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CASE REPORT

Congenital esophageal stenosis associated with tracheoesophageal fistula

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Abstract. Two patients with congenital esophageal stenosis associated with an H-type tracheoesophageal fistula in one and esophageal atresia with distal fistula in the other are presented. In both patients the correct preoperative diagnosis was made by esophagram. Satisfactory results were obtained with surgical excision. Histologic studies revealed fibromuscular stenosis and membranous esophageal mucosa, respectively.

Key words: Tracheoesophageal fistula – Esophageal atresia – Congenital esophageal estenosis

Introduction

Congenital esophageal stenosis (CES) can occur in distal esophagus associated with atresia (EA) or with an "H" (TEF) fistula. The first case of CES associated with EA was reported by Dunbar in 1958. Forty-six additional cases of CES associated with EA and/or TEF have been described since then. The diagnosis and management of CES is discused in this article.

Case reports

Case 1. A 3,000-g female neonate presented with cough and respiratory distress since the onset of feedings. At 18 h of age spontaneously resolving episodes of apnea and cyanosis were observed. Physical examination showed abundant salivation, superficial respirations and bilateral crepitations over the thorax. The abdomen was not distended.

Chest and abdominal radiographs were normal. Laboratory studies including electrolytes, blood gases, hemogram, and other evaluations were unremarkable. An esophagogram performed at 11 days of age showed an H-type tracheoesophageal fistula (TEF) at the level of the body of T1 and stenosis of the middle third of the esophagus (Fig. 1). Esophagoscopy showed the fistula opening on the anterior wall of the upper third of the esophagus with distal stenosis.

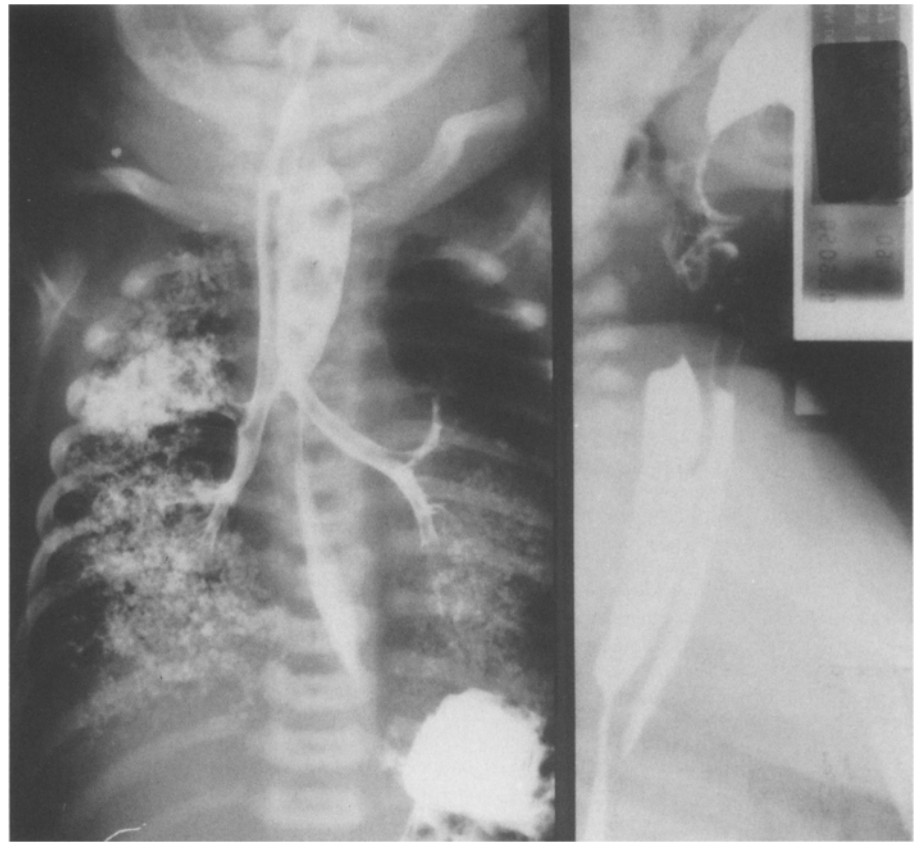

Fig. 1. H-type tracheoesophageal fistula and stenosis of middle third of esophagus

A right thoracotomy was undertaken and the TEF was excised and both ends sutured with a muscle sliver inserted between them. The esophageal stenosis was resected and an end-to-end anastomosis performed. Pneumonia and sepsis developed in the early postoperative period and were managed success-

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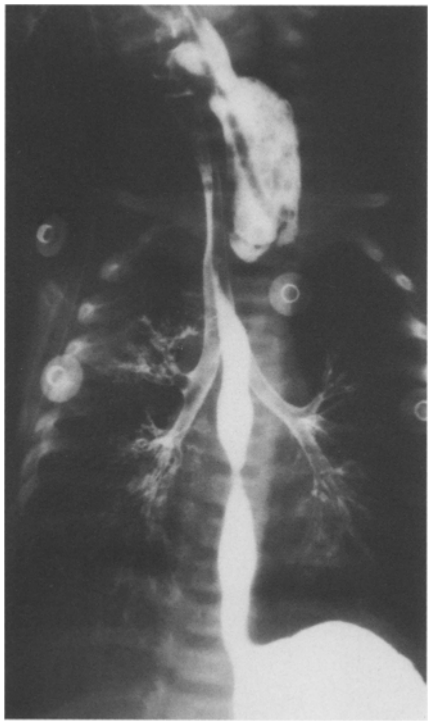

Fig. 2. Esophageal atresia with distal tracheoesophageal fistula and esophageal stenosis

fully with conservative treatment. *Oral feeding was initiated prior to a control esophagram.* Pathologic examination of the resected stenotic segment revealed fibromusclar hypertrophy.

Case 2. A full-term male 30 h of age was hospitalized for possible esophageal atresia (EA). He weighed 4,100 g; there was no history of polyhydramnios. Physical examination showed a well-appearing baby with hypersalivation and a non-distended abdomen. On a chest radiograph including the abdomen, the tip of a previously introduced catheter was seen in a dilated blind pouch at the level of the body of T2 and there was a small gastric air bubble. At the age of 1 week, a combined esophageal constrast study of the proximal and lower segments through a gastrostomy revealed EA with a distal TEF and esophageal stenosis 5 cm above the cardia (Fig. 2).

A double esophageal anastomosis was done after closure of the fistula and excision of the stenosis. The postoperative recovery was complicated by two anastomotic leaks, which were treated conservatively. Histologic examination of the stenosis revealed a membranous diaphragm.

Discussion

Approximately 4% of congenital anomalies of the esophagus are H-type TEFs. The first case of this type of anomaly was reported by Lamb in 1873 [4]. Esophageal membranes producing stenosis or obstruction are also uncommon observations. In 1791 Tenon described the first case of membranous obstruction of the upper third of the esophagus [9]. In 1929, Abel published the first case of esophagoscopic diagnosis of a congenital membrane of the lower segment of the esophagus [1]. Congenital esophageal stenosis (CES) associated with TEF has been reported by Dunbar [2], Stephens [8], Moyson [5], and Wolf et al. [10]. Eighteen of 71 cases of H-type fistula reviewed by Schneider and Becker [7] were associated with other anomalies, although none with a CES.

In 1958 Dunbar reported the first case of CES associated with EA, as was seen our second patient. Forty-six additional cases of CES associated with EA and/or TEF have been described since then [2]. In Nihoul-Fékété et al.'s recent review of 20 cases of CES, only 2 were found to be associated with EA or TEF [6].

Coughing and choking spells following feedings are characteristic clinical presentations of TEF; liquids cause more symptoms than solids. Abdominal distention is caused by air entering through the fistula during coughing or crying. These patients present with recurrent pneumonias. Esophagrams using hydrosoluble contrast material are the most valuable means of diagnosing TEFs. Pediatric fibrobronchoscopy is a helpful alternative to X-ray studies. Esophagoscopy is less accurate, but is an important aid for detecting other esophageal abnormalities [3].

In EA patients, the association with CES can easily be missed if it is not thoroughly searched for. It is important to verify patency of the distal esophagus at the time of primary anastomosis by intraoperative passage of a tube into the stomach. In our second patient, the preoperative diagnosis was suspected when too little gastric air was observed on X-ray film. The diagnosis was confirmed by contrast study.

Only 13 of 46 cases of CES associated with EA and/or TEF reported in the literature had detailed histologic findings. Of these, stenoses due to tracheobronchial rests were reported in 10 and in the remaining cases segmental fibromuscular stenosis and a membranous diaphragm were observed. A limited resection is required for stenosis caused by tracheobronchial rests. In fibromuscular stenosis and membranous diaphragms, hydrostatic balloon dilatation is sufficient.

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