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Two Roads to Diagnose Primary Esophageal Adenoid Cystic Carcinoma in the Mediastinum

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Adenoid cystic carcinoma (ACC) is a malignant epithelial tumor that predominantly originates in the salivary glands of the head and neck. However, ACC arising in the thoracic cavity is a rare entity. Diagnosis of primary esophageal ACC (EACC) involves use of gastrointestinal endoscopic ultrasound fine-needle aspiration; however a low diagnostic yield has been reported in the literature. We describe an uncommon presentation of EACC debuting with predominant airway obstructive symptoms and diagnosed through endobronchial ultrasound-guided fine-needle aspiration. We propose the use of an endobronchial sonographic approach to diagnose EACC and other esophageal malignant neoplasms with mediastinal involvement and airway proximity.

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During early embryonic development, the respiratory diverticulum originates from the primitive foregut where branching occurs, forming the trachea anteriorly and the esophagus posteriorly. As such, malignant processes occurring within the esophagus or trachea may have important diagnostic and clinical implications for one another. We report a rare esophageal malignant neoplasm located in the posterior mediastinum presenting with airway obstruction most likely related to the anatomic origin of these structures. Adenoid cystic carcinoma (ACC) is a malignant epithelial tumor that usually originates in the salivary glands of the head and neck.[1,2] Primary esophageal ACC (EACC) is extremely rare, accounting for <0.1% of primary tumors in the esophagus.[2,3] EACC usually has submucosal involvement on presentation; thus, it is believed to originate from the submucosal esophageal glands.[2] Approximately 60 cases have been reported in the literature.[4] The most common clinical presentation includes progressive dysphagia and food regurgitation. Initial diagnostic modalities for EACC include gastrointestinal endoscopic ultrasound fine-needle aspiration (EUS-FNA), but low diagnostic yields have been reported in the literature.[5] Moreover, many cases of EACC are diagnosed after surgical resection.[4] Herein, we present a case of EACC diagnosed through endobronchial ultrasound-guided fine-needle aspiration (EBUS-FNA).

An 81-year-old man with a 17-pack-year smoking history presented with a 6-month history of nonproductive cough and progressive dyspnea. In addition, he had chronic, mild, nonprogressive dysphagia. Pulmonary function tests revealed a severe obstructive ventilatory impairment. Chest computed tomography showed a 2.7 × 2.1-cm lesion in the subcarinal compartment of the mediastinum (Figure 1). In view of his predominant respiratory symptoms and the lesion's proximity to the airway, bronchoscopy was performed, which showed a left main obstruction due to apparent external mass effect with left lung collapse in all its segments (Figure 2A). Endobronchial ultrasound showed a lesion with mixed echogenicity (Figure 2B). EBUS-FNA revealed aggregates of atypical epithelial cells arranged in a tubular and cribriform pattern, microcystic spaces (Figure 3A), and globules of extracellular matrix surrounded by basaloid cells (Figure 3B). The immunostaining had strong positivity for CK7 (Figure 3C) in the ductal component and P63 in the myoepithelial component (Figure 3D); nonetheless, it was positive for CD117 (c-kit), S100, and smooth muscle actin, consistent with ACC. Subsequent upper endoscopy with EUS-

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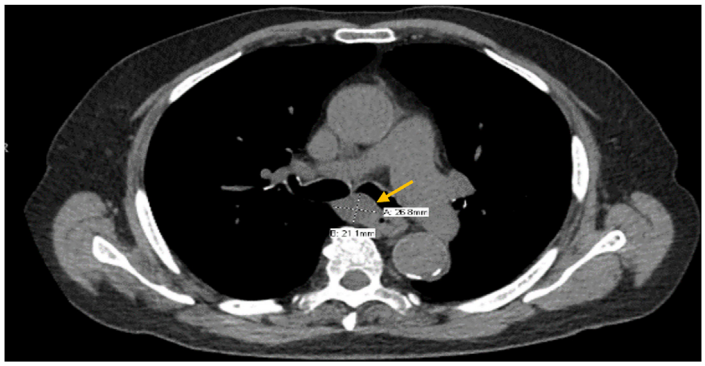

FIGURE 1 Chest computed tomography. Mediastinal 2.7 × 2.1-cm lesion in the posterior subcarinal compartment (arrow).

FNA showed a 2.8 × 2.7-cm hypoechoic lesion in the posterior mediastinum arising from the submucosal layer of the esophageal wall (Figures 2C, 2D). Therefore, it confirmed the esophageal origin of the ACC. No lymphadenopathy or metastasis was evidenced by bronchoscopic ultrasound, endoscopic ultrasound, or fusion positron emission tomography. The patient preferred nonsurgical management and thus is undergoing radiotherapy.

COMMENT

ACC is an uncommon malignant epithelial tumor that predominantly involves the minor salivary glands of the head and neck.[1,6] It is characterized by an indolent but aggressive course with early perineural invasion and

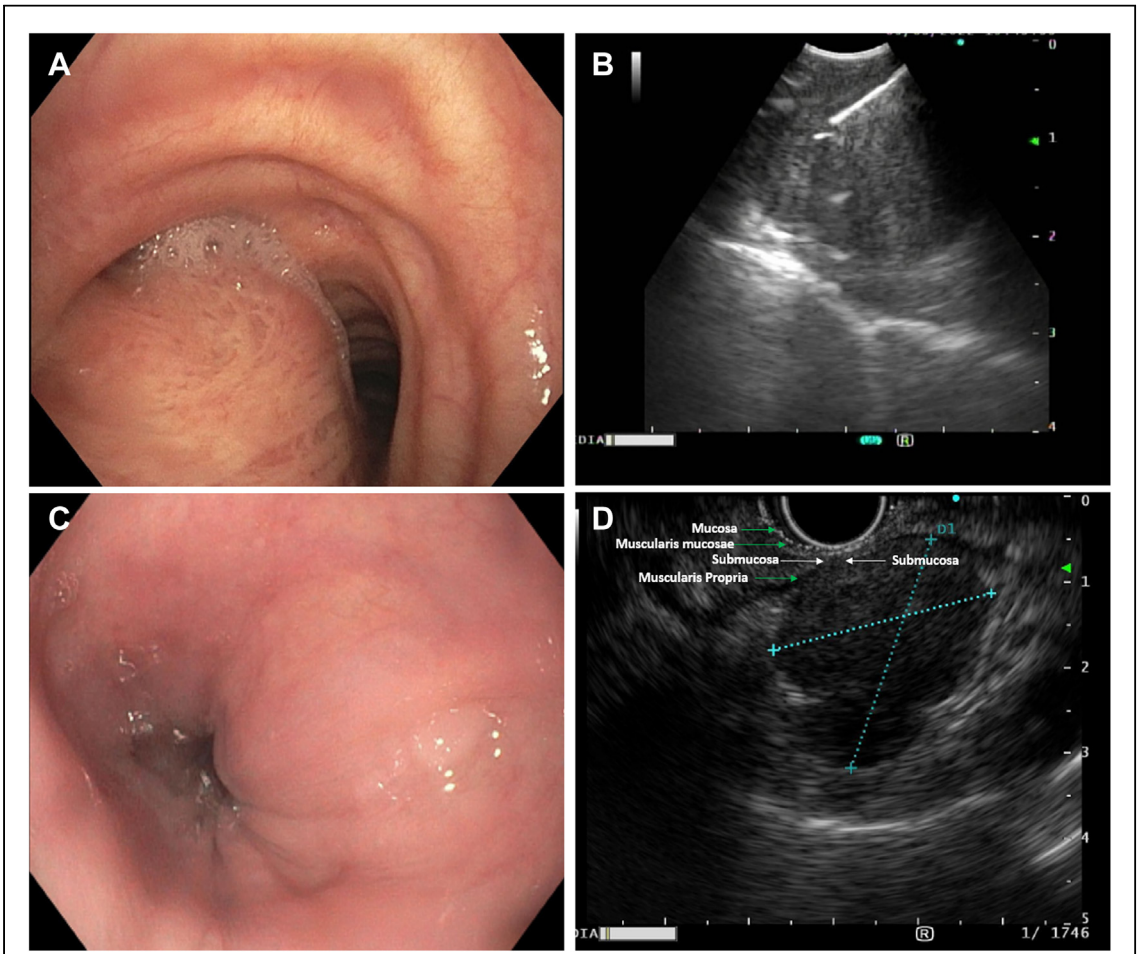

FIGURE 2 (A) Bronchoscopy showing main bronchi extrinsic compression from mediastinal mass. (B) Endobronchial ultrasound station 7 shows a mixed echogenic lesion with regular borders. (C) Upper endoscopy shows a subepithelial lesion at midesophagus with normal-appearing overlying mucosa and mild occlusion of the esophageal lumen. (D) Endoscopic ultrasound shows a hypoechoic lesion focally arising from the esophageal submucosal layer that extends into the posterior mediastinum (arrows).

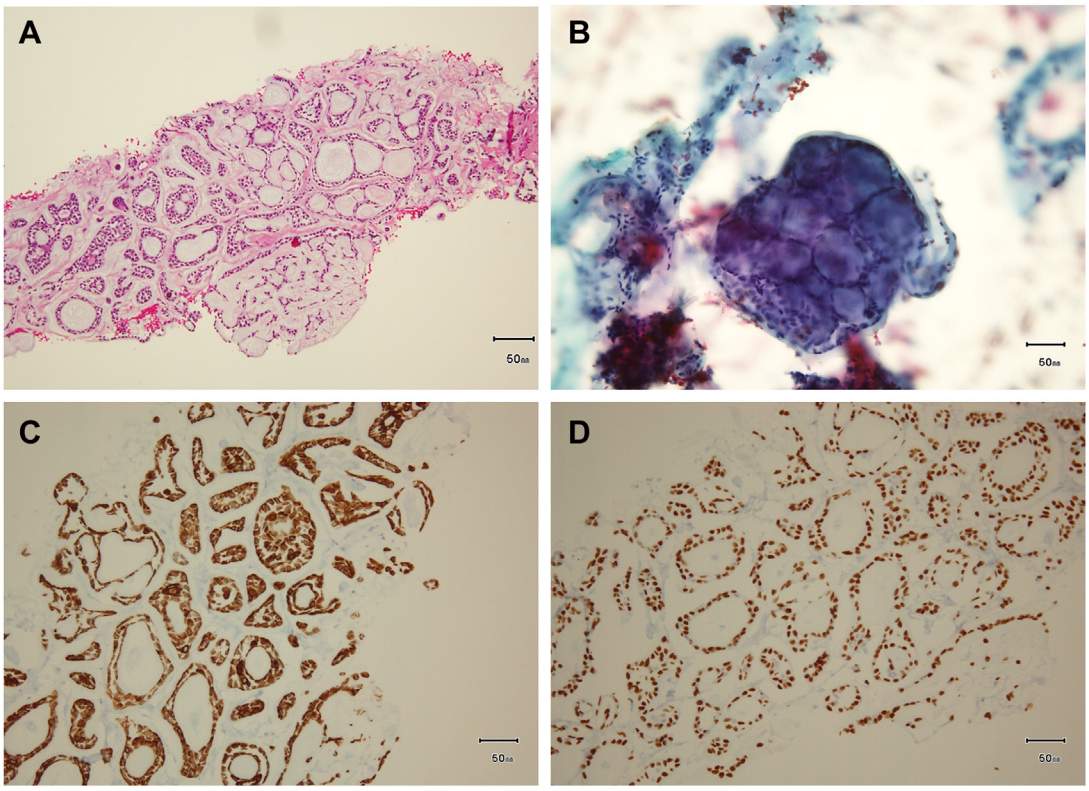

FIGURE 3 Pathologic findings of mediastinal tumor. (A) Mixed tubular and cribriform pattern (hematoxylin and eosin stain). (B) Papanicolaou smear shows metachromatic matrix globular spheres with defined borders, surrounded by basaloid cells. (C) Immunohistochemistry exhibits strong positivity for cytokeratin 7 (CK7) in the ductal component. (D) Immunohistochemistry: strong positivity for P63 in the myoepithelial component. (All images, magnification ×20.)

high rates of delayed distant metastases.[4,5] The incidence of primary EACC is estimated to be approximately 0.1% of esophageal tumors. These estimates, however, are derived mostly from the Japanese and Chinese literature as reports from Western countries are limited.[5] EACC has most frequently been reported involving the thoracic portion of the esophagus and less often in the lower or upper third.[4,7] The most common presentation is progressive dysphagia.[2] Nonetheless, our case shows an atypical manifestation with predominance of airway obstructive symptoms probably due to significant airway compression.

The diagnostic approach to EACC usually involves an initial biopsy through upper endoscopy. However, it is estimated that approximately 40% of cases are diagnosed correctly through EUS-FNA, with the rest being diagnosed during the postsurgical pathologic analysis.[5] Because of the predominance of respiratory symptoms in our patient and airway compression by the tumor, the diagnosis of ACC was carried out by transbronchial EBUS-FNA. EACC is commonly managed and treated with radical excision; evidence-based recommendations for use of chemotherapy or radiotherapy are lacking.[5] However, postoperative management of head and neck ACC includes adjunctive radiotherapy for improved locoregional control. Furthermore, definitive radiotherapy for unresectable tumors in combination with particle therapies has been associated with improvement in locoregional control and overall survival.[8] Thus, radiotherapy might be promising for poor surgical candidates.

ACC is classified into 3 main histologic patterns—cribriform, tubular, and solid. The solid type is associated with higher rates of metastases and worse outcomes.[1,4,6] The cribriform pattern is characterized by glandlike or cylindromatous microcystic spaces. The tubular form has well-formed ducts and tubules with central lumina filled with mucoid material, lined by inner epithelial cells and outer myoepithelial cells. Our case had a mixed tubular and cribriform pattern. ACC cells have a basaloid appearance with a small amount of cytoplasm and round hyperchromatic nuclei with a low mitotic activity.[6,7] Positive immunohistochemical staining, such as for P63,

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CK7, CD117(c-kit), S100, and smooth muscle actin, is diagnostic for EACC, all of which were positive in this patient.[3,5]

In conclusion, EACC is a rare tumor with scarce data in the literature. In this report, EBUS-FNA revealed a rare primary esophageal malignant neoplasm with histopathologic findings mostly seen in malignant lesions originating from the salivary glands. Respiratory symptoms secondary to mediastinal involvement with airway obstruction are even rarer, making this an atypical presentation. This case demonstrates the proof of principle of adding EBUS-FNA to the diagnostic armamentarium for EACC and other esophageal malignant neoplasms with mediastinal involvement and airway proximity. As such, we propose EBUS-FNA as an adjunctive diagnostic modality for esophageal tumors with airway proximity.

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PATIENT CONSENT

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