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CASE REPORT

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Leiomyoma of the esophagus and bronchus in a child

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Abstract Leiomyomas of the esophagus and/or the bronchus have rarely been reported in children. To our knowledge, the simultaneous presence of this tumor in both the esophagus and a bronchus in a child has not been previously reported. A 7-year old boy presented with respiratory and esophageal symptoms and was found to have a leiomyoma of the esophagus and right main bronchus. The esophageal leiomyoma was treated with limited myotomy, but bronchoscopic resection was possible for the bronchial lesion. The postoperative result was excellent, with normal swallowing and no residual respiratory problems at 1-year follow-up.

Key words Bronchus · Esophagus · Leiomyoma

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Introduction

Esophageal leiomyoma (EL) is rarely seen in children, Bourque et al. [2], having collected only 22 cases in an extensive review of the condition from the period 1916–1988. A leiomyoma arising from a main bronchus is even more rare; only five cases have been reported, two of them in children [4, 7, 8]. To our knowledge, the synchronous occurrence of leiomyomas in the esophagus and bronchus in a child has not previously been reported.

Case report

A 7-year old male was referred to King Faisal Specialist Hospital, Riyadh, with a history of a recurrent chest infection and cough with expectoration of whitish sputum for 1 year and dyspnea for 1 month. He also suffered from occasional dysphagia. Bronchoscopy and biopsy of a lesion from the right main bronchus at the referring hospital showed a bronchial leiomyoma (BL). On admission, the child's general condition was satisfactory with normal vital signs. There was decreased air entry on the right side of the chest; the heart and abdominal examinations were normal. Various laboratory investigations, including arterial blood gases on room air, were normal. A chest radiograph showed a widened mediastinum with atelectasis of the right middle lobe and part of the right upper lobe. Computerized tomography (CT) of the chest confirmed the presence of a soft-tissue mass in the subcarinal region, which surrounded the esophagus.

Magnetic resonance imaging (MRI) of the chest showed an elongated, thickened structure surrounding the esophagus from the upper thoracic region (T2-3) to the level of the diaphragm. There was also a localized filling defect in the upper margin of the right main bronchus (Fig. 1). An upper gastrointestinal (GI) contrast study revealed an irregular esophageal stricture 8–9 cm long together with a sliding hiatus hernia and marked gastroesophageal reflux (GER). The esophagus was dilated above the stricture, and there was extensive esophageal mucosal ulceration (Fig. 2). The patient was started on antireflux therapy for 4 months, including ranitidine and cisapride.

The patient underwent a bronchoscopy and right posterolateral thoracotomy. Near-total resection of the right main bronchus leiomyoma was carried out through a rigid ventilating bronchoscope using biopsy forceps. At thoracotomy, the esophagus was diffusely thickened from the level of the carina to the gastroesophageal junction. The esophageal and bronchial lesions were separate, there being no extension from one to the other. Multiple frozen-section biopsies from the esophagus revealed an inflammatory reaction except in a small area measuring 3 mm in the mid-esophagus, which was reported to contain leiomyomatous cells. Formal histologic examination of the esophageal and bronchial lesions confirmed the diagnosis of EL and BL. A limited myotomy and intraoperative esophageal dilatation were carried out. The operating findings suggested the presence of a multifocal EL. However, the main histologic feature was diffuse fibroinflammatory esophagitis, and an esophagectomy was considered unnecessary. The intra- and postoperative course was uncomplicated and the patient was discharged a few days later. His respiratory difficulty and occasional dysphagia were markedly improved.

The patient was readmitted 3 months later for re-evaluation. At bronchoscopy, a small tumor remnant was seen in the right main bronchus and was resected using a cup forceps. Esophagoscopy showed a lower esophageal stricture, which was easily dilated to 42F. A further bronchoscopy and

esophagoscopy 1 year after surgery showed no signs of recurrence, and the patient was asymptomatic. Although GER was noted on the first GI study, at the repeat esophagogram and esophagoscopy the reflux and esophagitis were markedly improved.

Discussion

Leiomyoma of the esophagus is an uncommon tumor in adults, and is rare compared with esophageal carcinoma [10]. An extensive review of the world literature until 1971 by Seremetis et al. [10] provided 838 cases, the youngest patient being 12 years old. Bourque et al. reviewed 22 cases in children, including 2 of their own cases, and found that overall 2.6% of all reported cases occurred in children [2]. The clinical picture of an EL is different in children compared to adults. It is more common in females; the diffuse form predominates, and 22% of cases are associated with a familial syndrome. Dysphagia is the commonest presenting symptom, followed by dyspnea, vomiting, retrosternal pain, and coughing. Achalasia was the initial diagnosis in a significant number of cases [2, 9]. The preoperative diagnostic workup should include a chest radiograph, upper GI studies, and CT scan. Endoscopy is essential, but biopsies rarely yield a positive result and may render enucleation difficult [12]. An accurate preoperative diagnosis is essential, however, to facilitate surgical planning. Various surgical procedures have been described to treat EL, including myotomy, enucleation, esophagectomy, esophagogastrectomy, and endoscopic removal [2, 10]. Esophageal resection carries a high morbidity and mortality, but may be necessary in cases with diffuse involvement of the esophagus [2, 10].

Leiomyoma of the lung is extremely rare, 66 cases having been reported until 1982 [13]. Le-Tian et al. [6] reviewed 27 patients with tracheobronchial tumors and found only 1 with a leiomyoma. In the majority of cases, the leiomyoma was localized in the periphery [8–10, 12, 13]. Leiomyomas arising in the main bronchi are extremely rare, only five cases having been reported [7]. Three out of 51 cases reviewed by Orlowski et al. [8] arose from the main bronchi. The clinical presentation of a respiratory tract tumor is determined by its location; peripheral tumors may be asymptomatic, but bronchial tumors usually produce obstructive symptoms with cough and expectoration of blood or blood-stained sputum [8, 13].

A chest radiograph may reveal evidence of bronchial obstruction with atelectasis and pneumonia, or a round shadow in peripherally located lesions [13, 14]. The diagnosis is established by bronchoscopy and biopsy. BLs arise from the smooth muscle and grow as polypoid tumors, usually with a wide base. Histologically, BLs are cellular neoplasms with minimal vascular or stromal components compared with peripheral leiomyomas, which tend to have a more fibrous and vascular component [14].

The surgical management of respiratory leiomyomas depends upon awareness of the nature of the tumor, its location, and the status of the distal lung tissue. White et al. [14] found that 65% of reported cases were managed by lobectomy or pneumonectomy because of advanced, irreversible lung disease or failure to appreciate the benign nature of the lesion. A conservative approach is generally preferable, which may include bronchoscopic resection, sleeve resection with end-to-end anastomosis, bronchotomy, and bronchoplasty and resection of the tumor using a neodymium-yttrium aluminum garnet (ND-YAG) laser [1, 5, 7, 11, 14, 15].

Charles et al. [8] reviewed eight cases of BL, including a unique case of a 48-year-old male who presented with a right upper lobe BL 15 years after excision of an EL. Our patient appears to be the only case with synchronous leiomyomas of the esophagus and bronchus with predominantly respiratory symptoms. Although only one focus of leiomyoma was identified in the esophagus, the operative findings suggested the presence of multiple tumors, which could only be confirmed by undertaking esophageal resection, which could not be justified. The need for a fundoplication as part of the treatment in this patient was

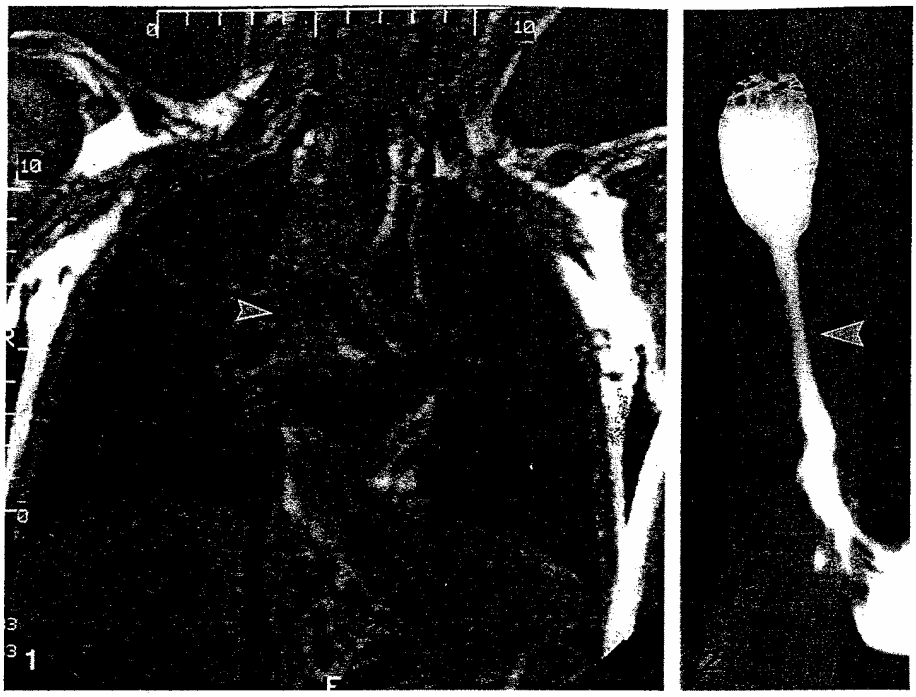

Fig. 1 Coronal MRI (T/W) scan showing intermediate-intensity mass involving esophagus down to cardia with second lesion involving right main bronchus (*arrowhead*)

Fig. 2 Contrast study of esophagus showing irregular narrowing distally (*arrowhead*) and mild dilatation proximally

discussed, but since the child showed a good response to medical and surgical treatment, we elected to observe him. If symptoms recur, however, we will consider antireflux surgery.

Our surgical approach represented teamwork of pediatric and otolaryngologic surgeons treating the esophageal and bronchial lesions on merit, but with a conservative approach to what is histologically a benign tumor. However, we plan to maintain close long-term follow-up to identify and manage recurrent tumors if and when they arise.

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