CLINICAL CASE

Oesophageal tracheobronchial remnants

Un cas de sténose congénitale de l’œsophage à cause de résidus trachéobronchiales

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Summary  Congenital oesophageal stenosis due to tracheobronchial remnants is a very rare condition characterized by the presence of tracheobronchial tissue in the oesophageal wall. The most common symptoms are dysphagia, regurgitation and hypersalivation. These usually appear in early infancy when solid food is introduced into the diet. Clinical diagnosis is difficult. Conventional radiology shows a dilated oesophagus without peristalsis, incomplete expansion of the gastro-oesophageal transition zone and delayed oesophageal emptying. Lesions suggesting oesophagitis may be present at endoscopy but pH-metry may be normal. The combination of stenosis with or without oesophagitis with normal pH-metry suggests that the patient’s symptoms may be due to factors other than intraluminal and that further investigation is needed. The treatment of this condition requires surgical resection of the stenosis with little morbidity or mortality. The presence of tracheobronchial remnants is confirmed upon histopathological examination of the resected bowel segment.

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Résumé  La sténose congénitale de l’œsophage en conséquence de résidus trachéobronchiaux est une anomalie rare définie par la présence de tissu d’origine trachéobronchiale dans la paroi oesophagienne. Les symptômes les plus fréquents sont la dysphagie, les régurgitations et l’hypersalivation. Ceux-ci se manifestent d’ordinaire dès l’introduction d’ingrédients solides dans l’alimentation. Le diagnostic sur base des données cliniques est difficile. La radiographie conventionnelle montre l’œsophage dilaté avec absence du péristaltisme, l’expansion incomplète de la zone de transition gastro-œsophagienne, et un ralentissement de la vidage de l’œsophage. L’endoscopie montre parfois des lésions suggestives d’œsophagite, tandis que la pH-métrie peut être normale. La combinaison d’une sténose avec ou sans œsophagite...
et une pH-métrie normale est suggestive d’une origine extra-œsophagienne des symptômes, qui devrait être explorée en détail. Le traitement spécifique de cette anomalie est la résection chirurgicale de la sténose et connaît en général peu de complications. La présence de résidus trachéobronchiaux peut être confirmée lors de l’examens histopathologique de la pièce chirurgicale.

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Introduction

Nausea, persistent vomiting, dysphagia, regurgitation, stridor during feeding and respiratory symptoms in newborns or infants require an immediate differential diagnosis. These symptoms are a major cause of psychological stress for the parents of the newborn. An infant who cannot sleep, has anorexia and weight loss must be rapidly diagnosed to relieve symptoms. Many different causes including malformations, functional disorders, neurological disturbances and even tumours can be responsible, and the diagnosis is often based on pathology. The possibility of achalasia, gastro-oesophageal reflux and congenital malformations such as oesophageal atresia, tracheo-oesophageal fistula, hiatal hernia, oesophageal duplication, congenital oesophageal stenosis due to membranous diaphragm and fibromuscular stenosis must be considered [1,2]. We report the clinical and histopathological features of a case of congenital oesophageal stenosis due to tracheobronchial remnants.

Case report

This report describes a boy born by caesarean section at 39 weeks gestational age. Body weight at birth was 2870 g and general development was normal at birth. At the age of six months, the baby was hospitalized for nausea, vomiting and stridor during feeding. Standard X-rays showed a dilated oesophagus without peristalsis, incomplete expansion of the gastro-oesophageal transition zone and delayed oesophageal emptying. pH-metry was normal and CT-scan failed to show any extrinsic compression. A tentative diagnosis of achalasia was made. At endoscopy, a 2 to 3 cm long filiform stricture of the distal oesophagus was observed. The stenosis could not be passed through with the paediatric oesophagogastroscope. Mucosal biopsies were taken and showed mild oesophagitis. The stricture was dilated twice with Savary bougies but the lesion and the patient’s symptoms persisted. A control endoscopy at the age of one showed a non-yielding supradiaphragmatic stricture with a diameter of 5.7 mm. Mucosal biopsies were again normal. Because the previous dilatation procedures had been unsuccessful the stricture was excised at thoracotomy and an end-to-end anastomosis was performed.

The surgical specimen consisted of a 1 cm long oesophageal segment with a markedly narrowed lumen and a rigid wall. Histological examination showed normal or keratinized multilayered squamous epithelium alternating with areas of thinned epithelium characterized by the presence of basal zone hyperplasia (Fig. 1). The muscularis mucosae was well-developed but showed focal fusion with the muscularis propria. The latter was interrupted in a small zone underlying an area of prominent mucosal inflammation. There was also localized fibrosis. Numerous normal submucosal glands were present. In addition, the disordered muscularis propria and adventitia contained cartilaginous plates and seromucinous glands connected by ducts with the oesophageal lumen (Fig. 2). Overall, this pattern provided a final diagnosis of congenital oesophageal stenosis due to tracheobronchial remnants. The subsequent clinical course was uneventful.

Discussion

Congenital oesophageal stenosis (CES) has been defined as an intrinsic narrowing of the oesophageal lumen caused by a malformation of the bowel wall. This condition is rare, with an estimated overall incidence of one in 25,000 to 50,000 live births [3,4]. For unknown reasons, the incidence of CES is higher in Japan [5—7]. There is no gender predisposition. The particular malformations which cause the stenosis can be divided into three categories: tracheobronchial remnants (TBR), a membranous diaphragm (MD) and fibromuscular stenosis (FMS) [3,5,8]. TBR is the most common cause, while MD is the least common variant [8,9]. TBR and FMS are frequently associated with oesophageal atresia and tracheo-oesophageal fistula [3,5,10—12]. Most cases of CES due to TBR occur in the lower third of the oesophagus, within 3 cm of the cardia, while MD and FMS are more commonly located in the middle part of the oesophagus [8,13]. Stenosis secondary to TBR often causes a high-grade obstruction, as in our case [8,14].

Figure 1 Normal multilayered squamous epithelium next to areas showing reactive changes. Haematoxylin-eosin, original magnification × 50.
Congenital oesophageal stenosis due to tracheobronchial remnants is characterized by the presence of tissue of a tracheobronchial origin in the oesophageal wall. The presence of these tissues causes the narrowing of the oesophagus [9,15]. TBR were first described more than 70 years ago by Frey and Duschl, in a 19-year-old girl who died with a diagnosis of achalasia. Microscopic foci of cartilage were found in the cardia during autopsy [16].

Tracheobronchial remnants in the oesophagus are due to an anomaly during separation of the respiratory tract anlage and the oesophagus which occurs around the 25th day of gestation. The causative factor could be intra-uterine stress or anoxia occurring in the first months of pregnancy [9,15,17,18]. The presence of TBR is often associated with disarray and thickening of the muscularis propria. It has been suggested that these changes may be due to a reduction of myenteric nitrergic neurons, but this hypothesis remains to be confirmed [5,19].

Congenital oesophageal stenosis is not necessarily symptomatic at birth [8,20]. The clinical signs of TBR usually first appear in early infancy at a mean age of three to four months, when solid food is introduced into the diet. Dysphagia, regurgitation and hypersalivation are the most common symptoms [15].

The clinical diagnosis of congenital oesophageal stenosis due to tracheobronchial remnants is difficult. A high index of suspicion, accurate anamnesis and a barium oesophagogram are the primary diagnostic tools. Although endoscopy locates the stenosis so oesophagitis can be ruled out, lesions suggesting oesophagitis can be present. Mucosal biopsies and even the surgical specimen may show clear-cut features of oesophagitis, as in our case. A normal pH-metry result rules out gastro-oesophageal reflux disease [15]. The combination of a stricture with or without oesophagitis and a normal pH-metry definitely suggest that the symptoms and lesions may be due to factors other than intraluminal causes and that further investigation is needed. Treatment of CES due to TBR requires surgical resection of the stenosis and there is little morbidity or mortality [15]. Diagnosis of TBR is made by histopathological examination of the resected segment.

References