Isolated microgastria

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Introduction

Microgastria (MG) is a rare congenital anomaly[1]. Associated anomalies in the form of VACTERL are common[1].

Case report

A two year old boy presented with repeated episodes of non projectile vomiting since birth. For the past six months the severity of vomiting had increased. Serial chest x-rays showed an increasing right paracardiac shadow which was treated as pneumonic consolidation by the referring doctor (Figure 1).

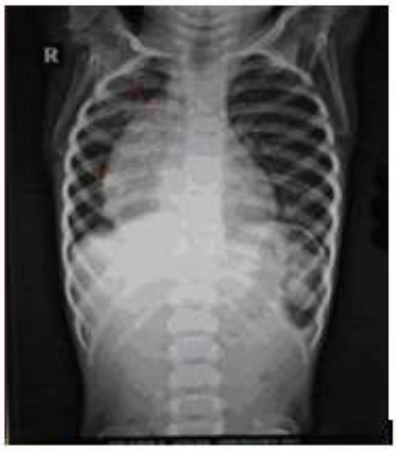

Figure 1: *Right paracardiac shadow*

On examination, pallor was present with weight and height below the 3rd percentiles. High resolution computed tomography showed an oesophagus dilated up to the gastro-oesophageal junction. The differential diagnoses were a) congenital oesophageal stenosis, b) reflux stricture with dilated oesophagus and c) achalasia cardia. Barium swallow showed a hugely dilated, tortuous oesophagus with retrograde peristalsis and delayed minimal emptying (Figure 2).

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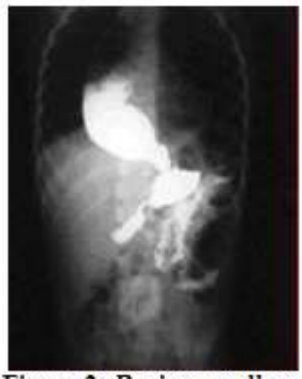

Figure 2: *Barium swallow*

Oesophageal manometry showed minimal to absent peristalsis with increase in lower oesophageal sphincter pressure. The general condition of the child was improved by inserting a feeding nasogastric tube under fluoroscopic guidance and administering small, frequent high protein feeds through it.

On laparotomy the findings were a hugely dilated, tortuous oesophagus with a stricture in the lower end and small tubular stomach in the midline (Figure 3).

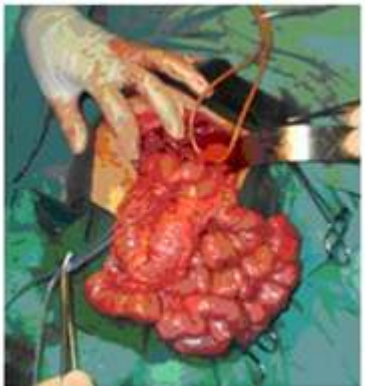

Figure 3: ***Laparotomy findings***

There was no malrotation and the spleen was normal. Gastric exclusion was done with Roux en Y end to end oesophago-jejunostomy. The postoperative period was uneventful. Feeds were started on the fifth postoperative day and gradually increased. He was discharged on the tenth day and was advised small frequent feeds. On follow-up, the child is tolerating feeds well and has gained weight over 1 year.

Discussion

Normally stomach undergoes a 90 degree clockwise rotation in the 5th week of development[2]. This occurs along with the migration of the pancreatic buds and differentiation of dorsal mesogastrium into splenic anlage. Arrest in early development of the foregut results in MG. As the stomach and spleen are both developed from dorsal mesogastrium, associated splenic anomalies are not surprising[3]. The association of limb, cardiac, tracheoesophageal, vertebral and renal anomalies has been attributed to impairment of early mesodermal development[4].

Prenatally MG mimics oesophageal atresia. Failure of visualization of fetal stomach early in second trimester suggests the possibility of congenital MG[5]. Clinically MG presents as feeding intolerance with or without vomiting and failure to thrive.

Caffey proposed that the oesophagus dilates to take the storage function of the inadequate stomach[6,7]. It is necessary to investigate the child for associated VACTERL anomalies.

The treatment of MG is individualized. A short period of conservative treatment is warranted in less severe forms of MG with diet modification i.e. frequent and small size meals with increasing quantity to dilate stomach[8,9]. However, if the stomach fails to enlarge with this treatment surgical intervention is necessary.

In patients with severe feeding intolerance with gross failure to thrive, as in this case, gastric exclusion with oesophago-jejunal anastomosis has produced satisfactory results[10,11]. Hunt and Lawrence originally described creation of a food reservoir for carcinoma stomach by creating a food pouch (Hunt-Lawrence pouch) from a segment of jejunum[8,9,12]. In 1980 Neifel et al used double lumen jejunal pouch in congenital MG. In this procedure Roux en Y jejunal loop is formed to prevent alkaline reflux and a food pouch is made by anastomosing in a side to side fashion. It also allows drainage of duodenal contents. This treatment provides an adequate pouch for food storage and decreases incidence of dumping, lessens requirement for frequent feeding and facilitates eating a balanced diet.

In conclusion MG is an extremely rare condition. When diagnosed, it needs workup for VACTERL association. Management options range from initial treatment with increasing volumes of feed to dilate the stomach, to surgical correction by either Roux en Y or double lumen pouch.

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