been operated for cleft lip and palate in infancy. The initial examination revealed adequate hydration and distended abdomen with absent bowel sounds. Abdominal X-ray (Fig. 1) revealed air-fluid levels, with a hugely dilated loop of bowel on the right side of the abdomen. Diagnostic laparoscopy revealed dilated large bowel in the right hypochondriac region and hence a laparotomy was performed. Laparotomy showed a hugely dilated terminal ileum, cecum, and ascending colon and volvulus of the cecum (Fig. 2). A right hemicolectomy was performed and end to end anastomosis performed between terminal ileum and ascending colon.

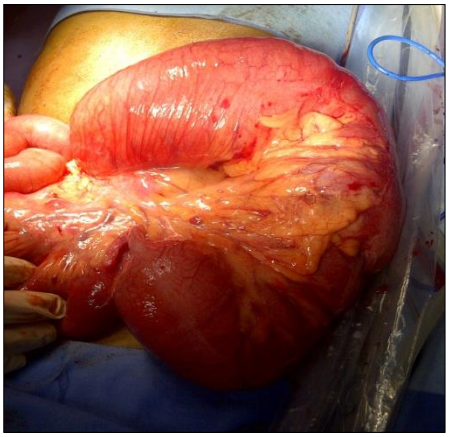

Figure 2: Operative figure showing hugely dilated cecum and colon.

The patient had postoperative wound sepsis with Klebsiella which was treated with appropriate antibiotics and the patient was discharged on 8th postoperative day.

DISCUSSION

Cornelia de Lange syndrome is a multi-system genetic disorder characterized by synophrys, low-set ears, small and widely spaced teeth, and a small and upturned nose; many affected may have behavior problems similar to autism. A surgical cause has to be ruled out in children presenting with bilious vomiting. Cecal volvulus is a rare cause of bowel obstruction in children.[1] Plain X-ray abdomen and barium enemas can aid in the diagnosis of

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this condition. Taneja et al. found 6 cases of cecal volvulus out of 189 children who were operated for acute intestinal obstruction.[1] In another case series of 28 children operated for intestinal obstruction only one case of cecal volvulus was reported, the authors reported idiopathic, bands or malrotation as causes of obstruction.[2] Rare association with the Cornelia de Lange syndrome has been reported [3] and our case is the first from the Indian subcontinent. Fundoplication,[4] laparoscopy,[5] and antegrade colonic enemas[6] have been reported as a predisposing factor for developing cecal volvulus. Mental disability is another known risk factor in these children and of the 40 reported cases of cecal volvulus, 13 children had a neuro developmental delay.[7] Multiple treatment options exist including conservative management, cecopexy, cecostomy and resection with end to end anastomosis, there is no consensus on the treatment and data suggest that resection and end to end anastomosis seems to have a better long term outcome.[3-7] This case highlighted the diagnosis of cecal volvulus and the rare but known association with Cornelia de Lange syndrome.

Consent to Publication: Author(s) declared taking informed written consent for the publication of clinical photographs /material (if any used), from the legal guardian of the patient with an understanding that every effort will be made to conceal the identity of the patient, however it cannot be guaranteed.

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