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CASE REPORT

Tracheobronchial remnant as a cause of oesophageal stricture in children – case report and review of the literature

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ABSTRACT

Tracheobronchial remnant (TBR), one of the causes of congenital oesophageal stenosis (COS), is a rare anomaly, the treatment of which is hampered by the lack of well-established therapeutic strategies. A definite diagnosis of TBR can be made only by histopathological examination of the resected segment. A 10-month-old girl was admitted to a district hospital due to dysphagia, intolerance of solid food, and regurgitation. The patient was referred to our hospital for continuation of the treatment. She underwent a laparotomy, segmental resection of the oesophageal narrowing, end-to-end anastomosis, and anterior fundoplication. Surgical excision of the stricture is the only effective treatment of a patient with TBR.

KEY WORDS:
oesophageal stenosis, tracheobronchial remnant, congenital oesophageal stricture.

INTRODUCTION

Congenital oesophageal stenosis (COS) is a rare anomaly in children with the incidence estimated at 1 : 25,000–50,000 live births [1]. COS may have various aetiologies, and its association with the other anomalies ranges from 17% to 33% [2]. COS is recognised most often in Japan, as shown by numerous publications from this country [3]. Nihoul-Fekete classified COS in to three histologic types: tracheobronchial remnant (TBR), fibromuscular thickening (FM), and membranous web (MW) [4–6]. Preoperative diagnosis may be difficult, and there is no unified therapeutic strategy. The authors present a case of a patient with oesophageal stenosis due to TBR.

CASE REPORT

A 10-month-old girl was admitted to a district hospital due to dysphagia and regurgitation. The symptoms were observed since her birth and gradually increased in severity. With introduction of diversified food, the patient started to have difficulties swallowing solids with episodes of choking. Moreover, she could only tolerate small portions of semi-fluid food.

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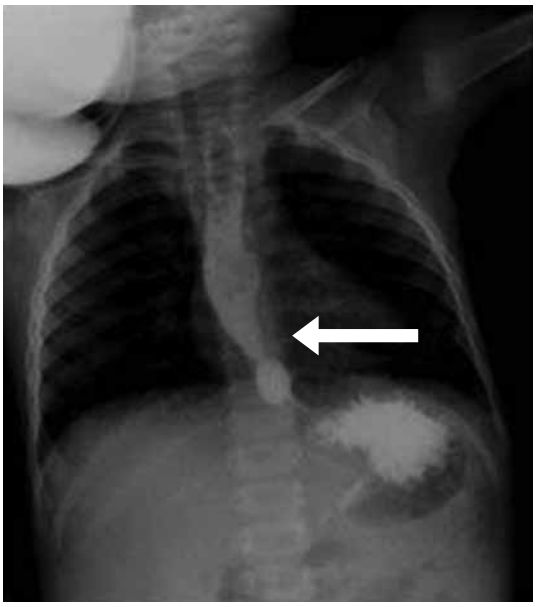

FIGURE 1. Barium oesophagogram in a patient with TBR – the arrow marks abrupt stenosis in the distal part of the oesophagus

Initially a barium oesophagography was performed. The investigation revealed a hiatal hernia 12 mm in diameter; the oesophagus above the hernia was dilatated to 14 mm with barium stasis, and the cardia was described as narrow and rigid (Fig. 1). Because the oesophageal stricture was suspected to be associated with oesophagitis, proton-pump inhibitors (PPIs) were included in the treatment. Manometric examination was impossible because the patient presented with severe gag reflex. Esophagoscopy showed normal upper 2/3 of the oesophagus, but a 1–1.5-cm-long rigid stricture was found in the distal third of the oesophagus. The diameter of the stricture was about 3 mm. What drew attention was that the endoscope 2.7 mm springed on the rigid narrowing, and thus the balloon dilatation was withdrawn.

The patient was then referred to our hospital and underwent interdisciplinary gastroenterological, radiological, and surgical consultation.

Based on the collected data, organic narrowing of the oesophagus was suspected, most likely TBR. The patient was qualified for surgical treatment. A midline laparotomy was performed. The distal, subdiaphragmatic part of the oesophagus was exposed and mobilised. The stricture was located 2 cm above the cardia. The extent of the stenotic segment was confirmed by an intraoperative esophagoscopy. Segmental resection length of about 8 mm was performed with subsequent end-to-end oesophageal anastomosis on a nasogastric tube using semi-continuous PDS 4.0 sutures. Crura of the diaphragm were approximated and sutured with insoluble sutures, and anterior fundoplication was performed. The abdominal wall was closed with peritoneal drainage installed.

The histological examination of the specimen revealed para-epidermoid epithelium, cartilage, and mucus glands of tracheal origin with nonspecific chronic inflammatory reaction (Fig. 2).

After the surgery, the patient received parenteral nutrition for seven days. The postoperative contrast study on the seventh day after surgery showed a smooth passage of barium (Fig. 3). On the 14th day the patient was discharged home eating full portions with no dysphagia.

Three weeks after the laparotomy, the patient was admitted to the hospital due to recurrence of dysphagia, regurgitations, and difficulty in swallowing saliva. Oesophagography showed a stricture in oesophageal anastomosis and insignificant sliding hiatal hernia (Fig. 3). Subsequent oesophageal endoscopy revealed a narrowing of the oesophagus, and thus a 9-mm balloon dilatation of the stenosis was performed. Additionally, a prokinetic agent was introduced into the treatment. The patient was discharged home being asymptomatic.

Esophagoscopy after one month showed no oesophageal stenosis and insignificant gastroesophageal reflux, which was treated with PPI. The patient did not require any additional balloon dilatation. Follow-up oesophagography showed smooth barium passage (Fig. 4). The follow-up after surgery is currently three years, and the

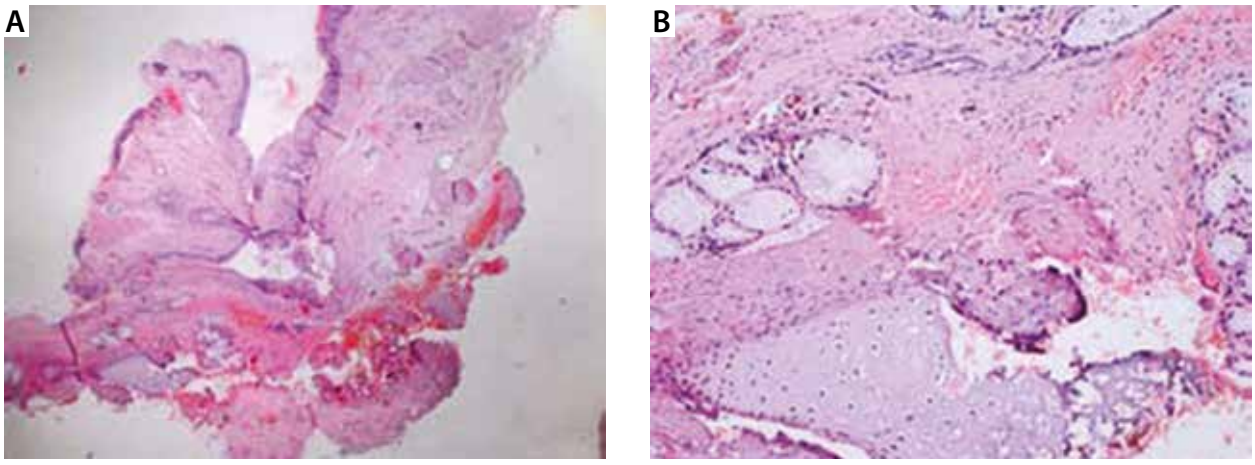

FIGURE 2. Tracheobronchial remnant – histopathological examination. Fragments of the airway wall with paraepidermoid epithelium and cartilaginous elements

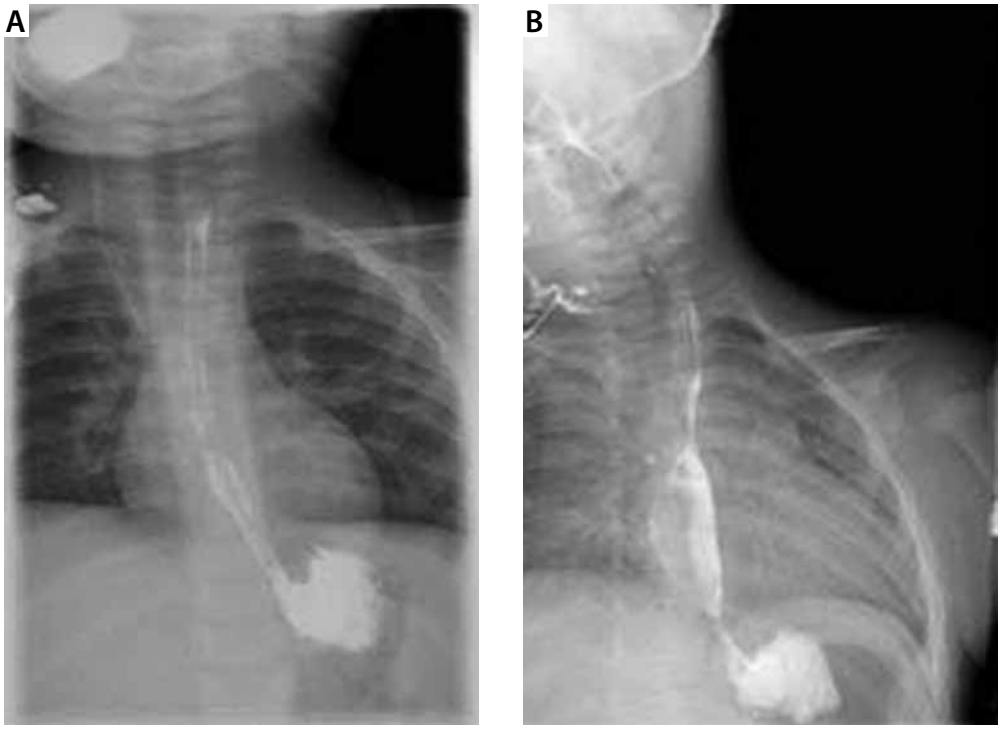

FIGURE 3. Oesophagography in a patient with TBR after oesophageal resection and partial fundoplication. **A)** one week after surgery – smooth contrast transition to the stomach, **B)** three weeks after surgery – slight narrowing in oesophageal anastomosis

patient remains asymptomatic, eats normally, and has gained weight.

DISCUSSION

Tracheobronchial remnant is a very rare congenital anomaly, and therefore it may cause diagnostic and therapeutic difficulties. It was first described by Frey and Duschel in l936 [7]. TBR is defined as abnormal presence of a ring of tracheal or bronchial cartilage in the wall of the distal oesophagus [8]. It is caused by abnormal separation in the embryonic period (around the 25th embryonic day) of the respiratory system from the foregut and can be associated with oesophageal atresia and tracheoesophageal fistula [5, 9]. The most common defect accompanying TBR is oesophageal atresia with or without fistula [9]. TBR might contain respiratory epithelium, bronchial glands, cartilage, or lymphatic tissue in the submucosa or muscularis propria and occur in the abdominal part of the oesophagus [10].

The presented case is, to our knowledge, the first described TBR case in Poland. The majority of available literature consists of case reports. The largest group of patients with TBR was described by Kim from South Korea – 29 children [11]. TBR can also be diagnosed in adult patients [12].

The diagnostic procedures are usually performed due to complaints of dysphagia, regurgitations, and difficulty in feeding or even in swallowing saliva. Dysphagia occurs usually after introduction of solid food; however, in the case of TBR it may occur in later infancy [13, 14]. The

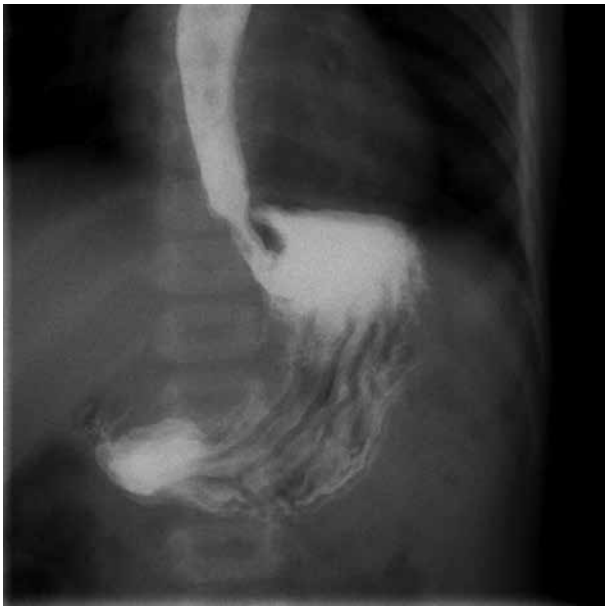

FIGURE 4. Oesophagography in a patient three years after the surgery – smooth barium passage

diagnosis of TBR is often delayed. Symptoms are similar to those occurring in oesophageal stenosis caused by achalasia, reflux oesophagitis, and other acquired causes of oesophageal strictures. The diagnosis of TBR may be suspected based on examinations such as oesophagoscopy and oesophagography, which remain the primary diagnostic tools. Oesophageal manometry test and endoscopic ultrasonography (EUS) are also useful in the diagnosis of COS and its differentiation [9, 13, 15]. A barium

TABLE 1. Series of pediatric patients with tracheobronchial remnant: review of the literature

Author	Year of publication	No. of patients	Methods of operation
Nihoul-Fekete *et al.*	1987	4	Segmental resection
Olguner *et al.*	1997	1	Segmental resection
Ramesh *et al.*	2001	1	Segmental resection
Usui *et al.*	2002	1	Segmental resection
Amae *et al.*	2003	6	Segmental resection
Maeda *et al.*	2004	1	Circular myectomy
Zhao *et al.*	2004	3	Segmental resection
Saito *et al.*	2008	1	Circular myectomy
Nemolato *et al.*	2008	1	Segmental resection
Urushihara *et al.*	2013	2	Segmental resection
Sagna *et al.*	2014	3	Segmental resection
Mai *et al.*	2015	1	Segmental resection
Kim *et al.*	2017	29	Segmental resection
Presented study	2018	1	Segmental resection

oesophagogram identifies tapered oesophageal stenosis with proximal dilatation in the lower part of the oesophagus. The oesophagoscopy reveals an annular rigid stricture, which is not prone to balloon dilatation but is easily performed in oesophageal achalasia. The manometric examination may exclude the disturbances of the basic pressure of the lower oesophageal sphincter, characteristic for achalasia. Definitive diagnosis can only be made after histopathological examination.

A high-frequency catheter probe EUS may be useful in the evaluation of mucosal and submucosal lesions of the oesophagus [15].

There is no uniform treatment strategy in patients with CES. Conservative procedures, such as balloon dilatation or bouginage, may be effective in cases of stenosis caused by FM or MW, but patients with TBR do not respond to such treatment and require surgical removal of the tracheal remnant through circular myectomy – enucleation or segmental resection of the stenotic segment and oesophageal anastomosis [3–5, 13, 14]. Reports from other centres are presented in Table 1. The length of the stenosis caused by TBR is usually about 1–2 cm, as presented in our patient [5]. We confirmed the extent of oesophageal stenosis and determined the resection with the assistance of intraoperative endoscopy. This procedure was also used and recommended by other authors [4].

A surgical treatment approach via laparotomy is commonly used, but thoracotomy can also be performed [16]. Segmental oesophageal resection and anastomosis require mobilisation of the upper end of the oesophagus to limit the tension as much as possible. Currently, minimally invasive methods may be used for partial oesophageal resection [16]. A laparoscopic or thoracoscopic approach can be performed even in small children. Urushihara described two cases of COS in children aged 23 months and 13 months (one with TBR), in whom he used laparoscopy and thoracoscopy to remove oesophageal stenosis [16]. Shortening of the oesophagus predisposes to gastroesophageal reflux (GER), therefore most authors carry out segmental oesophageal resection and end-to-end anastomosis together with fundoplication. In the case of circular myectomy, the most common complication is perforation of the oesophagus, fundoplication is not always performed during this surgical approach.

CONCLUSIONS

In summary, in the case of swallowing disorders in young children and after exclusion of other causes such as post-inflammatory stenosis, oesophageal reflux, or oesophageal achalasia, the possibility of occurrence of TBR should be considered. In these patients, oesophageal dilatations are impossible to perform and may lead to its perforation. Children with TBR should be operated in centres with long experience in oesophageal surgery. Surgical resection of stenotic distal oesophagus is recommended in patients with TBR, but in some cases excision of the tracheobronchial remnant through myectomy may be possible. Histopathological examination is necessary to confirm the diagnosis. Postoperative complications are uncommon, but patients should be followed up closely [9]. If post-operative stenosis in the anastomosis site is diagnosed, endoscopic examination and balloon dilatation should be performed.

DISCLOSURE

The authors declare no conflict of interest.

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