# Pleomorphic adenoma of the trachea: a case report

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## Introduction

Pleomorphic adenomas (PA) are the most common benign tumors of the salivary glands. These tumors comprise approximately 65% of all salivary tumors and typically emerge in the upper airway salivary glands including the parotid, submandibular, sublingual, and minor salivary glands [1-3]. Though rare, PAs have been described in other regions of the aerodigestive tract including the soft and hard palates, nasal septum, buccal mucosa, and upper lip [4]. In this report, we present an extremely rare case of pleomorphic adenoma in the trachea.

## Case Report

An 83-year-old female was referred to the Department of Otolaryngology-Head and Neck Surgery at the University of Virginia Health System for management of an obstructive tracheal mass. She initially presented to her primary care physician with 1 month of worsening shortness of breath and blood in her oropharynx of undetermined origin. She noted an unintentional 10 pound weight loss over the past 1.5 years. Her past medical history was significant for hypertension and rheumatoid arthritis. The patient was an alcohol user and endorsed a longtime exposure to second-hand smoke, though denied a personal history of smoking. Her physical exam was unremarkable and was not in respiratory distress. A flexible bronchoscopy performed in clinic revealed a solitary, polypoid mass with a wide-base and smooth, hypervascular surface. The mass was obstructing approximately 90% of the posterior tracheal lumen approximately four tracheal rings below the cricoid cartilage. A computed tomography study of her neck and chest was performed at an outside hospital, which demonstrated a smooth tracheal mass measuring approximately 1.6 cm by 1.3 cm.

The decision was made to proceed to the operating room to obtain a biopsy of the mass and debulk the tracheal portion of the tumor. The location of the tracheal mass was reconfirmed to be about 3 cm below the vocal fold edge. The excised tumor was a smooth, polypoid mass with hypervascularity on the surface and measured approximately 4 cm x 3.5 cm x 1 cm. After the tracheal mass was debulked and flush to the tracheal wall using bronchoscopic cautery forceps and monopolar scissors, a flexible esophagoscopy was performed. This demonstrated normal esophageal mucosa. There was compression along the anterior esophageal wall approximately 18 cm from the incisor, presumably due to mass effect from the tracheal mass. The patient tolerated the procedure and was discharged on POD#3 after an unremarkable hospitalization.

On microscopic examination, the tumor consisted of bland basaloid proliferation growing in orderly cords with multifocal areas of squamous differentiation, with adjacent areas of myxoid stroma. The pathologic differential diagnosis on hematoxylin and eosin staining included pleomorphic adenoma, basal cell adenoma, and neuroendocrine tumor. Immunohistochemically, the tumor was negative for chromogranin and synaptophysin and strongly positive for p63 and smooth muscle actin (SMA). Immunohistochemistry for c-kit highlighted scattered mast cells and GFAP stains scattered individual cells. These histopathology characteristics were consistent with benign mixed tumor and the diagnosis was confirmed during surgical pathology consensus conference.

The patient was seen approximately 2 weeks following her procedure and flexible laryngoscopy revealed a widely patent airway with no evidence of tumor recurrence. She has subsequently returned to her primary Otolaryngologist for continued follow-up.

## Figure 1

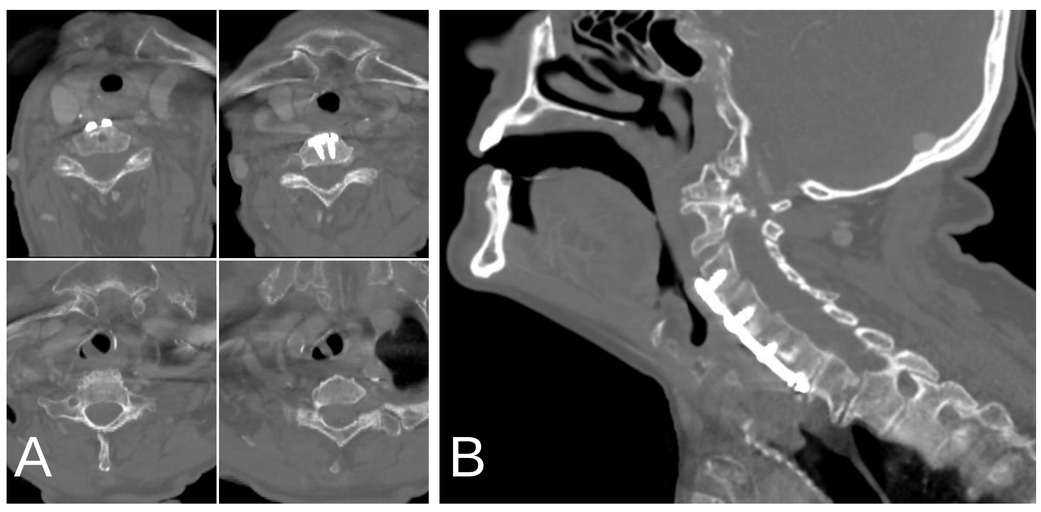


Figure 1: (A) Serial axial non-contrasted computer tomography images demonstrated an obstructive tracheal mass. (B) Sagittal oriented non-contrasted computer tomography image revealing the location of the tracheal mass approximately at the level of C6 to T1. Due to previous spinal and shoulder surgery, imaging was limited as the scan had multiple artifacts.

## Figures 2&3

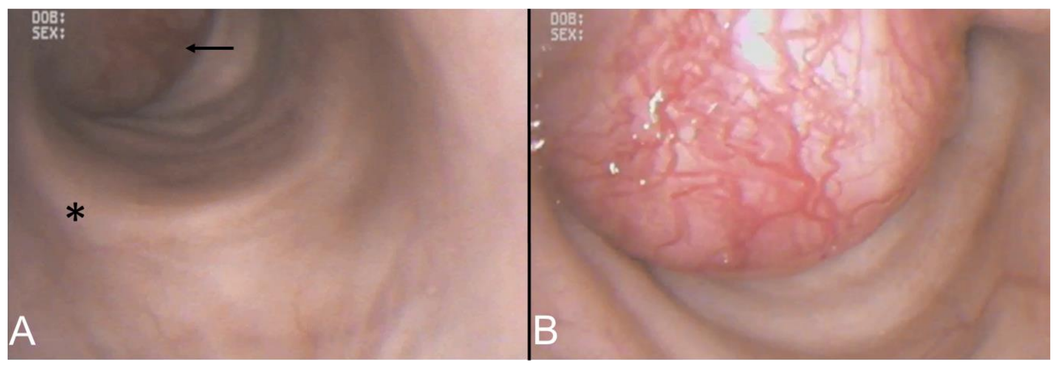


Figure 2: A. Flexible bronchoscopy in clinic revealed an obstructive mass (arrow) 4 tracheal rings inferior to the cricoid ring (asterisk). B. Closer view of the mass demonstrated a smooth, hypervascular surface and a wide-base pedicled to the lateral tracheal wall. The scope was circumnavigated passed the mass and revealed a patent trachea distally.

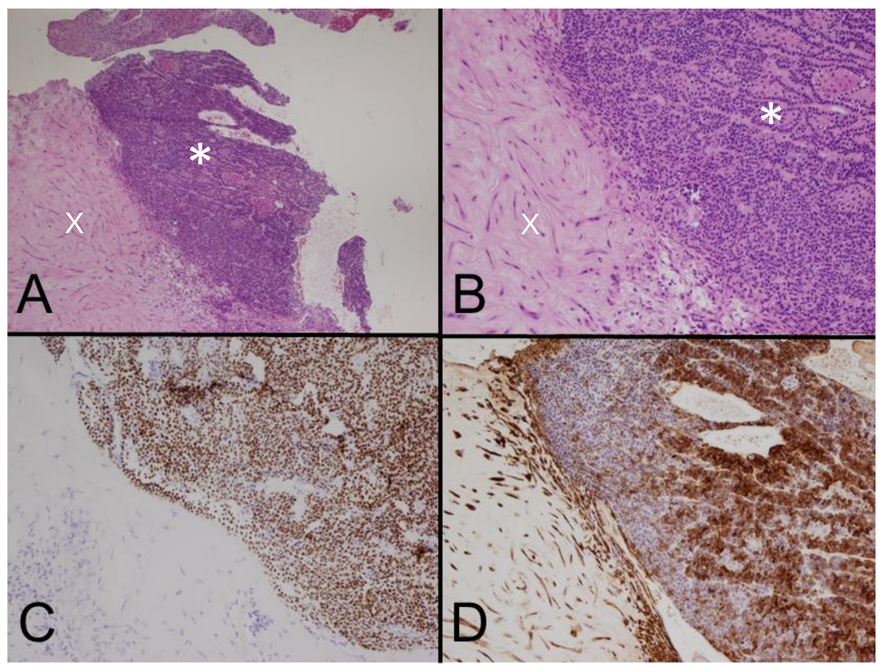


Figure 3: A. Hematoxylin and eosin stain at 40x magnification demonstrating basaloid and squamous epithelial component (asterisk) and stomal component (“x”) of the tumor. B. Hematoxylin and eosin at 100x magnification demonstrating the interface of the epithelial (asterisk) and stromal components (“x”) of the tumor. C&D. Immunohistochemical staining for p63 (C) and smooth muscle actin (D) are strongly positive. C-kit and GFAP stained scattered cells while chromogranin and synaptophysin stains were negative (data not shown).

## Discussion

PAs are benign tumors consisting of neoplastic myoepithelial cells, ductal cells, and stroma. The most common location of these tumors is the parotid gland, but they can also emerge in the submandibular, sublingual, and minor salivary glands [1-3]. PAs typically arise in 30 to 60-year-old females and their progression and growth is slow [2].

The prevalence of tracheobronchial PAs is unknown owing to its rarity outside the upper airway [4-6]. There have been only 6 articles that describe PAs in the lower airway of which only 3 were located within the trachea. The most commonly reported symptoms of tracheal pleomorphic adenoma are cough, sputum production, dyspnea, wheezing, and stridor. These symptoms may be confused with asthma, resulting in the delayed diagnosis and subsequent treatment of these tumors, which can lead to airway obstruction [7-10]. The differential diagnosis of a tracheal mass includes neoplasms such as squamous cell carcinoma, the most common primary tracheal malignancy, and adenoid cystic carcinoma. The latter is the second most common tracheal malignancy and is concerning due to its aggressive nature. Compared to PAs, adenoid cystic carcinoma presents sharply separated from the stroma and often with aggressive perineural invasion [4].

The treatment of salivary gland pleomorphic adenoma consists of wide excision with negative margins. In the case of tracheobronchial masses, there is not consensus on the treatment, but it is generally accepted that surgical excision is curative. Endoscopic resection may be performed if there are urgent respiratory symptoms, hemorrhage control is necessary, or in older patients who would not tolerate an open tracheal resection. Regardless of the surgery performed, either endoscopic or tracheal resection, close follow-up is warranted [11].

## Conclusions

Pleomorphic adenomas are benign, slow growing salivary tumors that rarely present in the lower airway. Given their non-specific symptoms, diagnosis and treatment is often delayed and can result in life threatening airway obstruction. Endoscopic excision can be performed in cases of respiratory symptoms or hemorrhage control, though close clinical follow-up is necessary to monitor for recurrence. Additional studies are required to define the clinical course and recurrence rate of lower airway pleomorphic adenomas to guide future management of these benign tumors.

## Contact

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