Pathology of Tracheal Tumors

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KEYWORDS

• Tracheal tumors • Pathology • Squamous cell carcinoma • Adenoid cystic carcinoma
• Epithelial precursor lesions

KEY POINTS

- In general, patients with tracheal malignancies show an unfavorable prognosis with reported 5- and 10-year survival rates of 5% to 15% and 6% to 7%, respectively, for all types of tracheal carcinoma.
- The most important prognostic factors in primary malignant diseases of the trachea are early diagnosis, tumor stage, histology, and treatment options.
- Because of their predominantly local growth pattern, malignant salivary gland–type tumors show a better outcome than other histologic types.
- Surgical cure of adenoid cystic carcinoma may be impossible because of its characteristic relentless growth along the perineural sheath.
- Survival in patients with resectable tumors is better than in nonresected patients, especially after histologically complete resection.
- Selection of patients for definitive surgery is the most important factor in improving the prognosis for patients with primary tracheal malignancies.

The trachea extends from the lower border of the cricoid cartilage to the carina, averaging a length of 11 to 12 cm in adults.[1] Malignant involvement of the trachea predominantly results from direct spread of neighboring tumors, whereas primary tracheal malignancies are rarely observed.

EPIDEMIOLOGY

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 frequently observed than cancers of the trachea, respectively.[2,3]

Squamous cell carcinoma and adenoid cystic carcinoma make up about two-thirds of adult primary tracheal tumors. A heterogeneous group of benign and malignant tumors accounts for the remaining third of tracheal neoplasms (see next section).[4]

WORLD HEALTH ORGANIZATION CLASSIFICATION

There is no separate histologic classification for tracheal neoplasms. Primary tumors of the trachea are not listed in the World Health Organization (WHO) classification of tumors of the lung, pleura, thymus, and heart, but are summarized in the WHO classification of head and neck tumors, together with neoplasms of the hypopharynx and larynx. In the histologic classification of tumors of the hypopharynx, larynx, and trachea, malignant epithelial tumors (including several variants of squamous cell carcinoma as well as malignant salivary gland–type tumors), neuroendocrine tumors, benign epithelial tumors, soft-tissue tumors (benign and malignant), hematolymphoid tumors, tumors of bone and cartilage, mucosal malignant melanomas, and secondary tumors are listed.[5] Most of these neoplasms occur in the hypopharynx

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or larynx, whereas only a low incidence is observed in the trachea.

TNM CLASSIFICATION

There is no generally accepted TNM classification for tracheal malignancies.[6] Because of the rareness of these tumors there are only proposed staging systems for primary tracheal carcinomas, which are not based on a large number of considered cases. To date their effectiveness has not been investigated prospectively.[4,7,8] Consequently, the application of a staging system for tracheal malignancies cannot be generally recommended.

Nevertheless, today a TNM classification can be used to allow a standardized description of the extent of tracheal malignancies. For this purpose, Table 1 shows a modification of the staging system proposed by Macchiarini[4] in 2006, adapted to the general rules of the TNM system according to the seventh edition of the TNM classification of malignant tumors.[6]

PREMALIGNANT LESIONS

According to the WHO classification, epithelial precursor lesions are defined as altered epithelium with an increased likelihood for progression to squamous cell carcinoma. The 2005 WHO classification distinguishes between squamous cell hyperplasia, mild dysplasia, moderate dysplasia, severe dysplasia, and carcinoma in situ. In the SIN (squamous intraepithelial neoplasia) concept, mild dysplasia corresponds to SIN1 and moderate dysplasia to SIN2, whereas severe dysplasia and

Table 1
TNM staging system for primary tracheal malignancies

T	Primary Tumor
Tx	Primary tumor cannot be assessed
T0	No evidence of primary tumor
Tis	Carcinoma in situ
T1a	<3 cm, limited to the mucosa
T1b	≥3 cm, limited to the mucosa
T2	Invasion of cartilage or adventitia
T3	Invasion of larynx, carina or main bronchus
T4	Invasion of other neighboring structures
N	Lymph Nodes
Nx	Regional lymph nodes cannot be assessed
N0	No evidence of regional lymph node metastasis
N1	Local lymph node metastasis
Upper third	Highest mediastinal, upper paratracheal, prevascular and retrotracheal lymph nodes
Middle third	Upper paratracheal, prevascular and retrotracheal, lower paratracheal, para-aortic (ascending aorta or phrenic) lymph nodes
Lower third	Upper paratracheal, prevascular and retrotracheal, subaortic (aortopulmonary window) lymph nodes
N1a	1–3 lymph node metastasis
N1b	>3 lymph node metastasis
N2	Regional lymph node metastasis
Upper third	Lower paratracheal, subaortic (aortopulmonary window) lymph nodes
Middle third	Highest mediastinal, subaortic (aortopulmonary window) lymph nodes
Lower third	Upper paratracheal, pulmonary ligament lymph nodes
M	Distant Metastasis
M0	No evidence of distant metastasis
M1	Distant metastasis
M1a	Metastasis to lymph nodes other than N1 and N2
M1b	Distant metastasis

Modified from Macchiarini P. Primary tracheal tumours. Lancet Oncol 2006;7:83–91; with permission.