

Pathology of Primary Tracheobronchial Malignancies Other than Adenoid Cystic Carcinomas



Darroch Moores, MD, FRCSC^{a,*}, Paresh Mane, MD, MCh^b

KEYWORDS

- Tracheal tumors • Pathology • Adenoid cystic carcinoma • Squamous cell carcinoma
- Mucoepidermoid carcinoma

KEY POINTS

- Most primary tracheal tumors are malignant.
- Malignancy of larynx and bronchi are much more likely than trachea.
- Tracheal tumors are most likely due to direct extension for surrounding tumors.
- Squamous cell carcinoma and adenoid cystic carcinoma make up about two-thirds of adult primary tracheal tumors.
- Because of their predominantly local growth pattern, malignant salivary gland-type tumors show a better outcome than other histologic types.

INTRODUCTION

Primary tracheal tumors (PTT) are relatively rare; their diagnosis is often late because the symptoms are noncharacteristic, and the signs only appear when more than 60% of the lumen is obstructed.¹ In adults, approximately 90% of all primary tumors of the trachea are malignant, in contrast to 10% to 30% in children. The incidence of tracheal malignancies is about 0.1 in every 100,000 persons per year, corresponding to approximately 0.2% of all tumors of the respiratory tract and to 0.02% to 0.04% of all malignant tumors. Malignancies of the larynx and bronchi are about 40 and 400 times more likely than cancers of the trachea, respectively.^{2,3}

Malignant involvement of the trachea predominantly results from direct spread of neighboring tumors, whereas primary tracheal malignancies are

usually rare. Compared with the bronchus and lung, their incidence is lower due to the limited surface area epithelium, greater mucociliary stream, and more laminar flow. Normally the trachea is lined by columnar ciliated respiratory epithelium, which has both mucinous and nonmucinous cells. Most of the tracheal neoplasms, both benign and malignant, are squamous rather than mucinous or glandular, thus reflecting a conversion of the epithelium to squamous. This is most likely due to the influence of smoking. The glandular neoplasms are similar to lung and salivary glands. In general, because most of these tumors are obstructing, they are detected early. Squamous cell carcinoma and adenoid cystic carcinoma make up about two-thirds of adult PTT.⁴ A heterogeneous group of benign and malignant tumors accounts for the remaining third of tracheal neoplasms.

A listing of these rarer tumors is as follows.

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^a Department of Surgery, Albany Medical College, 43 New Scotland Avenue, Albany, NY 12208, USA;

^b Department of Surgery, St. Peter's Hospital, 315 New Scotland Avenue, Albany, NY 12208, USA

* Corresponding author. 18 Burton Lane, Albany, NY 12211.

E-mail address: moores.darroch@gmail.com

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Benign

- Squamous papilloma (multiple or solitary)
- Pleomorphic adenoma
- Granular cell tumor
- Fibrous histiocytoma
- Leiomyoma
- Chondroma
- Chondroblastoma
- Schwannoma
- Paraganglioma
- Hemangioendothelioma
- Vascular malformations

Intermediate

- Carcinoid
- Mucoepidermoid
- Plexiform neurofibroma
- Pseudosarcoma
- Malignant fibrous histiocytoma

Malignant

- Adenocarcinoma
- Adenosquamous carcinoma
- Small cell carcinoma
- Atypical carcinoid
- Melanoma
- Chondrosarcoma
- Spindle cell sarcoma
- Rhabdomyosarcoma

SQUAMOUS EPITHELIUM TUMORS

Squamous Cell Carcinoma

Squamous cell carcinoma (Fig. 1) is one of the most frequent PTT, comprising about one-third of all tracheal neoplasms. Several variants of squamous cell carcinoma of the hypopharynx, larynx, and trachea are mentioned, including verrucous carcinoma, basaloid carcinoma, papillary

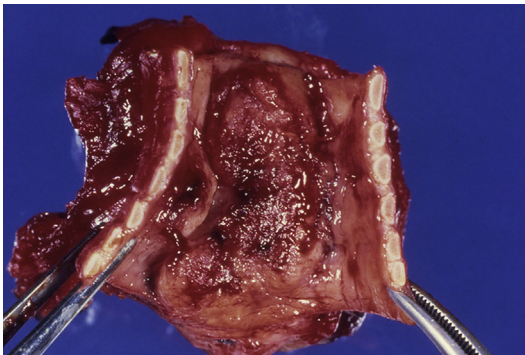


Fig. 1. Extensive squamous cell carcinoma of the lower intrathoracic trachea. Note the typical ulcerated surface of the tumor.

carcinoma, spindle-cell carcinoma, acantholytic carcinoma, and adenosquamous carcinoma. They are much less likely to happen in the trachea as compared with larynx or lung (75 and 180 times, respectively). Smoking is common in most patients. They are more likely in the posterior wall of the lower third. Macroscopically, tracheal squamous cell carcinoma usually grows as a polypoid and frequently ulcerative mass, projecting into the lumen of the trachea. In symptomatic stages, most of these lesions are easily detectable by bronchoscopy. Histologically these tumors are characterized by more or less well-defined squamous differentiation, with or without keratinization.⁵ The association with cigarette smoking may lead to metachronous or synchronous lesions of the oropharynx, the larynx, or the lungs.

Squamous Papilloma and Papillomatosis

Squamous papillomas of the trachea are rare benign tumors composed of stratified squamous epithelium with acantholysis and papillomatosis supported by fibrovascular core. They are multiple and recurrent or solidly exophytic growths into the tracheal lumen. In terms of the biology, squamous papillomas of the trachea are often associated with upper posterior laryngeal and all lower bronchial involvement. The patient presents with stridor, wheezing, dyspnea, chest pain, or hemoptysis. Lung parenchyma is affected in about 1% of cases, most of them complicated by necrosis cavitation or pneumonia. Human papilloma virus is a known cause of this tumor, and different types are detected.⁶ Squamous cell carcinoma may develop in squamous papillomas, although malignancy is rare in papillomatosis.⁷

Adenocarcinoma

Although the most common lung malignancy, adenocarcinomas are usually located in the lung periphery away from the central airways. Infrequently, they can arise as a primary tracheal tumor. When adenocarcinoma does involve the trachea, it is usually secondary to direct extension from an adjacent tumor or malignant mediastinal lymph node. Adenocarcinomas (Fig. 2) affect both sexes mostly in the fifth to eighth decades of life. Presentation is often obstructive. Gross adenocarcinomas are bulky tumors that may bulge into the lumen but may also invade through the tracheal wall into adjacent structures. Microscopically most of these are mucin producing. They form glands with epithelial cells containing large vesicular nuclei and prominent nucleoli. Differential diagnoses include adenoid cystic carcinoma, mucoepidermoid carcinoma, and secondary involvement of trachea from

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