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Bronchoscopic management of a primary endobronchial salivary epithelial-myoepithelial carcinoma: A case report

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ABSTRACT

Here, we discussed a 55 y/o African man who recently immigrated from Nigeria to the United States and who presented to Parkland Memorial Hospital with a productive, intermittent cough of one year duration. The cough was associated with shortness of breath and chest pain. Cough was not associated with voice hoarseness, hemoptysis, melanoptysis, and wheezing. He had a computed tomography (CT) scan of the chest that showed a 1.9 cm mass in the right main stem bronchus with ipsilateral right lower lobe consolidation and bronchiectasis. The patient was seen by pulmonology who recommended bronchoscopy for diagnosis and possible intervention. Bronchoscopy showed a 90% obstructing mass in the proximal right mainstem bronchus and bronchus intermedius. The mass was large and endobronchial, circumferential, exophytic, and polypoid. The decision was made to undergo bronchoscopic tumor ablation using electrocautery snare, argon plasma coagulation (APC), suction, and forceps. The tumor was successful ablated. Microscopic examination revealed eosinophilic ducts tightly coupled with a surrounding layer of clear cell myoepithelial cells and the diagnosis of epithelial-myoepithelial carcinoma (EMC) of the lung was made. The patient was discharged from the hospital with scheduled outpatient visits for monitoring of the carcinoma by pulmonology and thoracic surgery. Unfortunately, he was lost to follow up.

1. Introduction

Malignant salivary gland tumors represent ~3% of all head and neck malignancies diagnoses yearly in the United States. In contrast to the more common mucosal head and neck cancers where tobacco and alcohol use are known as specific carcinogens, no risk factor has been identified for these salivary malignancies. Most of these tumors affect the parotid gland and they are rarely found outside the head and neck. The World Health Organization classifies malignant salivary gland tumors as carcinomas, nonepithelial tumors, lymphomas, metastatic and secondary tumors, as well as unclassified tumors. The most common subtypes include: mucoepidermoid carcinoma (34%); adenoid cystic carcinoma (22%), and adenocarcinoma (18%) [1].

Currently the pathogenesis of these tumors is thought to be due to malignant transformation of reserved stem cells of the salivary duct system. The stage in the stem cell differentiation where the transformation to malignant cell occurs gives rise to the different types of neoplasms that can occur. For example, the intercalated duct stem cells give rise to adenoid cystic and acinic cell carcinoma while the excretory duct stem cells give rise to mucoepidermoid, squamous cell, and salivary duct carcinoma [2].

Salivary gland neoplasms are differentiated based upon their histology; however, histology can also divide the tumors in low-grade, intermediate-grade, and high grade which can give some indication of the clinical behavior and prognosis [3]. Regardless of the histological subtype, the treatment for salivary gland neoplasms is resection of the entire tumor with adjuvant chemotherapy and radiation if needed [4].

Epithelial-myoepithelial carcinoma (EMC) is an uncommon low to intermediate grade salivary gland carcinoma that is mostly found in the parotid gland where it comprises less than 1% of all salivary gland tumors. EMC is a biphasic tumor: It is composed of variable proportions of two cell types: An inner layer of duct lining cells and an outer layer of

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clear cells, which typically form double-layered duct-like structures [4].

A counterpart to salivary gland tumors are lung primary salivary gland type tumors (SGT). Pulmonary SGT encompass a small but distinct group of neoplasms that together account for <1% of all lung tumors with EMC making up a very small portion of the pulmonary SGTs (around 3–4%) [5]. SGTs of the lung are thought to arise from submucosal bronchial glands in the lower respiratory tract and have morphologic features like those of their salivary gland counterparts [6,7]. EMCs of the lung usually occur in the 5th-6th decade of life with no male to female predominance and no correlation has been found between smoking and these tumors. Macroscopic examination reveals a well circumscribed but non-encapsulated polypoid lesion [6,7]. Immunostatins are notable for the epithelial component being positive for pancytokeratin (AE1/AE3) and negative for smooth muscle actin (SMA) while the myoepithelial cells are variably positive for markers such as p40, S100, calponin, and SMA [6,7]. The Diagnosis of EMC is made by the histopathology alone. Due to its low rate of lymph nodes involvement and well as distant metastasis, the recommended therapeutic choice is surgical excision [8]. There is, however, no "gold standard" of management in these patients [9].

2. Case presentation

A 55-year-old African man with a past medical history of hypertension presented to Parkland Memorial Hospital with a chief complaint of a worsening productive cough that was recurring for the past year. Cough was described as intermittent with occasional yellow sputum production and associated with shortness of breath and chest pain. He denied voice hoarseness, hemoptysis, melanoptysis, and wheezing. Patient also noted 5-pound weight loss over the past 2–3 months, but denied fever, chills, or night sweats. Patient was a recent immigrant from Nigeria where he worked as a mechanic and had a remote smoking history of ~1 pack/year in the early 1990s. He had no personal history tuberculosis nor he had known tuberculosis contacts. He had no personal or family history of cancer and he denied sick contacts. His surgical history was non-contributory. On physical exam he was afebrile with a regular respiratory rate and no adventitious lung sounds where noted. The patient had been seen for the same complaint at an outside primary care clinic two weeks prior and a chest X ray revealed a mild tracheal deviation to the right (Fig. 1A).

A computed tomography (CT) of the chest with contrast was obtained (Fig. 1B & C) and it was notable for a 1.9 cm mass in the right mainstem bronchus with associated right lower lobe consolidation and bronchiectasis. Pulmonology was consulted and management options were discussed. The patient was ultimately recommended to undergo bronchoscopy for airway examination, biopsy of endobronchial lesion, and potential therapeutic intervention.

Bronchoscopy showed a 90% obstructing mass in the proximal right mainstem bronchus and bronchus intermedius. The mass was large and circumferential, endobronchial, exophytic, and polypoid (Fig. 2A). The mass was too large to pass the therapeutic bronchoscope for assessment of the distal airways. The lesion was pretreated with epinephrine and cold saline. The tumor was then excised piecemeal using an electrocautery snare, forceps and suction. Tumor coagulation was performed using APC for hemostasis. Finally, balloon dilation was performed in the right mainstem bronchus and bronchus intermedius (Fig. 2B). The excised tissue was sent for histopathologic examination.

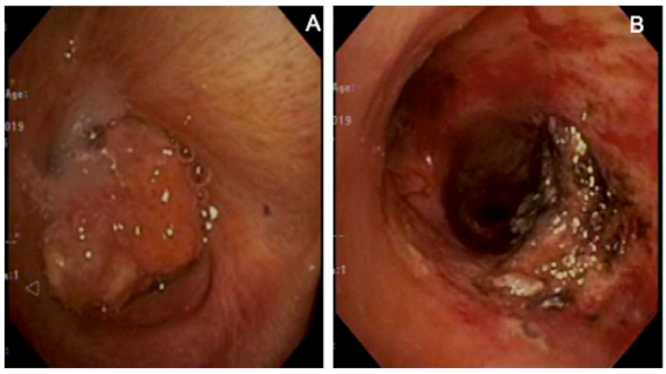

Fig. 2. Bronchoscopy Findings. [A] A 90% obstructing mass (circumferential, endobronchial, exophytic, and polypoid) in the proximal right mainstem bronchus and bronchus intermedius. [B] Right mainstem bronchus after airway recanalization.

Microscopic evaluation (Fig. 3) showed an infiltrating gland-forming neoplasm with two cell components: eosinophilic ducts, tightly coupled with a surrounding layer of myoepithelial cells with clear cytoplasm. Immunostains (Fig. 3) highlighted the two cell components, as the ducts were positive for AE1/AE3 and CD117 while the myoepithelial cells stained for SOX10, S100, p40 and p63.SMA was negative. The findings were diagnostic for epithelial-myoepithelial carcinoma. The tumor was interpreted as intermediate-grade on the basis of increased nuclear atypia and an elevated mitotic rate.

Patient was discharged from the hospital with scheduled outpatient visits for monitoring of the carcinoma by pulmonology and thoracic surgery. He, unfortunately, was lost to follow up.

3. Discussion

This is a rare case of patient with an endobronchial mass causing post-obstructive pneumonia, bronchiectasis and chronic cough. He was a male in his 50s with minimal smoking history and slow progressive symptoms over the preceding year. His tumor was found in the central airways and fit the description of a polypoid exophytic mass. His histology was notable for increased nuclear atypia which is unusual for this type of carcinoma.

Most of the 31 cases of pulmonary EMC reported in the literature were managed with surgical excision [9–12] with three other case reports documenting management by endobronchial intervention (Table 1). This management strategy is most likely due to its canonical salivary EMC counterpart, in which a wide surgical excision typically done [9,13]. This patient had a successful airway recanalization via flexible bronchoscope. Of note – due to the low rate of occurrence of these tumors and minimal data in the literature – there is no standard of

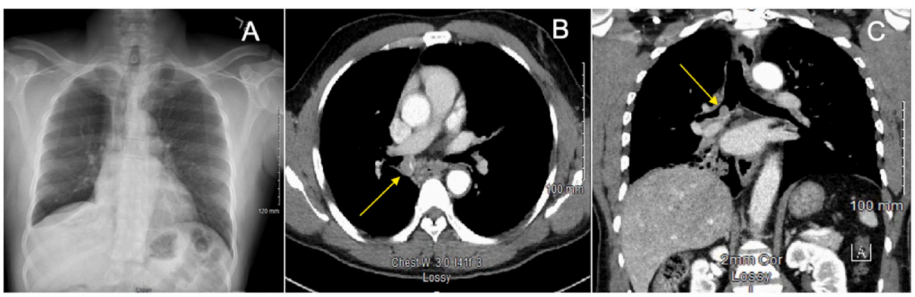

Fig. 1. Radiological Findings. [A] posterior-anterior plain chest film obtained 2 weeks prior to presentation with tracheal deviation to the right and hilar fullness. [B] Chest CT scan with contrast with axial view with right mainstem obstructing endobronchial mass (yellow arrow) [C] Coronal reconstruction with right mainstem endobronchial mass (yellow arrow) with evidence of post obstructive bronchiectasis. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

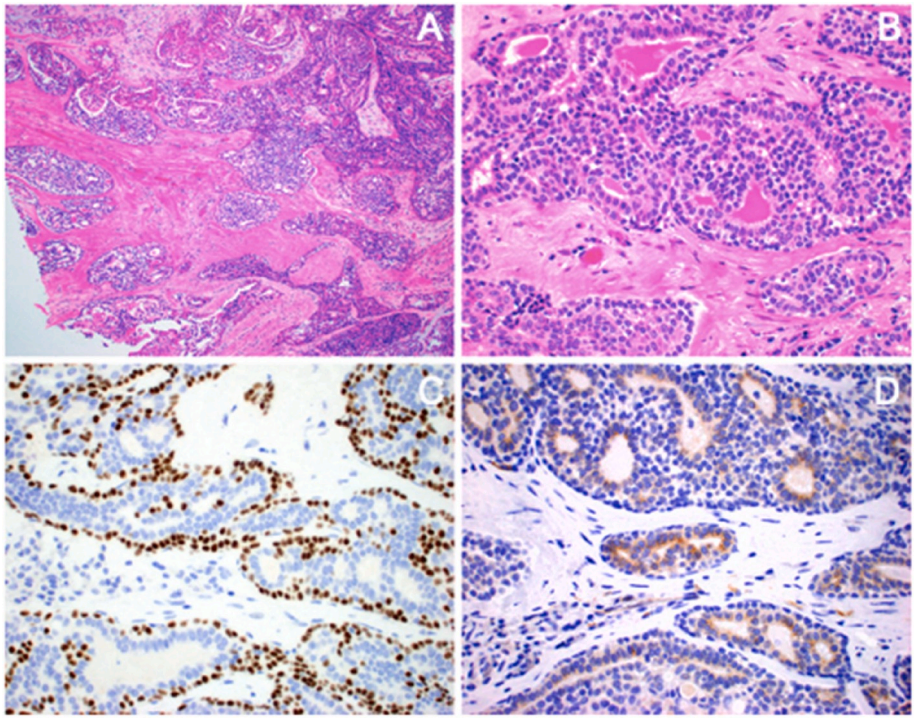

Fig. 3. Pathology Findings. [A] The tumor demonstrates an irregular, infiltrative pattern within a fibrotic stroma. [B] In the tumor nests, there are ducts that are tightly coupled to a surrounding layer of myoepithelial cells with pale cytoplasm. [C] The myoepithelial cells are positive for p40, while the ducts are not. [D] CD117 shows the opposite pattern, with staining seen only on the luminal surface of the inner ductal cells.

Table 1
Bronchoscopic management of EMCs in the Literature.

Reference	Location of Tumor	Management	Outcome
Chao TY et al. [16]	Left Main Stem Bronchus	Curative electrocauterization	No recurrence at 3 and 6 months
Muslimani AA et al. [17]	Left Main Stem Bronchus	Does not describe	Had recurrence at 8 months
McCracken D et al. [14]	Distal Trachea	Endobronchial laser ablation therapy with a Nd-YAG laser	No recurrence at 3 months

care defined in the management in these individuals.

Bronchoscopic interventions are utilized to successfully manage central airway obstruction due to benign or malignant tumors. Thermal therapies for tumor ablation or coagulation in the airways include electrocautery, argon plasma coagulation, neodymium-doped yttrium-aluminum-garnet laser (Nd-YAG) [14], and cryotherapy (either touch or spray) [15]. Choice of thermal tool is dependent on the location and characteristics of the airway lesion, operator preference, and device availability.

This patient originally underwent bronchoscopy for both diagnostic and therapeutic purposes. The tumor was excised for pathologic examination, and airway recanalization of right mainstem and bronchus intermedius was achieved. Given imaging with no nodal involvement, and after pathology returned with a diagnosis of EMC with known low risk of metastasis, management by the previously performed bronchoscopy was thought to be sufficient at this time. However, he was discharged with plans for serial clinic visits, imaging, and airway inspections, but unfortunately he was lost to follow up.

4. Conclusions

We conclude that EMC is a rare tumor that can be seen in patients who present with chronic cough without other presenting symptoms with an endobronchial lesion [9,14]. EMCs that are low to intermediate grade with no signs of lymph node involvement or distant metastasis can be successfully managed with bronchoscopic tumor ablation and/or if the patient is not a candidate for surgical resection. Given the potential risk of recurrence with EMCs, patients should have continued follow up with surveillance CT imaging and bronchoscopies for airway examination.

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Declaration of competing interest

The authors declare no conflict of interests regarding the data presented in this manuscript.

CRediT authorship contribution statement

Dalton T. Patterson: Writing - original draft, Investigation, Conceptualization, Visualization. **Quinn Halverson:** Writing - review & editing, Supervision. **Sarah Williams:** Writing - review & editing, Supervision. **Justin A. Bishop:** Writing - review & editing, Formal analysis, Visualization. **Cristhiaan D. Ochoa:** Conceptualization, Investigation, Visualization, Formal analysis, Supervision, Project administration, Funding acquisition, Writing - original draft. **Kim Styrvoky:** Conceptualization, Resources, Supervision, Writing - review & editing.

Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.rmcr.2020.101083.

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