Case report

Asymptomatic congenital oesophageal cyst infiltrating the lung: an unusual complication

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Abstract

An asymptomatic 14-year-old boy presented with minor chest trauma. Chest X-ray showed opacity in the upper zone of the left lung. Further investigations with magnetic resonance imaging showed it to be a posterior mediastinal mass with a fistula to the lung, along with cystic changes in the left upper lobe of lung. Left upper lobectomy with excision of the mass was performed. Histological examination later showed an oesophageal duplication cyst with a fistula to the left upper lobe of the lung. We present this unusual complication of an oesophageal cyst infiltrating the lung in an asymptomatic child. © 2000 Elsevier Science B.V. All rights reserved.

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1. Introduction

Oesophageal duplication cyst represents a rare congenital foregut anomaly. Its incidence has been noted as 1/8200 with male preponderance [1]. During 7th week of embryonic life the oesophageal epithelium proliferates until the lumen is nearly filled with cells. Irregular spaces within the cellular mass form communication channels or vacuoles. These vacuoles coalesce and a single lumen is restored. It has been postulated that oesophageal duplication cysts are formed due sequestration of these vacuoles and failure to recanalize the foregut lumen in a complete fashion between the 5th and 8th week of embryonic life [2]. The location of the cyst and the presence or absence of complications determine the presenting symptoms. To our knowledge an asymptomatic child with oesophageal cyst infiltrating the lung with fistula formation has not been described in the literature previously.

2. Case report

An asymptomatic 14-year-old male patient was referred to our hospital following minor chest trauma. His chest X-ray taken to rule out any chest injury, showed a homogenous soft tissue mass in the left hilar region, together with cystic changes of the upper lobe of the left lung. The other areas of the lung were normal (Fig. 1). Further investigations with computerized tomographic scan and magnetic resonance imaging showed a well circumscribed ovoid mass in the left paravertebral region centred at the level of T5/T6 vertebrae, extending anteriorly up to the hilum of the left lung. The mass was connected to multiple cavitating lesions in the posterior part of the upper lobe of the left lung by a band of soft tissue density (Fig. 2). Bronchoscopy showed extrinsic compression of the left mainstem bronchus with normal mucosa. Bronchial lavage fluid was negative for malignant cells. Culture done for mycobacterium tuberculosis was negative. The prospect of biopsy was fraught with danger of infection, seeding of malignant cells if the lesion was malignant, bleeding and pneumothorax [3], and decision was therefore, taken to proceed with surgical exploration. Surgical access was gained by left posterolateral thoracotomy. A solitary cyst measuring $3 \times 2.5$ cm was found in relation to the outer wall of the upper third of the oesophagus, this was infiltrating the upper lobe of the left lung and contained thick yellowish fluid, which did not yield any positive results by culture or microscopy examination. Frozen section examination of the cyst wall did not reveal any evidence of malignancy. Due to the extensive destruction of the lung parenchyma, the excision of the cystic mass was done along with left upper lobectomy. Patient had an uneventful postoperative period and was discharged home.
Fig. 1. Chest X-ray PA view showing hilar mass with left apical lung infiltration.

Fig. 2. Magnetic resonance imaging – sagittal view showing mass with cystic lesions in the upper lobe of left lung with the band of soft tissue density.
after 8 days. The histopathological examination showed pseudo-stratified columnar epithelium with two muscle layers thereby fulfilling the criteria for oesophageal duplication cyst [2], with fistulous connection to the lung. The lung showed inflammatory and cystic changes. On follow up at 6 months, patient did not have any respiratory or gastrointestinal symptoms, clinical examination was normal and the patient had a normal growth pattern.

3. Discussion

Oesophageal cyst is a rare entity. Arbona et al. after extensive searches of the literature were able to review and classify only 91 congenital oesophageal cysts reported in literature [2].

The modes of presentation of oesophageal duplication cyst are depending on the size and proximity to other organs. The location of the cyst usually determines the time of presentation and associated symptoms [1]. Upper oesophageal cysts usually lead to respiratory distress in infancy due to tracheobronchial compression, which in turn leads to development of emphysema and recurrent respiratory tract infection [2]. Middle oesophageal cysts can lead to cardiovascular symptoms, such as arrhythmias and mediastinal compression syndromes. Gastrointestinal disturbances can also occur and include dysphagia, vomiting and regurgitation. However, about 35% of patients with middle and lower oesophageal cysts are asymptomatic and diagnosis is made incidentally or at autopsy [2]. In our patient even though the cyst was in relation to the upper oesophagus and had caused a fistula to the lung the patient remained asymptomatic.

Oesophageal cysts are included in the differential diagnosis of mediastinal mass in the adults and children. The radiological evidence in this patient showed a mass in the upper mediastinum and extensive lung parenchymal destruction. This raised a strong suspicion about malignancy or tuberculosis on the basis of the radiological appearance. On the contrary the patient did not show any clinical features suggestive of either of these conditions.

Once the malignant nature of the lesion was ruled out on frozen section, excision of the irreversibly damaged lung was performed along with the cyst with a successful outcome. A possible explanation of this complication would be compression of the adjacent lung tissue by the cyst causing progressive emphysema and cystic changes. Eventually the foregut cyst eroded into the lung causing a fistula.

This may raise the possibility of two separate pathologies, i.e. oesophageal duplication cyst and congenital pulmonary cyst. This possibility was considered but a detailed histopathological examination of the lung specimen proved the cysts to be of inflammatory origin thereby ruling out the possibility of dual pathology.

The asymptomatic nature of presentation, ominous looking radiological findings but an entirely benign pathology with an unusual complication prompted us to report this case. Of all the 220 case reports available about duplication cysts of oesophagus, such a complication of a fistula formation to the lungs has so far not been described.

References