

Congenital esophageal stenosis: a rare case of dysphagia

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ABSTRACT

Congenital esophageal stenosis (CES) is a rare anomaly, resulting from incomplete separation of the respiratory tract from the primitive foregut at the 25th day of life. First clinical signs are abnormalities of the swallowing mechanism caused by the intrinsic narrowing of the esophagus. Diagnosis is usually delayed, requiring an accurate history and high level of suspicion, alongside with an esophagogram. Definite diagnosis is only confirmed by histological examination. Treatment usually involves surgery, depending on the severity, location and type of stenosis. We report the case of an 18 months old toddler diagnosed with CES. The characteristic radiographic and CT features are presented as well as the histology.

CASE REPORT

CASE REPORT

An 18 months old male infant, born at term with hypospadias, was brought to our hospital with a 12 month history of postprandial vomiting of undigested food particles beginning in the weaning period, with no report of weight loss. A previous admission had taken place in other hospital, with no conclusive diagnosis. Clinical examination was unremarkable. Differential diagnosis was wide and included gastroesophageal reflux (GER), achalasia and stenosis secondary to GER, foreign body, congenital or neoplastic causes.

An esophagogram using barium as contrast media was performed, revealing an asymmetric, regular, tapered and aperistaltic narrowing of the distal esophagus, approximately 2-3 cm long. Proximal dilatation of the esophagus and tertiary contractions were found, while GER was not seen (Fig. 1 and Fig. 2). Esophagoscopy showed a circumferential, slightly non-central narrowing, 2cm proximal to the esophagogastric junction (Fig. 3). No signs of esophagitis were found nor was biopsy performed. Manometry pointed to a distal tumor versus

a primary phase of achalasia. Given the worsening of the clinical condition, a feeding tube was placed.

Oral and intravenous contrast enhanced computed tomography (CT) of the chest was done for better characterization of the lesion and pre-surgery assessment, using a GE Light Speed Plus 4 slice scanner. The scan revealed a diffuse, circumferential, asymmetric and discretely lateralized to the left, non enhanced wall thickening of the distal esophagus (Fig. 4 and Fig. 5). The lesion showed a maximum thickness of 10mm, and 2.5cm of longitudinal length, being localized 2.5cm distal to the carina while sparing the last 2 cm of the esophagus. No cleavage plane with the descendent aorta or left atrium was found, neither were hilar or mediastinal lymph nodes. Excision of a 5cm segment by thoracotomy with an end-to-end esophageal anastomosis was performed under a probable diagnosis of congenital esophageal stenosis (CES) versus neoplastic stenosis. Histology confirmed CES due to tracheobronchial remnants (TBR) (Fig. 6). Post-operative recovery was uneventful, with steady weight gain and normal development at follow-up.

DISCUSSION

CES is a rare anomaly, with an incidence estimated at 1 per 25,000 to 50,000 live births, with a slightly male predominance [1]. It is believed that it results from an intrauterine stress or anoxia at the 25th day of life, leading to incomplete separation of the respiratory tract from the primitive foregut [1]. An association with other anomalies, particularly esophageal atresia, is reported in 17-33% of the cases [1,2]. Three anatomic types of CES are distinguished: esophageal membranes (EM) or web, fibromuscular stenosis (FMS) and TBR. The last is the most common form, involving preferentially the lower third of the esophagus, while the other two are more commonly seen in the middle third [3]. The symptoms vary depending on the location and severity of the stenosis. Generally, high lesions present with respiratory symptoms, while low lesions present with vomiting. Usually symptoms start around the weaning period with dysphagia to solids. However some patients may become used to live with minor degrees of stenotic lesions, eventually seeking medical attention only as young adults due to food impaction, despite a life-long history of dysphagia [4]. A high index of suspicion, an accurate history and an esophagogram are cardinal for the diagnosis. Still, diagnosis is only reached postoperatively in most cases [5].

The main clinical differential diagnosis include stenosis secondary to reflux esophagitis, achalasia, extrinsic compression by a vascular ring, foreign body and neoplasm [1,2]. Esophagogram is the main diagnostic tool in CES, generally revealing a concentric, aperistaltic, sometimes asymmetric narrowing of the upper, mid, or distal (frequently within 3.5cm of the gastric cardia) esophagus, with variable length (about 1-2cm), smooth contours and tapered borders. It is usually associated with proximal dilatation and GER can also be found [1,4,5,6]. Normal initial radiographic findings do not rule out CES [5]. In the esophagogram, patients with reflux esophagitis stenosis generally have severe GER, showing areas of lack of distensibility or transverse and converging folds, possibly associated to other inflammatory findings such as thickened and irregular folds. Stricture caused by achalasia occurs rarely in infants and tends to involve the esophagogastric junction, showing a long, tapered, narrow segment resembling a "bird's beak", with proximal dilatation. Peristalsis is disorganized, although it can be entirely absent. The vascular ring due to double aortic arch occurs as bilateral and posterior compressions of the esophagus, which remain constant regardless of peristalsis. The right indentation is usually slightly higher than the left, and the posterior compression is usually rather wide and courses in a downward direction as it goes from right to left. On the other hand esophageal neoplasms of the thoracic esophagus, also very rare in infants, may involve the entire circumference of the esophagus, producing an abrupt stricture with irregular margins [7].

Endoscopy identifies stenosis, rules out esophagitis and foreign bodies and allows biopsy, when possible [3]. pH monitoring and manometry may also be useful for diagnosis [5]. More recently, endoscopic ultrasonography has also been advocated as a helpful tool for CES diagnosis and pre-treatment assessment, since it may distinguish TBR with cartilage from FMS [1,8,9]. For greater diagnosis accuracy and

pre-surgery study, magnetic resonance imaging (MRI) can also be performed, being preferred to CT scan given the young age group of the majority of the patients. When MRI is not available, CT scan with oral contrast is also an option. On MRI, congenital esophageal stenosis due to TBR will appear as an asymmetric wall thickening in the upper- mid- or distal esophagus, possibly with some hypointense areas on the T2-weighted sequences corresponding to the cartilage islands. On CT scan, wall thickening might also be depicted. However, the definitive diagnosis is always histological [1,3,8].

The appropriate choice of treatment depends on the location, severity and type of stenosis. CES caused by TBR tends to require surgical correction, whereas CES caused by FMS is usually treated only by bougienage or dilatation [9]. The EM type is typically managed by endoscopic dilation or excision [10]. When indicated, excision of the affected segment by laparotomy or thoracotomy with an end-to-end esophageal anastomosis is commonly performed, though circular myectomy and laparoscopic esophageal stricturoplasty has also been reported [6,11]. Fundoplication can also be added to the surgical procedure [6].

TEACHING POINT

Congenital esophageal stenosis is a rare but clinically important anomaly which commonly manifests as swallowing abnormalities around the weaning period, usually requiring surgical treatment. Clinical history alongside with esophagogram can point to the diagnosis, which can only be confirmed by histopathology.

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FIGURES

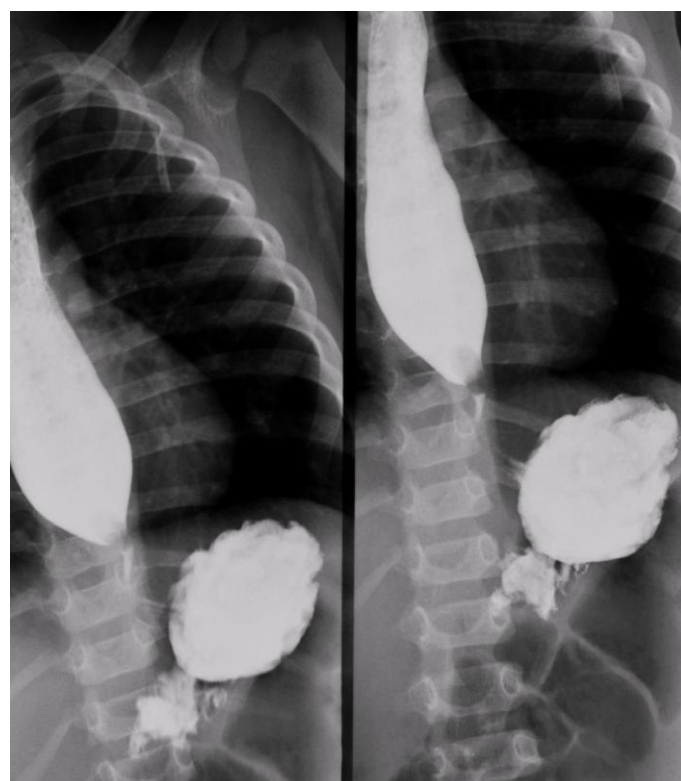


Figure 1: 18 month old male with congenital esophageal stenosis. Esophagogram using barium as contrast media with both images in AP projection. The exam revealed an asymmetric, regular and tapered narrowing of a short segment of the distal esophagus. Tertiary contractions were observed along the esophagus, except in the narrowed segment, which showed no peristalsis. The proximal portion of the lower esophagus was greatly dilated.

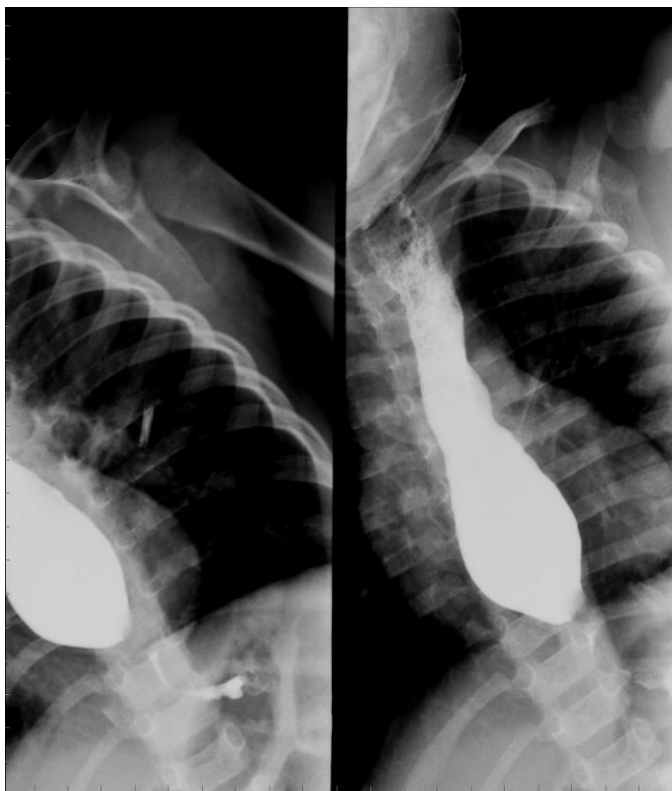


Figure 2: 18 month old male with congenital esophageal stenosis. Esophagogram using barium as contrast media, shows an AP projection (left sided image) and an unsuccessful attempt to obtain a lateral projection (right sided image), due to poor collaboration of the patient. An asymmetric short narrowing of the distal esophagus is observed, as well as proximal dilatation of the esophagus. Gastroesophageal reflux was not identified.

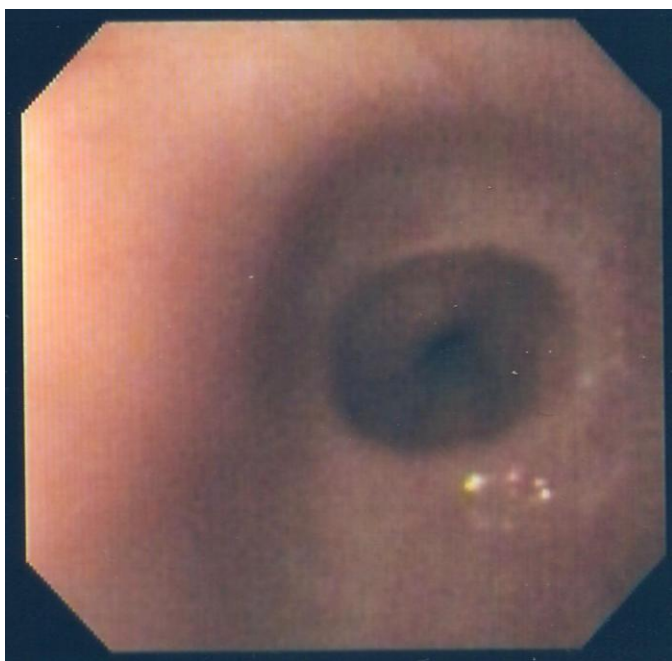


Figure 3: 18 month old male with congenital esophageal stenosis. Esophagoscopy showed a circumferential, slightly non-central narrowing at the distal esophagus, 2cm proximal to the esophagogastric junction.

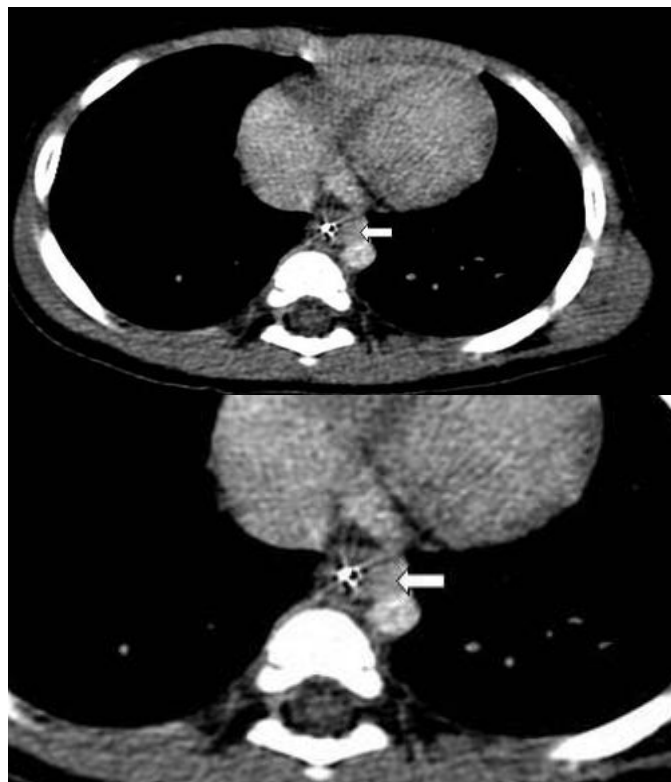


Figure 4: 18 month male with congenital esophageal stenosis. Axial contrast enhanced CT of the chest in the venous phase (lower part magnified view) demonstrates an asymmetric and circumferential, slightly lateralized to the left, wall thickening of a segment of the distal esophagus (arrow). No mediastinal lymph nodes are observed. The feeding tube is visualized in the esophageal lumen. (Protocol: GE Light Speed Plus 4 slice scanner, 9-28 mAs, 120kV, 5mm slice thickness, 22ml of non ionic contrast, Ultravist).

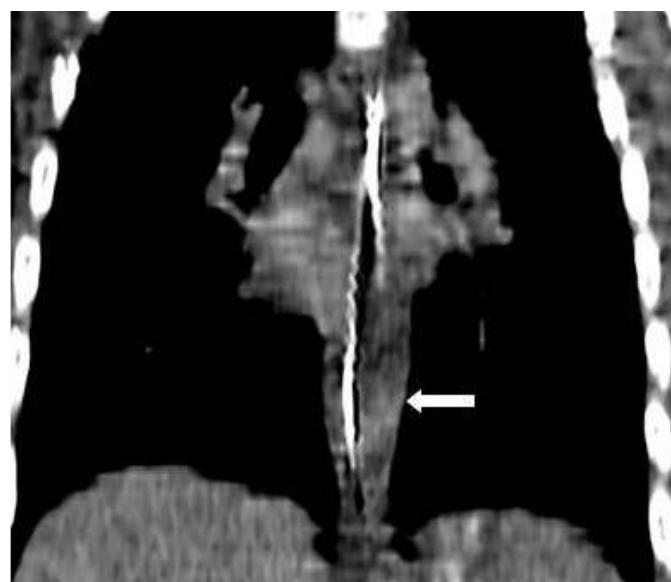


Figure 5: 18 month male with congenital esophageal stenosis. Coronal CT reconstruction of the chest shows a circumferential, diffuse, slightly asymmetric and regular thickening of the distal esophageal wall (arrow). The feeding tube is visualized in the esophageal lumen. (Protocol: GE Light Speed Plus 4 slice scanner, 9-28 mAs, 120kV, 5mm slice thickness, 22ml of non ionic contrast, Ultravist).

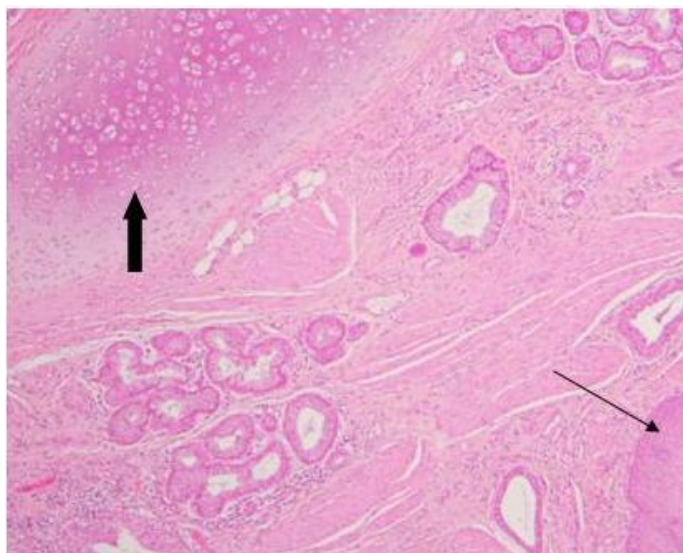


Figure 6 (left): 18 month old male with congenital esophageal stenosis. Photomicrograph (original magnification, × 100; hematoxylin-eosin [H-E] stain) of the resected specimen of the esophagus shows normal squamous lining of lumen (thin arrow) and an island of cartilage within the wall (thick arrow).

Etiology	Probably results from intrauterine stress or anoxia at the 25th day of life, leading to incomplete separation of the respiratory tract from the primitive foregut
Incidence	1 per 25,000 to 50,000 live births
Gender Ratio	Slight male predominance
Age Predilection	Most of the times, symptoms start around the weaning period (3.2-4.5 months). However, in some cases the diagnosis is only made during adulthood.
Risk Factors	Unknown.
Anatomic Types	3 types: esophageal membranes or web (EM), fibromuscular stenosis (FMS) and tracheobronchial remnants (TBR)
Treatment	EM type: endoscopic dilation or excision FMS type: bougienage or dilatation TBR type: surgical correction
Prognosis	Generally good
Findings on Imaging	<p><u>Esophagogram:</u></p> <ul style="list-style-type: none"> - normal findings - concentric, aperistaltic, sometimes asymmetric narrowing of the upper, mid, or distal (frequently within 3.5cm of the gastric cardia) esophagus, with variable length (about 1-2cm), smooth contours and tapered borders - proximal dilatation and GER can occur <p><u>Endoscopy:</u></p> <ul style="list-style-type: none"> - narrowing of the upper, mid- or distal esophagus <p><u>Endoscopic ultrasonography:</u></p> <ul style="list-style-type: none"> -asymmetric thickness of the muscular layer at the narrow portion of the upper, mid- or distal esophagus -hyperechoic lesions with acoustic shadow (cartilage islands) can be found within the thickened segment <p><u>CT:</u></p> <ul style="list-style-type: none"> - diffuse, circumferential, asymmetric and non enhanced wall thickening of the upper, mid- or distal esophagus - no lymphadenopathy <p><u>MRI:</u></p> <ul style="list-style-type: none"> - diffuse, circumferential and asymmetric wall thickening of the upper, mid- or distal esophagus with intermediate signal

Table 1: Summary table of congenital esophageal stenosis

Differential Diagnosis	Esophagogram	CT	MRI
Congenital Esophageal Stenosis	-normal findings -concentric, aperistaltic, sometimes asymmetric narrowing of the upper, mid, or distal (frequently within 3.5cm of the gastric cardia) esophagus, with variable length (about 1-2cm), smooth contours and tapered borders -proximal dilatation and GER can occur	-diffuse, circumferential, asymmetric and non enhanced wall thickening of the upper, mid- or distal esophagus -no lymph nodes	-diffuse, circumferential and asymmetric wall thickening of the upper, mid- or distal esophagus
Stenosis secondary to reflux esophagitis	-severe GER -areas of lack of distensibility or transverse and converging folds -presence of associated inflammatory findings (thickened and irregular folds)	-uniform, circumferential wall thickening; -involvement of a long esophageal segment -target sign [12]	
Achalasia	-involves the esophagogastric junction -shows a long, tapered, narrow segment resembling a “bird’s beak” -presence of proximal dilatation -peristalsis is disorganized or absent	-uniform dilatation of a long segment of the esophagus -no wall thickening -normal-appearing boundary surfaces and mediastinal fat -abruptly narrowing of the esophagus at the esophagogastric junction -no evidence of intramural or extrinsic obstructive lesion [12,13]	
Vascular ring (double aortic arch)	-bilateral and posterior compressions of the esophagus, which remain constant regardless of peristalsis -right indentation is usually slightly higher than the left -posterior compression is rather wide and courses in a downward direction as it goes from right to left	Angiography: -“four artery sign” consists of two dorsal subclavian arteries and two ventral carotid arteries evenly spaced around the trachea -left or anterior arch has a course similar to normal left aortic arch -right or posterior arch has a course to the left behind the esophagus and joins the left arch -descending aorta is usually left sided -right arch is usually larger [14]	Angiography in T1WI: -“four artery sign” -presence of a left and right aortic arches that arise from the ascending aorta -both arches give rise to a subclavian and common carotid artery -both arches join posteriorly, encircling the trachea and esophagus -descending aorta is usually left sided -right arch is usually larger and more cephalad positioned [15]
Foreign body	-helpful in demonstrating radiopaque foreign bodies (60% of the cases) -for non radiopaque foreign bodies the esophagogram shows a filling defect [16]	-variable appearance	-variable appearance

Table 2: Table differential diagnoses of congenital esophageal stenosis

Differential Diagnosis	Esophagogram	CT	MRI
Neoplasm	-involves the entire circumference of the esophagus -abrupt stricture with irregular margins	-focal, polypoid, or circumferential wall thickening -presence or absence of intense enhancement -asymmetric thickness of a short segment -presence of mass effect -loss of fat planes -presence of periesophageal lymph nodes [12]	T1WI: -intermediate signal intensity, similar to soft tissue [17] - best to detect lymph nodes [18] T2WI: -intermediate signal intensity [19] -expansion of the esophageal lumen, giving a circular appearance [19] DWI: -high signal that exceeds the signal intensity of the surrounding background (lung or air) -Lymph nodes: 1- short axis diameter >5 mm 2- long axis-to-short axis diameter ratio >2 3- signal intensity higher than that of the spinal cord [20]

Table 2: Table differential diagnoses of congenital esophageal stenosis (continued)

ABBREVIATIONS

CES = Congenital esophageal stenosis
 TBR = Tracheobronchial remnants
 GER = Gastroesophageal Reflux
 CT = Computed Tomography
 EM = Esophageal Membranes
 FMS = Fibromuscular Stenosis
 MRI = Magnetic Resonance Imaging
 T1WI= T1 weighted imaging
 T2WI= T2 weighted imaging
 DWI= diffusion weighted imaging

KEYWORDS

Congenital esophageal stenosis; tracheobronchial remnant; choristoma

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