Case Reports

CONGENITAL TRIPLE ATRESIA OF THE ESOPHAGUS, DUODENUM AND RECTUM - A DIAGNOSTIC DILEMMA

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Introduction

The early disturbance of organogenesis that results in esophageal atresia (EA) also affects other systems. The VACTERL association is a well known combination of defects for esophageal atresia and ano-rectal malformation. The associated anomalies often significantly alter treatment and affect the survival. The association of esophageal atresia with tracheo-esophageal fistula and anorectal malformation is common and is seen in 10% of cases while its association with duodenal atresia is not so common. Here is a case of triple atresia involving the esophagus, duodenum and ano-rectum.

Case Report

A 24 hour old neonate born after 36 weeks gestation, was admitted with complaints of regurgitation of all the feeds. Examination revealed a 2.4 kg baby with scaphoid, soft abdomen and absent anal opening. A number 10 red rubber catheter passed into the esophagus, met an obstruction about 10 cms from the hips. A x-ray of the chest and abdomen taken with red rubber catheter in situ showed a blind upper esophageal pouch. The same radiograph also revealed distention of the stomach and first portion of the duodenum with air giving the characteristic appearance of "double-bubble". No intestinal air was seen beyond the duodenum (Fig 1). A diagnosis of triple atresia (esophageal, duodenal and rectal) was made.

A right postero-lateral thoracotomy was carried out and after ligation of fistula, a primary anastomosis between the proximal and distal ends of esophagus was performed. At the same time duodenostomy, gastrotomy with a transanastomotic feeding tube and left sigmoid colostomy were also accomplished. The patient withstood the surgery well.

The post-operative course was remarkably smooth for 4 days. The gastrotomy and thoracotomy tubes drained well. However, on the 5th post operative day saliva was noticed in the drainage tube, this gradually increased in quantity. Due to anastomotic leak the condition started deteriorating, which was managed by repeated throat suction, continuous gastrotomy tube decompression, parenteral nutrition and broad-spectrum antibiotics. The patient's course, however was steadily downhill and he expired on 8th post

operative day. The child died due to pulmonary complications and sepsis as a result of anastomotic leakage.

Discussion

Tracheo-esophageal obstruction is known to exist in about 10% of cases of ano-rectal malformations, while the reported incidence of duodenal atresia along with ano-rectal malformations is 1% to 2% [1]. The overall association of esophageal atresia gastrointestinal anomalies occur in 24% of cases [2] and these include anorectal atresia, duodenal atresia, ileal atresia, malrotation, annular pancreas and pyloric stenosis. Ano-rectal malformation is the commonest associated anomaly.

In the presence of triple atresia the diagnosis is du-

Fig. 1 : Antero-posterior and lateral, chest and abdominal radiograph demonstrating the blind upper esophageal pouch of esophageal atresia with catheter in situ (shown with arrow in lateral view clearly). The characteristic double bubble appearance of stomach and proximal duodenum is also seen due to duodenal atresia. No air is visualized in rest of the abdomen.

bious because apart from obvious ano-rectal anomalies, the other associated anomalies of gastrointestinal tract may be unseen. The presence of duodenal atresia obviates possibility of using the inverted film in assessing the level of the imperforated anus, because gas does not reach the rectum. In our case, it was high ano-rectal malformation with recto-urethral fistula with absent anal opening, so along with esophageal atresia the baby was diagnosed clinically having ARM. But, in cases of rectal atresia with normal anal opening, the situation becomes more complex. Irving suggested that the use of ultrasonography makes it easier to look for a duodenal atresia in the case of type II atresia without tracheo-esophageal fistula where the radiological examination alone will not show the typical lesion [3].

Spitz in a review of 18 patients of esophageal atresia and duodenal atresia emphasized that these babies are at high risk with mortality rates ranging from 67% to 94% [4]. Jackson and colleagues infer that the majority of these deaths are caused by failure to recognize the second abnormality pre-operatively [5]. In the present case, the x-ray of chest and abdomen was taken with red rubber catheter in situ to confirm the diagnosis of esophageal atresia and the same film revealed the typical double bubble appearance of gas shadow in the upper abdomen.

The management of triple atresia is challenging. These patients are usually managed by staged procedures but recently, a case of VACTERL syndrome with antenatally diagnosed duodenal atresia was operated successfully in one stage by radical surgery for esophageal atresia with tracheo-esophageal fistula and duodenal atresia [6]. The overall prognosis of VACTERL syndrome with duodenal atresia is very poor.

References

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