Computed tomography diagnosis of malrotation with midgut volvulus and superior mesenteric vein thrombosis beyond infancy

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ABSTRACT

Malrotation can be difficult to diagnose beyond the newborn period because of its non-specific symptoms and clinical findings. We present an unusual case of malrotation with midgut volvulus and superior mesenteric vein thrombosis in an adolescent. An 11-year-old girl presented to the paediatric emergency department with persistent vomiting, dyselectrolytemia, and metabolic alkalosis. An unremarkable abdominal radiograph and ultrasonography examination prompted a computerised scan of the abdomen. The diagnosis of malrotation with midgut volvulus and superior mesenteric vein thrombosis was made. The findings were confirmed on laproscopy and the patient underwent successful Ladd’s procedure. This case report emphasizes the importance of imaging, especially computed tomography, in making accurate diagnosis of malrotation and its complications, beyond the newborn period.

Keywords: Computed tomography, malrotation, superior mesenteric vein thrombosis, volvulus.

INTRODUCTION

Midgut malrotation is a congenital anomaly in the embryological development of the fetal intestinal rotation. More than 90% of patients present by 1 year of age. Midgut malrotation beyond infancy is rare and its incidence has been reported to be between 0.0001% and 0.19%. The diagnosis in this age group is fraught with immense difficulty. Failure to diagnose this condition promptly can lead to catastrophic complications. We report a case of an adolescent with acute on chronic presentation of malrotation with midgut volvulus and superior mesenteric vein thrombosis. The patient presented in a critical condition with severe dyselectrolytemia and metabolic alkalosis.

CASE REPORT

An 11-year-old female presented to the paediatric emergency department with recurrent episodes of bilious vomiting and pain abdomen for the last 15 days. The child was a normal term delivery and developed recurrent episodes of vomiting, till the age of 6 months. The condition of the child improved spontaneously and subsequently there were occasional episodes of vomiting and pain abdomen.

The general physical examination was unremarkable except for pallor and reduced weight. The laboratory examination was suggestive of severe dyselectrolytemia with hyponatremia, hypokalemia, metabolic alkalosis, and ketonuria.

The child was being treated as case diabetic ketoacidosis outside our institution, based on clinical presentation and raised blood glucose levels on two occasions with ketonuria.

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A supine abdominal radiograph and erect radiograph of chest were unremarkable. The ultrasound of abdomen did not reveal significant findings. Considering the clinical diagnosis of obstruction, a contrast-enhanced computed tomography (CECT) of abdomen was planned.

CECT abdomen showed small bowel loops in the right and large bowel loops in left abdominal cavity suggestive of malrotation [Figure 1]. There was inversion of superior mesenteric artery (SMA) and superior mesenteric vein (SMV) relationship, SMA seen left of SMV [Figure 2]. The stomach and duodenum were mildly dilated suggestive of obstruction. The whirlpool sign, clockwise swirling of bowel and mesentery along the axis of SMA, characteristic of volvulus was seen [Figure 3]. The SMV was thrombosed and replaced by multiple collaterals in the mesentery [Figure 4]. The uncinate process of pancreas was hypoplastic. Based on CECT findings, diagnosis of malrotation of bowel with chronic midgut volvulus and superior mesenteric vein thrombosis was made.

The patient was taken up for immediate laparoscopy, which confirmed malrotation with 270° twist of the bowel. There were dense Ladd's bands and adhesions, with thrombosis of SMV. An engorged mesenteric...
Congenital malrotation of the midgut often presents within the first month of life. Paediatric radiologists are therefore consciously aware of this abnormality and its associated imaging features. Because presentation is non-specific and the index of suspicion is very low in the older population, the clinical diagnosis is usually not considered in the initial evaluation. Prompt recognition and surgical treatment usually leads to a successful outcome. Delay in diagnosis can lead to grievous complications of intestinal necrosis.

Radiographic diagnosis of malrotation with volvulus had traditionally been made by upper gastrointestinal contrast studies in infants. But in older children, when clinical symptoms are non-specific, imaging findings on conventional radiograph and ultrasonography may be inconclusive. CECT plays a significant role in diagnosing this unusual entity as well as demonstrating its complications.

CT scan is increasingly used preferentially and is now considered the investigation of choice, providing diagnostic accuracy of 80%.[3] CT scan shows the abnormal anatomical arrangements of the midgut with the duodenum not crossing the spine, the small bowel loops being placed in right abdominal cavity and the large bowel loops on left. Deviation from the normal positional relationship of SMV and SMA, originally described by Nichols and Li,[4] also serves as a useful indicator for diagnosis of midgut malrotation. Patients with gut malrotation often have an underdeveloped or absent uncinate process of the pancreas.

The CT appearance of midgut volvulus is characteristic. The shortened mesentery allows the small bowel and mesentery to twist and wrap around the narrowed SMA pedicle to create a distinctive “whirlpool” appearance on CT scan. This pattern was first described by Fisher in a patient with midgut volvulus.[5] CT scan may also demonstrate small bowel obstruction secondary to internal herniation in patients with malrotation.

An unusual complication of malrotation with chronic midgut volvulus is superior mesenteric vein thrombosis.[6] Approximately 15% of patients with malrotation and volvulus develop complete obstruction of the mesenteric vessels resulting in bowel infarction, most commonly in infancy. However, patients with chronic malrotation and intermittent volvulus usually have significant collateral mesenteric circulation. This prevents the bowel infarction seen in acute vascular occlusion.

Surgical management of intestinal malrotation was first described by Ladd in 1936 and remains the mainstay of management today.[7] Although symptomatic midgut malrotation requires surgical intervention in all cases, management of asymptomatic individuals is still controversial. It is increasingly being argued that all suitable patients with intestinal malrotation should undergo surgical correction, irrespective of age, as it is impossible to predict which patients may develop catastrophic complications.[8]

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**REFERENCES**