CONGENITAL ESOPHAGEAL WEB : REPORT OF TWO CASES AND REVIEW OF THE LITERATURE

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SUMMARY:

Congenital Esophageal Stenosis (CES) though rare in infants and children, congental esophageal web (CEW) as a cause of CES is extremely rare. We report two children 4 and 9 years of age with CEW. They presented with progressive dysphagia after the introduction of solid food, failure to thrive and anemia. The diagnosis of esophageal web was confirmed by esophagogram and endoscopy. They were treated with dilatation and dysphagia improved markedly. Follow-up esophagogram showed improvement in the caliber of the stenosed area. A brief review of the literature and different therapeutic approach for CEW is discussed.

INTRODUCTION:

True congenital stenosis of the esophagus is a rare condition, which has been historically confused with esophageal stricture secondary to inflammation.[1] It is defined as intrinsic stenosis of the esophagus, present at birth, caused by congenital malformation of the esophageal wall architecture.[1] We report two children who had upper esophageal web and review the literature relevant to this entity.

CASE 1:

A 9-year-old Saudi boy referred to our hospital with history of progressive dysphagia following the introduction of solid food at age of 6 months. The patient had a history of chocking and vomiting soon after eating solid foods. Fluids and semi-solid diet was however, tolerated well. There was no history of corrosive ingestion or trauma. Clinical examination showed a pale, small for age child with no dysmorphic features, his weight and height below 5th percentile. Systemic examination was unremarkable. Laboratory investigations revealed Hb 5 gm/dl, urea and electrolytes were within normal range. Esophagogram showed evidence of smooth, thin, shelf-like indentation in the proximal esophagus at C_{5-6} level anteriorly. This was in keeping with the diagnosis of esophageal web (Fig 1A). Examination of esophagus under general anaesthesia using flexible esophagogastroscope revealed a thin web at post cricoid region at a distance of 15 cm from the incisor teeth. Esophageal dilatation was performed by savory dilators. The dilatation went up to 33F easily. Post dilatation course was uneventful and dysphagia improved markedly. In the 18 months follow up period the patient could swallow solid foods without difficulty and there were no complaints. Esophagogram done one month post dilatation showed less prominent indentation and the caliber of the stenosed area was wider (Fig 1B).

CASE 2:

A 4-year-old Saudi boy referred from another hospital with progressive dysphagia since early infancy. The main complaint was difficulty in swallowing solid food. There was no history of corrosive ingestion or trauma. Esophagoscopy and dilatation was attempted in referring hospital. Physical examination revealed pale, small for age child with weight and height below the 5th percentile. Systemic examination was unremarkable. Laboratory investigations showed Hb of 8 g/dl, urea, creatinine, liver function tests were within normal limit. Esophagogram revealed an anterior shelf-like indentation in the proximal esophagus (hypopharynx region) keeping with the diagnosis esophageal web (Fig 2A). Rigid esophagoscope was introduced under G.A., and a thick web in the anterior wall of the esophagus below the level of cricopharyngeal muscle was detected. Esophageal dilatation was performed using Savory dilators. Dilation went up to 45F easily. The patient made an uneventful recovery, and there were no complaints in the follow-up of 18 months period. The patient's dysphagia improved and he could tolerate solid diet comfortably. One month post dilatation esophagogram confirmed a significant dilation of the narrow segment (Fib. 2B).

DISCUSSION:

Congenital Esophageal Stenosis (CES) is a rare condition in infants and children. The incidence of CES is approximately 1 in 25,000 to 1 in 50,000 live births, with an associated anomalies reported up to 33%.[1,2] There are three forms of CES, these are: 1) Membranous web or diaphragm, 2) Fibromuscular thickening, 3)Tracheo bronchial remnants. All forms of CES may have their origin during the period of tracheobroncial and esophageal formation and separation which occur around the 25th embryonic day.[3] Majority of patients with CES are detected at the time of introduction of solid food at the age of 4 months onwards. The symptoms include dysphagia, excess salivation, regurgitation, aspiration episodes, foreign body impaction and failure to thrive.[4] The mainstay of diagnosis are esophagogram and esophagoscopy. Investigations such as pH monitoring, manometry and endoscopic biopsy are performed to rule out conditions mimicking CES.[1]

Tenon[5] in 1791 described the first case of membranous obstruction of the upper esophagus whereas, Rossi[6] reported first case of web causing complete obstruction of the distal esophagus in 1826. The presence of tracheobronchial remnants in CES were initially reported by Frey and Duschel[7] in (1936). Spitz[8] in 1973 demonstrated a clear congenital basis of CES. As of 1995, fewer than 500 cases of CES have been reported in the world literature.[4] Congenital esophageal web or diaphragm is the rarest of the three forms of CES[4]. It has been considered to represent a missed form of esophageal atresia and may be analogous to membranes in other parts of GIT.[1] It is commonly located in the middle or lower esophagus and partially obstructing the lumen.[1] Symptoms typically begin with

introduction of solid food. Both the patients in this report presented with dysphagia since the time of introduction of solid food suggesting the condition was congenital.

Various operative and non-operative procedures have been tried for treatment of Congenital Esophageal Web, these include dilatation using bougienage or hydrostatic balloon, endoscopic excision using laser or electrocautery and resection and primary anastomosis through cervical, thoracic or laparotomy approach.[1,4,9,10,11,12,13,14,15] Successful endoscopic excision of congenital web has been reported.[10] Good long term results have been reported for dilatation and resection.[8] A single dilation was showed to have relieved symptoms in two cases in this report for a period of 18 months follow up. It is also possible that the patients may have had a permanent cure by a simple dilation procedure. This suggests that non-operative treatment in the form of dilatation can be tried first, if the outcome is not satisfactory surgery may be considered.

LEGENDS:

FIG. (1A) : Esophagogram showed evidence of smooth, thin, shelf-like indentation in the proximal esophagus at C5-6 level anteriorly.

FIG. (1B) : Esophagogram one month post dilatation revealed less prominent indentation and the caliber of stenosed area become wider.

FIG. (2A) : Esophagogram revealed anterior shelf-like indentation in the proximal esphagus.

FIG. (2B) : One month post dilation esophagogram confirmed a signficant dilation of the narrow segment.

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