Congenital esophageal stenosis owing to tracheobronchial remnants

Estenose congênita do esôfago por remanescentes traqueobrônquicos

Estenosis congénita del esófago por remanecientes traqueobrónquicos: relato de cuatro casos

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ABSTRACT

Objective: To emphasize the need of an accurate diagnosis of congenital esophageal stenosis due to tracheobronchial remnants, since its treatment differs from other types of congenital narrowing.

Case description: Four cases of lower congenital esophageal stenosis due to tracheobronchial remnants, whose definitive diagnosis was made by histopathology. Except for the last case, in which a concomitant anti-reflux surgery was not performed, all had a favorable outcome after resection and anastomosis of the esophagus.

Comments: The congenital esophageal stenosis is an intrinsic narrowing of the organ's wall associated with its structural malformation. The condition can be caused by tracheobronchial remnants, fibromuscular stenosis or membranous diaphragm and the first symptom is dysphagia after the introduction of solid food in the diet. The first-choice treatment to tracheobronchial remnants cases is the surgical resection and end-to-end anastomosis of the esophagus.

Key-words: choristoma; esophagus; esophageal stenosis; constriction, pathologic.

RESUMO

Objetivo: Enfatizar a necessidade de um diagnóstico preciso de estenose congênita do esôfago por remanescentes traqueobrônquicos, já que seu tratamento difere dos outros tipos de estreitamento congênito.

Descrição do caso: Quatro casos de estenose congênita do esôfago inferior causada por remanescentes traqueobrônquicos, cujo diagnóstico definitivo foi obtido por exame histopatológico. À exceção do último caso, em que não se realizou cirurgia antirrefluxo concomitante, todos apresentaram evolução satisfatória após ressecção e anastomose do esôfago.

Comentários: A estenose congênita do esôfago consiste no estreitamento intrínseco da parede do órgão associada à malformação de sua estrutura. Pode ser causada por restos traqueobrônquicos, espessamento fibromuscular ou diafragma membranoso e tem como primeira manifestação clínica disfagia após introdução de alimentos sólidos na dieta. O tratamento de escolha para os casos de remanescentes traqueobrônquicos é a ressecção do segmento estenosado com anastomose término-terminal.

Palavras-chave: coristoma; esôfago; estenose esofágica; constrição patológica.

RESUMEN

Objetivo: Enfatizar la necesidad de un diagnóstico preciso de estenosis congénita del esófago por remanecientes traqueobrónquicos, una vez que su tratamiento difiere de los otros tipos de estrechamiento congénito.

Descripción del caso: Cuatro casos de estenosis congénita del esófago inferior causada por remanecientes traqueobrónquicos, cuyo diagnóstico definitivo fue obtenido por examen histopatológico. Excepto por el último caso, en el que no se utilizó cirugía antirreflujo concomitante, todos presentaron evolución satisfactoria después de resección y anastomosis del esófago.

Comentarios: La estenosis congénita del esófago consiste en el estrechamiento intrínseco de la pared del órgano

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Conflito de interesse: nada a declarar

Recebido em: 18/7/2012
Aprovado em: 24/1/2013

asociado a la malformación de su estructura. Puede ser causada por restos traqueobrónquicos, espesamiento fibromuscular o diafragma membranoso y tiene como primera manifestación clínica disfagia después de la introducción de alimentos sólidos en la dieta. El tratamiento de elección para los casos de remanecientes traqueobrónquicos es la resección del segmento estenosado con anastomosis término-terminal.

Palabras clave: coristoma; esófago; estenosis esofágica; constricción patológica.

Introduction

The first report of congenital esophageal stenosis (CES) was attributed to Frey and Duschl, in 1936. The authors described the case of a 19-year-old girl, whose death was attributed to the diagnosis of achalasia and who was found to have cartilage in the cardia during necropsy. CES is a rare condition, associated with malformation of the esophageal structure[1], which may be concomitant with esophageal atresia. Tracheobronchial remnants (TBR) are the most frequent cause, but membranous diaphragm and fibromuscular (FMS) stenosis are other possible etiologies for this type of narrowing[2]. In the differential diagnostic procedure, the physician should consider achalasia and acquired forms of narrowing, such as those secondary to peptic esophagitis by gastroesophageal reflux and caustic injuries. Dysphagia is the initial clinical manifestation, which occurs after the introduction of solid food. An indicative history leads to investigation through esophagography and endoscopy, but the definitive diagnosis is only possible by histopathology.

Unlike other types of stenoses that respond to endoscopic dilatation and the construction of an anti-reflux valve as a treatment option, in patients with TBR the resection of the stenotic segment followed by end-to-end anastomosis is the treatment of choice, since in this type of stenosis, dilatation may result in esophageal perforation and its consequences.

In the present study, four clinical cases of CES due to TBR were reported with the aim to point out the importance of considering such pathology in the face of clinical cases of dysphagia, once an accurate diagnosis is mandatory for choosing an appropriate therapeutic approach.

Case Description

This is a retrospective study including all cases of TBR patients treated at Hospital Antônio Pedro, Lagoa Hospital, and in private practice. The cases were monitored and operated on by the same surgeon from 2001 to 2011. The inclusion criterion was a diagnosis confirmed by histopathological findings. Using a descriptive research method, the information contained in the medical records was used and compared with data obtained in the current literature. The study was approved by the Research Ethics Committee of Hospital Universitário Antônio Pedro.

The first case refers to a black male patient, admitted to the service of Pediatric Surgery at Hospital da Lagoa, at 18 months old. The parent referred vomiting after eating from 4 months old and inability to eat solid food. An esophagogastroduodenal series (EGDS) was requested, which demonstrated a ring-like stenosis over the distal third of the esophagus (Figure1). The presence of a whitish stenotic ring at 22cm from the incisors prevented the progression of the endoscopic device into the esophagus. There were no signs of changes in the integrity of the mucosa, or of gastroesophageal reflux, and the appearance was compatible with choristoma. The infant underwent exploratory laparotomy. After identification of the cartilaginous ring by palpation, the anesthesiologist was asked to pass a Foley catheter to define the lower limit of the resection. After this maneuver, it was proceeded to the resection of the esophageal stenotic segment, followed by end-to-end anastomosis and Thal fundoplication. The child evolved without postoperative complications, and the histopathology confirmed the heterotypic diagnosis.

The second case was a Caucasian female infant, less than 24 hours old, submitted to surgical repair of esophageal atresia with distal tracheoesophageal fistula in the service of Pediatric Surgery at Hospital da Lagoa. The patient evolved without postoperative complications, with introduction of oral diet on the seventh day. After completing 1 month of age, a control esophagography showed wide anastomosis and gastroesophageal reflux until the upper third of the esophagus. In this examination, the radiological report made no reference to the distal esophageal stenosis, identified in the EGDS. After clinical treatment for reflux, the infant was brought to our clinic at 7 months of age complaining of pneumonia and choking during feeding. A new contrast study showed stenosis of the lower esophagus, confirmed during endoscopy. There was an obstacle to the passage of the endoscope by a rigid circular ring, suggestive of choristoma (Figure 2). During the examination, there was no episode of gastroesophageal reflux and the mucosal integrity was intact. Exploratory laparotomy was indicated and it was proceeded

to resection of the esophageal stenotic segment, with end-to-end anastomosis and Thal fundoplication. During the surgery, the remaining cartilage was not identified and the postoperative endoscopic exam guided the limits of esophageal resection. The child recovered without complications and histopathology revealed choristoma, with identification of small plates of cartilage and mucous glands.

The third case was a black female patient, who was admitted at the service of Pediatric Surgery at Hospital Universitário Antônio Pedro at 23 months old, with history of vomiting after meals since 4 months old. Up to that moment, she had been receiving clinical treatment for gastroesophageal reflux, without responding to the therapy. Two months before the hospitalization, a contrasted study in the esophagus showed stenosis over the lower third of the esophagus, and a proximal filling defect. Then, an upper gastrointestinal endoscopy (UGIE) showed intact mucosal integrity, normal color, with crescent-shaped narrowing in the distal esophagus, and food retention. With the diagnostic hypothesis of choristoma, exploratory laparotomy was indicated. During the surgery, it was not possible to feel the cartilage, so endoscopic aid was necessary for the segmental resection of the esophagus. After anastomosis, Thal fundoplication was added to the procedure. In the histopathologic examination of the object, cartilage rings and aggregate mucous glands were found (Figure 3). During the patient's follow-up period, the esophagography and the endoscopy were normal.

The last case was a Caucasian female patient, admitted to Hospital Universitário Antônio Pedro at 4 years of age. The history made reference to regurgitation and vomiting after eating since 6 months of age, when solid food was introduced to the diet. Empirically, clinical treatment for gastroesophageal reflux disease was performed, with no improvement. She was submitted to UGIE for removal of foreign body (food remains). During the examination, the mucosa was intact and there was a crescent-shaped narrowing in the distal esophagus. The area of stenosis was then dilated, with temporary improvement. An esophagography confirmed the stenosis over the transition from the middle to the lower third of the esophagus (Figure 4). The patient underwent a right posterolateral thoracotomy, the stenotic segment was resected, and traffic was restored by end-to-end anastomosis. The esophagus was tapered, without palpable cartilage and, therefore, without definition of the resection area. Endoscopy was again required to establish these limits. The histopathologic examination of the resected esophageal segment revealed esophageal

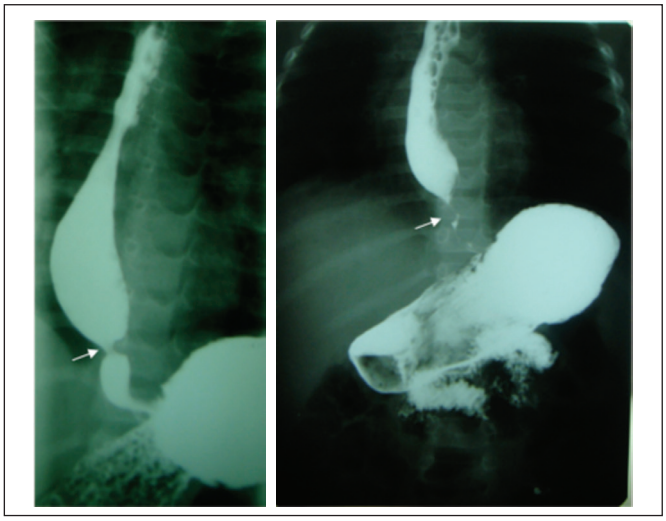

Figure 1 - Ring-like stenosis (arrow) at the distal third of the esophagus (case 1)

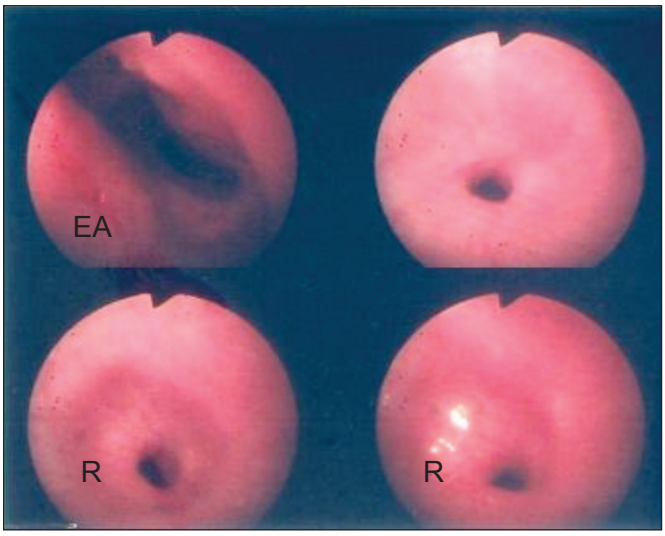

Figure 2 - Rigid circular distal ring (R), suggestive of choristoma. Broad esophageal anastomosis (EA) (case 2)

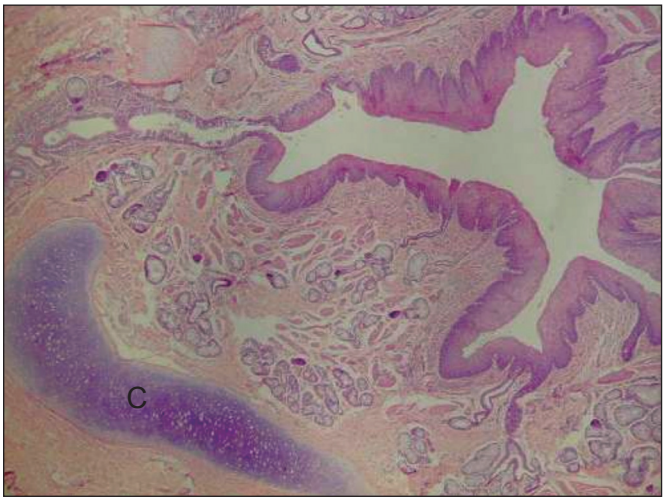

Figure 3 - Cartilage ring (C) and mucous glands over the esophageal wall (case 3)

mucous glands and respiratory epithelium. Approximately 2 months after surgery, there was recurrence of stenosis. Nissen fundoplication was indicated and the child is under a dilation program at the present moment.

Discussion

The incidence of CES ranges from 1:25,000 to 1:50,000 live births and there is a slight predominance in males[3]. Defined as an intrinsic narrowing of the esophagus, they are associated with malformation of the structure of the esophageal wall. Congenital esophageal stenoses may be caused by tracheobronchial remnants, fibromuscular stenosis, or membranous diaphragm[4].

CES due to TBR, also referred to as choristoma or heterotopy[5], is rare, but corresponds to one of the most frequent causes of lower CES, especially in Japan[6]. The presence of a cartilaginous ring in the esophageal wall is the likely result of a failure in embryonic separation of the esophagus from the respiratory tract on the 25th day of gestation. This deposit of cartilage may involve completely or incompletely the distal third of the organ, causing partial obstruction of its lumen[7]. Although sequestration of cartilage should occur in the upper esophagus, this is found in a more distal position, usually within 3cm of the cardia[7,8], due to normal growth[6]. Indeed, the tracheobronchial remnants may contain cartilage, respiratory epithelium, or seromucous glands — each alone or in combination. In cases where there is no cartilage, they may not develop stenosis[9].

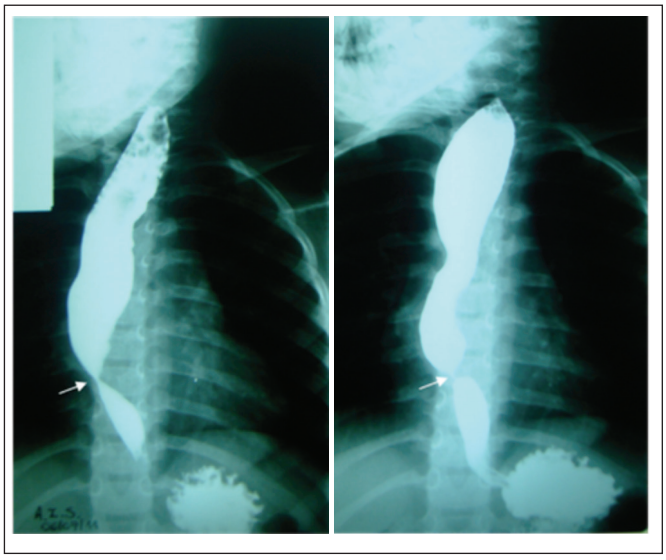

Figure 4 - Stenosis (arrow) over the lower third of the esophagus (case 4)

The association of esophageal choristoma with other congenital malformations is especially rare, being esophageal atresia with or without tracheoesophageal fistula the most frequent anomaly[3,6,9,10]. In the postoperative of atresia, special attention should be given to the control contrasted study, with may reveal this lesion early[10]. It should be considered, however, that a normal esophagography at this point does not rule out CES[9]. Other malformations may be present, such as anorectal malformations, congenital heart disease, trisomy 21, microphthalmia with coloboma of iris, atresia or duodenal duplication, malrotation, Meckel's diverticulum, celiac disease, microgastria, diaphragmatic hernia, tracheomalacia, tracheoesophageal fistula in "H," vesico-ureteral reflux, and hemangioma[3,4,6,10].

The onset of symptoms usually occurs at the time of transition from liquid to solid diet, when the patient begins to show dysphagia. Regurgitation and vomiting are the most frequent complaints[7]. Other symptoms may be: hypersalivation, stridor during feeding, development deficits, recurrent pneumonia secondary to aspiration, upper respiratory tract infections, and foreign body or food impactions[3,8,9]. The severity of symptoms appears to be proportional to the degree of involvement of the esophageal wall[7].

The diagnosis should be suspected from the correlation of the clinical, endoscopic and esophagography findings. The contrast study demonstrates segmental narrowing of the esophagus, with upstream dilation. The extent of stenosis should be measured, and extrinsic compression, foreign body, and fistula should be excluded[1]. During endoscopy, the impossibility of progression of the device and the absence of signs of esophagitis rule out other causes, such as achalasia, peptic stricture or stenosis by ingestion of caustic substances. The monitoring of pH and the esophageal manometry are useful to exclude or associate gastroesophageal reflux disease[1,2,9,11]. To distinguish FM stenosis from stenosis due to TBR an endoscopic ultrasound may be used[3,4]. In fact, the diagnosis is only definitive with the completion of the histopathology, which will identify in the resected esophageal segment, the presence of cartilage, seromucous glands, and pseudostratified ciliated columnar epithelium, alone or in combination[9]. Zhao *et al*[3] observed a delay from 2 to 2 and 1/2 years between the onset of symptoms and the diagnosis of TBR. The early diagnostic differentiation is essential, therefore, in order not to delay definitive treatment.

Although there are reports of endoscopic dilatation as treatment for CES due to TBR, its results are ineffective[3,4] and its use should be limited to cases without cartilage[9], as it can result in serious complications such as perforation

of the esophagus[1,3,10,11]. The treatment of choice should be surgery[3,4,10]. The operation consists of segmental resection of the esophagus, via abdominal or thoracic, and primary anastomosis. During the procedure, the vagus nerve must be dissected and isolated with wire to prevent its damage. The identification of the cartilage ring by palpation can be difficult, and at this moment, the use of pre-operative or endoscopic passage of a balloon catheter is most useful in defining the boundaries of resection[4,11,12]. The endoscopic ultrasound is another resource to be used in the attempt to identify the structure of cartilage during surgery[4]. In cases located in the distal esophagus and addressed via abdominal, fundoplication is associated to the procedure to prevent gastroesophageal reflux and protect the suture. Surgical treatment has low morbidity and mortality, being stenosis of anastomosis the prevailing postoperative complication, but which responds to dilation[3]. In selected cases, enucleation of the remaining cartilage, myotomy, and myectomy may be treatment options[4,12,13].

Thus, the etiologic diagnosis of a CES owing to TBR is essential to define the gold standard treatment that comprises resection of the esophageal stenosis, end-to-end anastomosis, and the construction of an anti-reflux valve. However, dilatation of the esophagus as an alternative therapy can be disastrous, causing perforation of the organ and serious consequences.

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