

Congenital esophageal stenosis and antral web

A new association and management challenge

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ABSTRACT

يعد تضيق المريء الخلقي من الأمراض النادرة التي قد ترتبط بغيرها من الأمراض الشاذة، كما يعد الوتر الغاري من الأمراض الخلقية النادرة أيضاً، ولذلك فإن حدوث المرضين في آن واحد من الأمور النادرة جداً. نستعرض في هذا البحث حالة طفل مُصاب بهذا التلازم النادر وقد تم تشخيص حالته مبدئياً على أنها حالة عدم تكون جزء من المريء (رتق المريء) مع وجود ناسور بين القصبة الهوائية والمريء. ورغم التدخل الجراحي الناجح إلا أن الطفل مر بمراحل صعبة جداً وكان هناك مشقة في معرفة سبب عدم تحسن حالته حتى تم إثبات إصابته بكل من تضيق المريء الخلقي والوتر الغاري. ويهدف هذا البحث إلى إلقاء الضوء على الصعوبة التي تواجه الأطباء في تشخيص وعلاج هذه الحالة النادرة.

Congenital esophageal stenosis is a rare condition that may be associated with other anomalies. Antral web is also a rare congenital condition. Association of both conditions is extremely rare. We present this rare association in preterm baby in whom initially misdiagnosis of esophageal atresia and trachea-esophageal fistula then followed by a dilemma in differentials diagnosis to explain the uneventful postoperative recovery. The aim of this report is to highlight the diagnostic and therapeutic difficulty of this rare condition.

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aim of this case report is to highlight the diagnostic and therapeutic difficulty of these conditions.

Case Report. A preterm baby girl, second twin, was born at the 31st week of gestation and weighing 1.14 kilograms. She developed respiratory distress after birth. Excessive frothing was noticed and a 10 French size catheter was passed, but obstructed at 12 centimeters from the upper lip. Radiograph film shows dilated proximal esophagus and gaseous abdomen. The case was diagnosed as EA with TEF and underwent thoracoscopic repair. Intraoperatively, there was no fistula; instead, an esophageal stenosis was noted. The stenotic area was located with the help of orogastric tube. Resection and primary anastomosis were carried out. On the sixth postoperative day, an esophagogram showed no leak and good progression of the contrast through the anastomotic site. The pathology report was consistent with pin hole membranous esophageal stenosis (Figure 1). When feeding started, the baby did not tolerate and developed severe gastroesophageal reflux, choking, and recurrent aspiration. Maximum medical treatment failed to control the reflux.

Abdominal radiograph showed a distended gas-filled stomach (Figure 2). Ultrasound showed a normal pylorus. We suspect a congenital gastric outlet obstruction for which upper gastro-intestinal study was performed, but failed twice due to life threatening aspiration, and with limited evaluation show dilated stomach and severe gastroesophageal reflux disease (GERD), but the contrast material passed through the antrum and pylorus to the duodenum, and there was no evidence of malrotation. Laparotomy was performed with the intention to look for a surgical cause of obstruction and for antireflux procedure. Intraoperatively, there was an antral mucosal diaphragm with a central fenestration (Figure 3); the valve was excised. Postoperatively, the feeding was successfully tolerated gradually until reaching full oral feeding within one month. She was followed up at an outpatient clinic with no complaints and progressive weight gaining.

Congenital esophageal stenosis (CES) is associated with other congenital anomalies in up to 33% of cases and the esophageal atresia (EA) and tracheoesophageal fistula (TEF) is the most common association.^{1,2} The

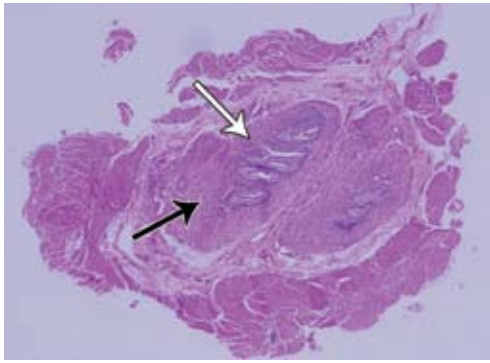


Figure 1 - Microscopic picture (hematoxylin and eosin) of the resected stenotic esophageal segment. Note the narrowed lumen (white arrow) and significant muscular hypertrophy (black arrow) of the esophageal wall.

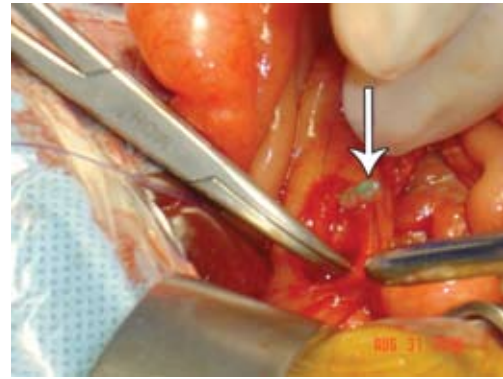


Figure 3 -Intraoperative picture of the antral web with tube passed through it (white arrow).

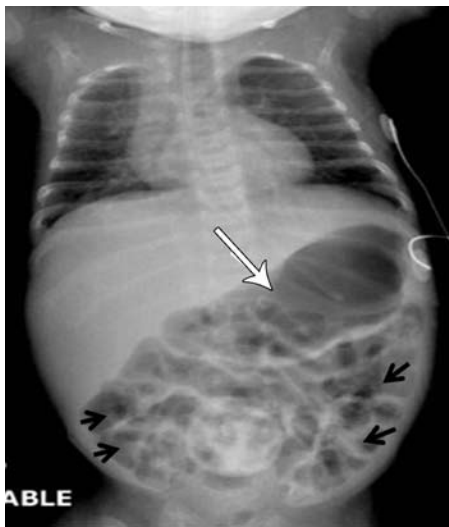


Figure 2 -Plain abdominal radiograph shows gastric distension (white arrow) with no evidence of bowel obstruction (black arrows).

Discussion. Although CES is an infrequent entity, but many cases have been reported during the last years. Its incidence falls between 1 per 25,000 and 1 per 50,000 live births and the incidence is higher in Japan although the reason for this is unknown.¹⁻³ Congenital esophageal stenosis defined as an intrinsic stenosis caused by a congenital malformation of esophageal wall that is not necessarily symptomatic at birth. They classified the etiology as: 1) tracheobronchial rests (TBR); 2) a membranous diaphragm (MD); and 3) segmental hypertrophy of muscularis and diffuse fibrosis of submucosa (FMS); the stenosis due to TBR is the most common, while the MD is the least common variety.⁴ Congenital esophageal stenosis is associated with other congenital malformations in approximately 17-33% of cases; EA and TEF being the most prevalent. Other malformations include cardiac anomalies, congenital

microgastria, congenital duodenal obstruction (atresia, web, annular pancreas and duplication), intestinal malrotation, intestinal atresia, anorectal malformations, diaphragmatic hernia, asplenia, celiac disease, vesicoureteral reflux, microphthalmos with iris coloboma, Goldenhar syndrome, Down's syndrome and other chromosomal anomalies.¹⁻⁷ We add antral web to this spectrum of anomalies. Resection of the membrane is usually the only treatment available for a complete esophageal membrane. For incomplete membranes, endoscopic incision of the membrane using cautery or a laser should be possible and dilatation of the web with various instruments has been reported.⁸ Congenital gastric outlet obstruction (GOO) in a newborn is a relatively rare condition. This may be caused by a pyloric membrane/web, pyloric atresia, or antral membrane/web.⁹ The Antral webs are thin, fenestrated diaphragms with bilateral mucosal surfaces lacking a supporting muscular layer; they are typically located 1-2 cm from the pylorus. Depending on the size of the fenestration, they have been associated with varying degrees of gastric outlet obstruction. There are several reports of older children experiencing early satiety and pain without vomiting secondary to non-obstructive antral webs.¹⁰ In contrast, our case presented early in neonatal period with obstructive antral web, which not detected even by radiological study.

We conclude that if the CES has been treated and the patient still symptomatic there should a suspicion that another distal obstructive anomaly is present.

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