CASE REPORT

Pleomorphic adenoma presenting with a mediastinal massYoung Kyung Lee¹, Yee Hyung Kim², Gou Young Kim³ & Hyo Chul Youn⁴¹ Department of Radiology, Seoul Medical Center, Seoul, Korea² Department of Pulmonary and Critical Care Medicine, Kyung Hee University Hospital at Gangdong, School of Medicine, Kyung Hee University, Seoul, Korea³ Department of Pathology, Kyung Hee University Hospital at Gangdong, School of Medicine, Kyung Hee University, Seoul, Korea⁴ Department of Thoracic and Cardiovascular Surgery, Kyung Hee Medical Center, School of Medicine, Kyung Hee University, Seoul, Korea**Keywords**

Chest CT; endoscopic ultrasound; mediastinum; pleomorphic adenoma.

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Abstract

Pleomorphic adenoma is the most common type of salivary gland tumor. Although it rarely affects the trachea and bronchus, it usually grows in the airway and presents as an endobronchial mass. We recently experienced a case of pleomorphic adenoma originating in the bronchus and growing outward into the mediastinum. We present this case with review of the computed tomography (CT) scan of the patient's chest, the endoscopic ultrasound (EUS), and the histopathology report.

Introduction

Pleomorphic adenoma (benign mixed tumor) is the most common type of salivary gland tumor.¹ A few cases of pleomorphic adenoma involving the trachea or bronchus have been reported.^{2,3} However, cases presenting with a mediastinal mass are very rare.^{4,5} Herein, we aim to describe a case of primary pleomorphic adenoma presenting with a mediastinal mass that was confirmed by surgical biopsy.

Case description

A 54-year-old woman visited our hospital to be evaluated for abnormalities that were previously found during a computed tomography (CT) scan of the chest. She had no relevant medical history and was a non-smoker. That patient had begun to develop blunt chest pain upon bending forward about two weeks prior to presentation. Her vital signs were stable. Rales or wheezing were not audible on auscultation. Physical examination of the head and neck did not reveal any mass-like lesions. The patient's blood chemistry and cell counts were within normal limits. Tumor markers, including α -FP, CEA and β -HCG, were 2.2 ng/mL, 3.1 ng/mL, and

1.3 mIU/mL, respectively, and were all within normal range. Electrocardiogram (ECG) showed incomplete right bundle branch block with normal sinus rhythm. Echocardiography revealed no specific abnormalities. Pulmonary function was also normal.

Chest radiography did not show any abnormal findings. However, a chest CT scan revealed a well-circumscribed, posterior mediastinal mass adjacent to the pulmonary artery and vein (Fig. 1). The mass was $5.9 \times 4.5 \times 2.5$ cm and was well enhanced with contrast media. Multiple low-densities were seen within the mass. Neither parenchymal lung masses nor any mediastinal lymphadenopathy were observed. In addition, chest CT and bronchoscopy did not reveal any endobronchial abnormalities. Upper gastrointestinal endoscopy was performed to rule out esophageal malignancy, but revealed no evidence of a tumor originating in the esophagus. Endoscopic ultrasound (EUS) revealed a well-marginated, hypoechoic, solid mass with internal multi-cystic changes (Fig. 2).

On the fifth day following her hospital visit, the patient underwent video-assisted thoracic surgery (VATS) for biopsy. Thoracoscopy showed that a $6.0 \times 4.5 \times 2.8$ cm firm, well-circumscribed, solid mediastinal mass with a cystic compo-

Figure 1 Chest computed tomography (CT) scans taken at the time of the hospital visit showed a large, well-circumscribed, well-enhanced mediastinal mass with internal multiple low-attenuation sites at the level of the subcarina.

ment was located in the subcarinal area (Fig. 3). It was firmly adhered to the left main bronchus. The cross section of the mass showed a well-defined lobulating, gray/yellow, firm, and myxoid solid area with cystic change. No hemorrhage or necrosis was noted (Fig. 4A). Microscopically, the mass was composed of epithelial and stromal elements. The epithelial component was glandular in nature with epithelial and myoepithelial two-cell linings. The stromal component had a nonspecific fibromyxoid appearance with spindle cells (Fig. 4B). Special immunohistochemical stains for myoepithelial cells, such as P63 and SMA, were positive. No features suggestive of malignancy were noted. A final diagnosis of

Figure 2 Endoscopic ultrasound (EUS) revealed a well-margined, hypoechoic, solid mass with internal multi-cystic changes (thick arrows) adjacent to the descending aorta (thin arrow).

Figure 3 Thoracoscopy revealed that a $6.0 \times 4.5 \times 2.8$ cm firm, well-circumscribed, solid mediastinal mass was located in the subcarinal area (A). It was firmly adhered to the left main bronchus. During removal of the mass, the left main bronchus was torn. Figure 3B shows the image after removal of mediastinal mass.

primary benign pleomorphic adenoma growing into the mediastinum and the trachea was reached.

Two years of follow-up chest CT scans and physical examinations revealed no recurrence of the primary tumor.

Discussion

Primary tracheal tumors represent only 2% of reported upper airway tumors.⁶ Pleomorphic adenoma, which is the most common type of primary mixed tumor of the parotid gland, rarely develops in the trachea. About 30 cases have been reported in the literature.^{2,3,7,8}

The pathogenesis of pleomorphic adenoma in the trachea remains a matter of controversy. Although these lesions are traditionally thought to originate from the epithelium of the submucosal glands,⁹ their occurrence in locations unrelated to any gland contradicts this hypothesis. It is possible that this

Figure 4 Grossly, the cut surface shows a well-defined whitish to yellowish, partly myxoid solid and cystic mass (a). Microscopically, the tumor has a biphasic appearance, composed of islands of epithelial and myoepithelial cells, and a fibromyxoid matrix (hematoxylin and eosin [H-E] stain, $\times 100$) (b).

neoplasm may originate from primitive stem cells that have the capacity to differentiate into ductal structures, myoepithelium matrices, and chondromyxoid matrices.¹ It is also possible that ectopic tissues can exist in organs other than the salivary gland or that a primary pleomorphic adenoma of the salivary gland can metastasize to other sites.^{4,5}

Most of the cases reported in the literature presented with endobronchial nodules or masses. This indicates that pleomorphic adenoma usually grows in the trachea or bronchus. Therefore, bronchoscopy plays a central role in diagnosis. However, in our case, we found a tumor originating from the left main bronchus that grew outward into the mediastinum, resulting in a large mediastinal mass. Blunt chest pain upon bending forward was the patient's sole symptom. No endobronchial abnormalities were identified by bronchoscopy, which is a very unusual feature of this type of tumor. Based upon the findings from radiological and histological exami-

nations, we were able to confirm a mediastinal primary pleomorphic adenoma originating from the bronchus. Although two previous cases of pleomorphic adenoma cases involving the mediastinum were found in the literature, to the best of our knowledge, there are no cases in the literature similar to the case we present here.^{4,5}

Because of its rarity, the radiological characteristics of mediastinal pleomorphic adenoma are not well known. In our case, a mediastinal pleomorphic adenoma presented with a well-circumscribed and well-enhanced mass with multiple internal low-attenuation sites on chest CT. In addition, EUS showed a well-margined, hypoechoic, solid mass with internal multi-cystic changes. The findings from EUS for a mediastinal pleomorphic adenoma have not been described before. The diagnostic value of positron emission tomography (PET) scans in the differentiation of malignant and benign salivary gland tumors was limited, because the standardized uptake value (SUV) of the pleomorphic adenoma was increased moderately.^{10–12} Given the cases of salivary gland tumors, the SUV of mediastinal pleomorphic adenoma would also be increased. Therefore, PET scans would not differentiate pleomorphic adenoma from other malignancies.

The biological behavior and clinical course of pleomorphic adenoma are not well described. Only cases that showed malignant and aggressive behavior have been described.¹³ Metastasis to other organs – so called, “benign metastasizing pleomorphic adenoma” – has been reported.¹⁴ Thus, such tumors should be considered potentially malignant, and long-term follow-up is mandatory after complete resection. Our patient was followed for two years after resectional surgery, and there was no recurrence during this period.

In conclusion, pleomorphic adenomas can present as mediastinal masses, even in patients without medical histories of this tumor.

Disclosure

No authors report any conflict of interest.

References

- 1 Moran CA, Suster S, Askin FB, Koss MN. Benign and malignant salivary gland-type mixed tumors of the lung. Clinicopathologic and immunohistochemical study of eight cases. *Cancer* 1994; **73**: 2481–90.
- 2 Yan HC, Shen CY, Chiang CH *et al.* Pleomorphic adenoma of the trachea: report of two cases. *J Formos Med Assoc* 1991; **90**: 1124–7.
- 3 Kajikawa S, Oki M, Saka H, Moritani S. Pleomorphic adenoma of the trachea. *Respiration* 2010; **80**: 433–4.
- 4 Feigin GA, Robinson B, Marchevsky A. Mixed tumor of the mediastinum. *Arch Pathol Lab Med* 1986; **110**: 80–1.

- 5 Steele NP, Wenig BM, Sessions RB. A case of pleomorphic adenoma of the parotid gland metastasizing to a mediastinal lymph node. *Am J Otolaryngol* 2007; **28**: 130–3.
- 6 Schneider P, Schirren J, Muley T, Vogt-Moykopf I. *Primary tracheal tumors: experience with 14 resected patients*. *Eur J Cardiothoracic Surg* 2001; **20**: 12–8.
- 7 Baghai-Wadji M, Sianati M, Nikpour H, Koochekpour S. Pleomorphic adenoma of the trachea in an 8-year-old boy: a case report. *J Pediatr Surg* 2006; **41**: e23–26.
- 8 Paik SS, Jin YH, Park CK, Shin DH, Chung WS, Lee JD. Pleomorphic adenoma of the trachea. *J Korean Med Sci* 1997; **12**: 564–6.
- 9 Davis PW, Briggs JC, Seal RM, Storrington FK. Benign and malignant mixed tumours of the lung. *Thorax* 1972; **27**: 657–73.
- 10 Matsuda M, Sakamoto H, Okamura T *et al.* Positron emission tomographic imaging of pleomorphic adenoma in the parotid gland. *Acta Otolaryngol Suppl* 1998; **538**: 214–20.
- 11 Rubello D, Nanni C, Castellucci P *et al.* Does 18F-FDG PET/CT play a role in the differential diagnosis of parotid masses. *Panminerva Med* 2005; **47**: 187–9.
- 12 Uchida Y, Minoshima S, Kawata T *et al.* Diagnostic value of FDG PET and salivary gland scintigraphy for parotid tumors. *Clin Nucl Med* 2005; **30**: 170–6.
- 13 Demirag F, Topcu S, Kurul C, Memis L, Altinok T. Malignant pleomorphic adenoma (malignant mixed tumor) of the trachea: a case report and review of the literature. *Eur Arch Otorhinolaryngol* 2003; **260**: 96–9.
- 14 Vivian MA, Sahni VA, Lowe AC, Silverman SG. Benign metastasizing pleomorphic adenoma presenting as a solitary kidney mass: imaging features. *Urology* 2012; **80**: e17–18.