

Apple-Peel Syndrome" A Case Of Malrotation With Atresia Of Proximal Small Bowel

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INTRODUCTION:

Malrotation of gut is a serious congenital abnormality of GI tract as it predisposes to duodenal obstruction and midgut volvulus which can lead to ischaemic necrosis of small bowel. Underdiagnosis of midgut volvulus carries a high mortality rate. During fetal life the gut begins as a straight midline tube which elongates and rotates around SMA of 270 degrees in anticlockwise direction to form the entire adult bowel. Malrotation is a generic term used to describe any variation in the position of intestines and is in itself not symptomatic. Most patients (70%) have isolated malrotation of the bowel but there is an increased incidence of associated duodenal stenosis and atresia [1]. We present a case of malrotation with atresia of jejunum and ileum.

CASE REPORT:

A full term normal institutional delivery newborn baby presented with complaints of excessive regurgitation and inability to pass meconium in first 48 hrs. Mother was an unbooked case and had no antenatal USG done. The haematological and biochemical parameters were normal. X-ray of abdomen revealed scanty air in the lower abdomen. Contrast study was done by administering diluted non ionic contrast perorally through nasogastric tube. The stomach was normal in appearance. The duodenojejunal junction was to the right of vertebrae (Fig 1). The proximal jejunum was dilated and showed hold up of contrast. No contrast passed into distal jejunum and ileum. Per-rectal administration of contrast showed irregularly contracted rectum visualized in its proximal 2cm only. USG and colour Doppler showed normal SMA and SMV relationship. A diagnosis of malrotation with atresia of small bowel involving distal jejunum and ileum was offered. The patient was operated at a tertiary care centre and there was atresia of distal jejunum and complete ileum. Associated malrotation was seen.



Pic 1. Contrast study showing displaced duodeno-jejunal junction to the right of midline and absence of passage of contrast into the distal jejunum and ileum.

DISCUSSION:

Jejunal and ileal atresias have a common etiology and are due to an intrauterine vascular accident. The vascular insult may be primary or secondary event. Atresias of jejunum and ileum are more common than stenosis.

The term "apple peel atresia" describes a particular type of intestinal atresia characterized by an occlusion of the jejunum, resorption of the mesentery, and wrapping of a

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large part of the small intestine in a spiral around a thin vascular stalk consisting of the left branch of the ileocolic artery [2]. This condition accounts for approximately 5% of all intestinal atresia [3]. It is often familial, and an autosomal recessive mode of inheritance has been suggested. Associated anomalies including ocular abnormalities and microcephaly are rather infrequent and occur in 15% of cases of apple peel intestinal atresia [4]. Our case possibly fits into this description. Occasional case report of this entity with cystic fibrosis has been described.

A second more complex type of intestinal atresia is the syndrome of multiple intestinal atresia with intraluminal calcifications. Both these syndromes are thought to have an autosomal recessive association. Most of the newborn babies present with bilious vomiting and abdominal distension [5].

On plain X-ray the gas shadows can be seen upto the level of atresia with proximal bowel dilatation. With stenosis rather than atresia bubbles of distal gas are present. There may be fine intraluminal calcification seen associated with distal atresia. On contrast study the exact location of atresia can be delineated. Sometimes contrast enemas are requested to rule out more distal atresias.

It must however be kept in mind that in a child with malrotation with bilious vomiting and obvious signs of peritonism the radiological investigation should stop at the level of plain X-ray and surgery is indicated as soon as possible. In all other cases contrast studies are indicated.

Malrotation on the other hand has no specific plain radiographic findings even when associated with volvulus. The intestines in malrotation are often malfixed. The purpose of upper GI study in such cases to locate

duodeno-jejunal junction. On supine radiograph the normal duodeno jejunal junction lies to the left of the left sided pedicle at the height of duodenal bulb. When malrotation is present it is normally displaced to the right side. However it should also be kept in mind the normal duodeno jejunal junction may be mobile esp in babies less than age of 4 yrs. It can be displaced temporarily by a distended colon or stomach, an enlarged spleen, an indwelling nasogastric tube or manual palpation [6]. On a lateral view the junction of second and third parts of the duodenum is normally retroperitoneal but it turns sharply anterior in malrotation. The more distal jejunal loops lie to the right of midline. The caecal pole may lie more higher and to the left than normal in patients with malrotation. A barium meal is preferred study compared to that of the barium enema as the position of ileo caecal junction may be normal in a large number of patients of malrotation.

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