Clinical History:

A full term two days old Saudi newborn boy presented with bilious vomiting. He was well in himself and had stable observations. On examination, his abdomen was soft, non-tender and not distended. Routine blood tests were normal. He underwent upper gastrointestinal contrast study which showed a rare cause of his condition.

Imaging Findings:

Initial plain abdominal X-ray showed mildly dilated stomach with paucity of air in distal large bowel. Upper gastrointestinal contrast study was performed by a dedicated paediatric radiologist which showed dilated stomach and proximal duodenum with hold-up of contrast at 2 ½ hours. Patient was kept under careful monitoring due to chances of aspiration in a vomiting neonate, with subsequent supervised gastric emptying through nasogastric tube. Ultrasound showed a heterogeneous epigastric mass. The colour Doppler revealed characteristic clockwise winding of superior mesenteric vein around the corresponding artery describing the typical “Whirlpool Sign” which was pathognomonic of midgut volvulus. On the basis of radiological findings laparotomy was performed which confirmed the diagnosis of malrotation with midgut volvulus. Interestingly 540 degree twist of small bowel was noted. Ladd’s procedure was performed with untwisting of bowel. Total parenteral nutrition was commenced post-operatively and the patient was discharged home after three weeks.

Discussion:

Malrotation with midgut volvulus is an acute paediatric surgical emergency. It results from the failure to rotate midgut derivatives around their mesenteric axis of superior mesenteric artery (SMA) during first trimester of fetal growth. The incidence of midgut malrotation is 1 in 500 live births with male to female ratio of 2:1 [1]. However, in symptomatic malrotation, the incidence of midgut volvulus is reported to be 42.1%. [2]. Sixty percent of patients diagnosed with malrotation are neonates with the remaining 40% equally divided between infants and children [3].
can be associated with other congenital anomalies, such as intestinal atresia, omphalocoele and gastroschisis. It is thought that short mesenteric attachment brings the duodenum and the caecum nearer than usual, resulting in twisting of the midgut at distal duodenum and proximal jejunum on the SMA trunk which leads to duodenal obstruction and bowel ischaemia. Necrosis can occur within 2 hours of initial presentation.

Bilious vomiting is invariably the most common presentation of malrotation with volvulus in early neonatal period. The patient can also present with features of acute abdomen such as intermittent abdominal pain, distension, diarrhoea or constipation, haematochezia, and very rarely shock. Plain abdominal X-rays can depict features of duodenal obstruction. Upper gastrointestinal contrast study can describe the classic spiral corkscrew feature of the involved intestine [4]. It should be performed by a paediatric radiologist because of inherent chances of aspiration in a vomiting child which in turn can prove fatal. The ultrasound and computed tomography may provide a great deal of help to locate the abnormal position of SMA. Ultrasound with colour Doppler should be sufficient to show the whirlpool sign, first described in 1992 [5]. CT should be performed in complicated cases and should not delay the surgery which is the mainstay of management to prevent or minimize the complications. Urgent laparotomy is indicated in case of peritonitis. Broad spectrum antibiotics are mandatory when necrosis is suspected. The mortality rate ranges from 3-15% in spite of aggressive surgical management [6]. The prognosis of midgut volvulus is directly related to the stage of presentation, degree of bowel ischaemia, early diagnosis and timely surgical management.

**Differential Diagnosis List:** Whirlpool sign in midgut volvulus, Annular pancreas, Duodenal atresia

**Final Diagnosis:** Whirlpool sign in midgut volvulus

**References:**

Figure 1

a

**Description:** Complex mass in the epigastrium with the typical whirlpool appearance. **Origin:** Althamer E, King Saud Medical City, Riyadh, Kingdom of Saudi Arabia

b

**Description:** Vascular origin of the mass. **Origin:** Althamer E, King Saud Medical City, Riyadh, Kingdom of Saudi Arabia
Description: Superior mesenteric vein forming a twist around the superior mesenteric artery. Origin: Althamer E, King Saud Medical City, Riyadh, Kingdom of Saudi Arabia
Description: Gastroduodenal obstruction with paucity of distal gases. Origin: Althamer E, Paediatric Radiology, King Saud Medical City, Riyadh, Kingdom of Saudi Arabia
Description: Ladd band - a cause of malrotation in this patient. Origin: Jawad A, Paediatric Surgery, King Saud Medical City, Riyadh, Kingdom of Saudi Arabia