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The current knowledge of tumors of the trachea, carina, and the main and lobar bronchi, is reviewed, including institutional experience at Massachusetts General Hospital.

Tracheobronchial Gland Tumors

Henning A. Gaissert, MD, and Eugene J. Mark, MD

Background: Tracheal tumors are uncommon, making up only 0.2% of all respiratory malignancies in the United States. One consequence of this low incidence is that few centers accumulate meaningful experience. Another is the lack of awareness of effective therapy. Bronchial gland tumors demonstrate oncologic diversity and include benign, low-grade, and high-grade malignant tumors.

Methods: We reviewed the present knowledge of bronchial gland tumors of the trachea, carina, and bronchi, including the epidemiology, presentation, evaluation, tumor types, and treatment options.

Results: The malignant bronchial gland tumors, adenoid cystic carcinoma and mucoepidermoid carcinoma, are far more common than benign mucinous cystadenoma or pleomorphic adenoma. Complete resection of localized tumors has excellent long-term results in symptomatic benign tumors. The disease-free survival after resection of malignant tumors is limited by distant metastasis and regional disease, while local recurrence is uncommon. Postoperative mediastinal radiation is now accepted adjuvant therapy. Experience at our institute demonstrates a significant survival advantage for patients with complete resection compared to unresectable patients.

Conclusions: Expanding knowledge of diagnostic evaluation and surgical therapy can improve the long-term survival of patients with tracheobronchial gland tumors.

Introduction

Tumors of the seromucous glands are part of the spectrum of tracheobronchial neoplasms. These neoplasms share essential histologic features and clinical behavior with salivary gland tumors of the neck. The collective term *bronchial adenoma* once described all tumors of the large airways that were not lung cancer and alluded in a pars-pro-toto manner to the bronchial glands. This designation has become obsolete as it wrongly conveys histologic homogeneity and a benign disposition for tumors that are overwhelmingly malignant and of which

From the Departments of Surgery (HAG) and Pathology (EJM), Massachusetts General Hospital, Harvard Medical School, Boston, Massachusetts.

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Address correspondence to Henning A. Gaissert, MD, Massachusetts General Hospital, Blake 1570, 55 Fruit Street, Boston, MA 02114. E-mail: hgaissert@partners.org

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Abbreviations used in this paper: MGH = Massachusetts General Hospital, ACC = adenoid cystic carcinoma.

Table 1. — Histology of Primary Tracheal Tumors in Published Studies*

	Epidemiologic Studies			Radiotherapy	Clinical Studies Surgery			
	Manninen et al ² 1991	Gelder et al ⁴ 1993	Licht et al ³ 2001	Chao et al ⁵ 1998	Pearson et al ⁶ 1984	Regnard et al ⁷ 1996	Perelman et al ⁸ 1996	Grillo et al ⁹ 1990
No. of patients	95	321	109	42	44	208	120	198
Tumor type: **								
Adenoid cystic carcinoma	6.3	11	7.3	7.1	64	31	55	40
Squamous cell carcinoma	72	54	68	67	20	45	17	35
Adenocarcinoma	13	4	10	4.8		1.9	0.8	0.5
Large cell carcinoma		5.9	1.8			0	0	0
Small cell carcinoma	7.4	5	7.3	4.8		0	2.5	0.5
Anaplastic carcinoma	2.1		5.5	4.8		0	0	0
Mucoepidermoid carcinoma			0.9			2.4	0.8	2
Other	0	6.5	3.7	0	16	19	23	21
No diagnosis	0	14	0	12	0	0	0	0

* Among bronchial gland tumors, only adenoid cystic and mucoepidermoid carcinomas are common enough to be recognized.
 ** Numbers after tumor type indicate percentage of patients.
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only a minority exhibits glandular features. While glandular architecture is observed in both benign and malignant tumors, neither the most common cause of malignant airway obstruction, metastatic carcinoma, nor the most common primary tumor, squamous cell carcinoma, is associated with bronchial glands. The other common parenchymal lung cancer, adenocarcinoma, is extremely uncommon in the trachea, though it is sometimes observed in series of lobar sleeve resections.

This article summarizes the current knowledge of tracheal and bronchial exocrine gland tumors and their treatment. In our review, we reiterate relevant observations about airway tumors that are not specific to bronchial gland tumors. The invasive and often transmural nature of malignant disease, for example, should guide its treatment and explains why complete resection by segmental tracheal or bronchial resection has produced the best long-term outcome in patients with localized disease. For the purpose of this review, we restrict our comments, in particular on surgical therapy, to tumors of the trachea, carina, and the main and lobar bronchi, and exclude neuroendocrine tumors.

Epidemiology

No dedicated epidemiologic observations of the rare glandular tumors are available, and risk factors are not known. Tracheal tumors in general are uncommon, making up only 0.2% of all respiratory malignancies in the United States¹ and occurring at a rate of 1 per 1 million per year in Finland.² National epidemiologic studies of tracheal carcinoma from Finland and Denmark have demonstrated both disappointingly poor long-term survival and exceedingly low rates of surgical resection (less than 10%).^{2,3} The Danish investigators

reasoned that a lack of treatment led to poor outcome and criticized a nihilistic attitude among physicians toward surgical treatment. The most common tracheal gland tumor, adenoid cystic carcinoma (ACC), is no exception, although the slow progression of disease, in contrast to bronchogenic carcinoma, permits prolonged survival in many patients, even with palliative therapy alone. Epidemiologic and clinical series report considerable differences in the histologic composition of airway tumors, as demonstrated for tracheal tumors in Table 1.^{2,9} These differences reflect treatment selection and variations in resectability between benign, low-grade, and highly malignant lesions. The proportion of bronchial gland tumors in the trachea is far higher than in the bronchi due to the preferential location of the most common glandular tumor, ACC. For example, approximately 40% of 360 primary tracheal tumors,¹⁰ but less than 15% of primary bronchial tumors,¹¹ at Massachusetts General Hospital (MGH) derived from airway glands (Table 2).

Table 2. — Salivary Gland Tumors Among 360 Primary Tracheal (1962–2002) and 78 Primary Low-Grade Malignant and Benign Bronchial Tumors (1972–1995) at MGH

Location	Type	No. of Patients
Trachea	Adenoid cystic carcinoma	135
	Mucoepidermoid carcinoma	14
	Pleomorphic adenoma	3
Bronchi	Adenoid cystic carcinoma	2
	Mucoepidermoid carcinoma	7
	Polypoid glandular neoplasm	1
	Mucous gland cystadenoma	2
Total		164
Data from references 10, 12, 13.		

Presentation

Tumors located in the trachea or the main bronchus cause progressive airway obstruction with variable time course and nonspecific symptoms. Cough, shortness of breath, or wheezing often lead to treatment for chronic obstructive lung disease, and patients may receive oral corticosteroids. As a result, the diagnosis is usually delayed by observation of presumed asthma or obstructive lung disease unless hemoptysis, due to tumor growth or mucosal erosion, prompts bronchoscopic evaluation. Lobar or main bronchial obstruction can lead to unilateral wheezing, which may become obvious to the careful examiner. As shown in Table 3, the tumor type influences the duration of symptoms. Benign tracheal tumors and ACC have the longest mean symptom duration of 24 months.^{10,12} Symptoms lasted for less than 4 months in 3 patients at MGH with pleomorphic adenoma, 2 having hemoptysis and 1 having stridor. In patients with mucoepidermoid carcinoma, symptoms lasted for 11 months.¹² As explained below, due in particular to the slow growth of ACC, the duration of symptoms alone predicts neither malignant potential nor resectability of a tumor.

Evaluation

Imaging studies usually precede bronchoscopy, unless the patient arrives in respiratory distress. The radiographic evaluation of airway tumors has advanced from traditional to sophisticated techniques. The purpose of newer modalities is a more precise assessment of tumor length and depth of invasion. Whether these advantages influence clinical outcome has not been determined. Chest radiographs may demonstrate collapse of a lobe or an entire lung due to complete obstruction or to hyperinflation of the lung peripheral to a bronchial tumor that causes a ball valve effect. Standard tomography, depicting the air shadow of the trachea and the approximate length of tumor-related narrowing,¹³ has been replaced by computed tomography (CT) with multiplanar reconstruction. CT of the chest may demonstrate a tracheal or bronchial mass but underestimates the extent of submucosal invasion. Newer software algorithms allow three-dimensional

and luminal evaluation by CT (virtual bronchoscopy) but lack the detail of real bronchoscopy and do not reliably determine tumor length. Invasion of the great vessels, however, can be identified.¹⁴ Magnetic resonance imaging (MRI) has been reported in ACC¹⁵; however, its perceived advantage of multiplanar reconstruction may be less apparent now that CT permits imaging along similar planes. MRI may have a role in individual patients, either when CT is unavailable or when radiation must be avoided. Endoluminal ultrasound of the trachea may distinguish between compression and infiltration of the trachea by an extrinsic tumor,¹⁶ but detection of submucosal infiltration in the presence of a normal mucosal surface, by far the most common cause of positive tracheal resection margins in ACC, has so far not been described. Optical coherence tomography has depicted microstructures such as mucosa, cartilage, and glands¹⁷ and may provide sufficient anatomic detail of light-microscopic quality to guide surgical therapy in the future. Despite the present shortcomings of locoregional staging, only 17 (8.2%) of 208 patients with primary squamous or adenoid cystic tracheal carcinoma underwent operative exploration without resection at MGH.¹⁰ Patients with malignant tumors undergo a search for metastatic disease that typically includes examinations of the lung, brain, bone, adrenal glands, and liver.

Every patient suspected of having a tracheal tumor should undergo endoscopic examination of the airway by an endoscopist experienced in rigid bronchoscopy and in judging resectability, no matter how debilitated, seemingly unresectable, or highly obstructed. When performed with care, endoscopy is well tolerated, provides a diagnosis, and may be used for dilatation of a stricture or for removal of a tumor to relieve symptoms. The procedure requires close cooperation with an anesthesiologist trained in the management of airway obstruction and is preferably conducted under general anesthesia with rigid endoscopes. Biopsies are obtained if a carcinoma is suspected or the tumor is extensive. Additional mucosal biopsies beyond gross tumor are occasionally important to determine the length of tumor. Laryngoscopy precedes bronchoscopy if the tumor involves the subglottic airway or vocal cord dysfunction is known.

Tumor Types

The minor salivary glands are located in the submucosa of trachea and bronchus where they are evenly distributed throughout the trachea in humans.¹⁸ Morphologically, these glands resemble the major salivary glands of the neck and those in the oropharyngeal mucosa, and they are subject to salivary gland-like tumors, though at a much lower frequency than those in the neck. There

Table 3. — Symptom Duration in Some Bronchial Gland Tumors

Tumor	Months
Adenoid cystic carcinoma, resectable	18.3
Adenoid cystic carcinoma, unresectable	23.7
Mucoepidermoid carcinoma	11
All benign tumors	20
Data from references 10, 12.	

are four major types, of which only one, mucous gland adenoma, is known exclusively in its benign form. Adenocarcinoma has been excluded from this review as virtually all adenocarcinomas arise from the pulmonary parenchyma. True origin from the rare tracheo-bronchial glands may occur but at such low incidence as to preclude any specific statements. Studies that report a higher incidence of tracheal adenocarcinoma (Table 1) do not provide pathologic review and should be suspected of including metastatic lesions with secondary tracheal invasion.

Mucous Gland Adenoma (Mucinous Cystadenoma)

Biology: Spencer¹⁹ reported 5 of these tumors, all from main or lobar bronchi. Since then, more than 10 additional bronchial and 3 tracheal tumors have been reported.²⁰⁻²² Cystically, dilated acini are a characteristic finding and have led to the alternative term *mucous gland cystadenoma*. Low-grade mucoepidermoid carcinoma is the main diagnostic alternative, but less differentiated cells usually give this carcinoma away.

Pathology: These rare tumors are spherical, smooth, and sometimes pedunculated. The few case reports describe them in the main or lobar bronchus, though they have also been found in the lung and the trachea. The tumor is composed of mucus-containing acini lined with cuboidal cells in a single layer with a pale, foamy cytoplasm.²¹ The epithelial lining may show stratification or papillary infolding.²³ Nuclei have an open chromatin pattern and a prominent nucleolus. The stroma is composed of hyaline connective tissue. The tumor is well demarcated (Fig 1) but has no capsule. Therefore, neoplastic acini merge with those of the normal bronchial mucous gland. There are no mitotic figures, and a malignant variant has not been reported.

Pleomorphic Adenoma

Biology: This tumor arises commonly in the major salivary glands — the parotid and submandibular gland,

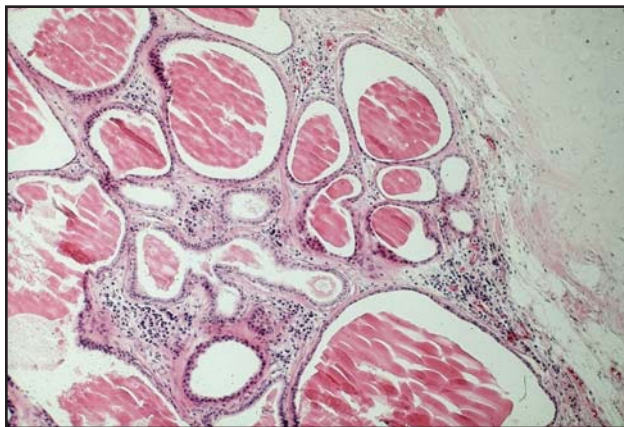


Fig 1. — Low power view of bronchial mucinous cystadenoma elevating intact epithelium (periodic acid-Schiff).

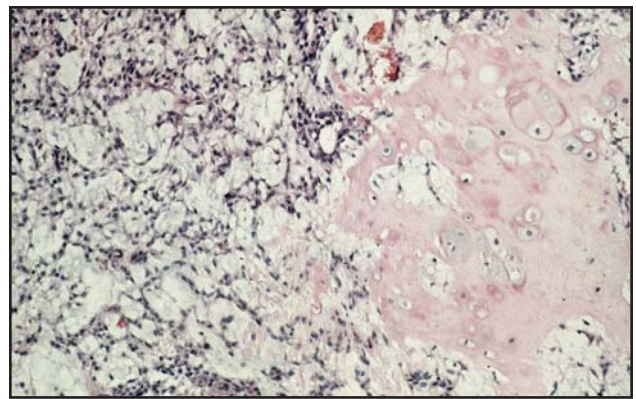


Fig 2. — Pleomorphic adenoma demonstrating epithelioid component (left) and mesenchymal and chondroid component (right) with distinct interface.

for example — and is rare in the trachea and bronchi. Most pleomorphic salivary tumors are benign, though late recurrence of malignant pleomorphic adenoma was reported, with metastasis in lung and chest wall arising 11 years after resection.²⁴

Pathology: The term pleomorphic reflects the capacity for display of both epithelial and mesenchymal differentiation and accounts for the alternative name of mixed tumor (Fig 2). The epithelial elements are variable in discreteness and include glandular or trabecular or solid patterns. Ductular structures may contain secretory material. The mesenchymal elements are typically chondroid but can be myxoid or collagenous. Usually these are transitions between epithelioid and mesenchymal elements. Necrosis and malignant-appearing nuclei and high mitotic rate are features of potential malignancy but are uncommon findings. When a carcinoma does arise in pleomorphic adenoma, it is described as carcinoma ex pleomorphic adenoma. Some examples of pleomorphic adenoma have only epithelioid or mesenchymal tissue. Such cases pose a particular problem in histologic diagnosis because the characteristic bimorphic nature of the tumor is not present. These examples are termed monomorphic adenomas. Prognosis is not influenced by the relative proportions of epithelial and mesenchymal elements, but the histologic differential diagnosis is influenced depending on the specific findings.

The differential diagnosis includes cellular and solid variants of ACC, low-grade mucoepidermoid carcinoma, chondroma, bronchial hamartoma, low-grade adenocarcinoma of salivary gland type, and epithelial-myoepithelial carcinoma.

Mucoepidermoid Carcinoma

Biology: This tumor arises from submucosal glands and is exclusively malignant, occurring in low- and high-grade variants. Low-grade variants may grow as well-demarcated or pedunculated lesions (Fig 3). Heitmiller et al²⁵ reported 18 mucoepidermoid tumors, 3 of which were in the trachea. Three patients had

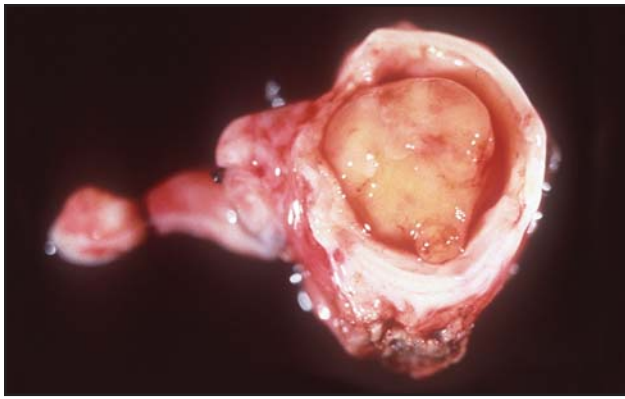


Fig 3. — Mucoepidermoid tumor protruding into bronchus with ball-valve effect.

high-grade tumors, all resulting in death due to disease progression, while there were no recurrences after complete resection for low-grade lesions. A series from Taiwan found high-grade tumors in 8 of 11 patients with a 5-year survival rate of only 25%.²⁶

Pathology: Salivary glands and salivary gland tumors have both a squamoid component lining ducts and a mucinous component lining glands, so mucoepidermoid carcinoma reflects differentiation along both lines. Mucoepidermoid carcinomas can be considered in terms of the relative squamoid or mucinous component as well as in the degree of malignancy. The squamoid component may be epidermoid with prominent eosinophilic cytoplasm and intercellular bridges, and in these regions the differential diagnosis on a biopsy will include squamous cell carcinoma. The mucinous component may be cuboidal cells with basophilic cytoplasm and small rudimentary glands (Fig 4), and the differential diagnosis will include adenocarcinoma.

Cells intermediate between squamous and mucinous cells, commonly referred to as transitional cells, often exceed the clearly squamoid or clearly glandular components. These tumors produce a greater differential diagnosis, particularly on a small biopsy, where metastases as well as solid forms of other salivary gland tumors can arise.

The tumors are conventionally graded into either two grades (low-grade or high-grade) or three grades (well differentiated, moderately differentiated or poorly differentiated), depending on the grading system. Increasing grade correlates with poorer prognosis.

A peculiar variant of mucoepidermoid granulomas produces cysts, often with scant and attenuated epithelium. Because of the cystic rather than solid nature of such a lesion, the differential diagnosis may include mucus retention or mucocele or benign cyst. If both squamous and glandular components have malignant features, the differential diagnosis would include adenosquamous carcinoma. Mucoepidermoid tumors typically are centrally located and adenosquamous carcinomas typically are in the parenchyma of the lung.

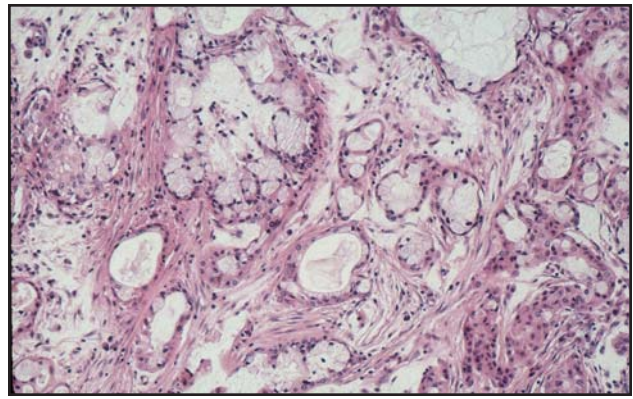


Fig 4. — Glandular component of mucoepidermoid carcinoma with well-formed glands.

Adenoid Cystic Carcinoma

Biology: This salivary gland tumor is a slowly progressive malignancy, with late metastasis to lung, bone, and brain. The most common site in the airway is the trachea, and bronchial lesions are uncommon. Although equal numbers of the two most frequent primary airway tumors in clinical series suggest that ACC is as common as squamous cell carcinoma, the latter probably has a higher incidence due to its more rapid progression.

ACC occurs between the second and ninth decade of life. The mean incidence is in the fifth decade, 10 years earlier than in squamous cell carcinoma. The sex distribution is nearly even. As a reflection of slow progression, patients may present after years of gradually worsening symptoms of airway obstruction (Fig 5) resembling benign tumors in average symptom duration.

Pathology: ACC typically has long, cylindrical structures lined by small cuboidal cells with deeply eosinophilic cores of basement-membrane-like material. The cylinders account for the alternative name of cylindroma. The eosinophilic cores will stain with periodic



Fig 5. — ACC eroding through bronchial wall and causing luminal obstruction.

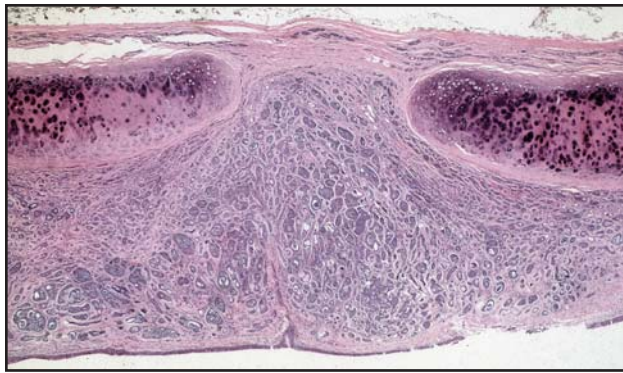


Fig 6. — Cross-section of bronchus demonstrating adenoid cystic tumor filling the lamina propria and forming a tongue invading between two plates of cartilage.

acid-Schiff. Mucinous material is also produced. Relatively solid nests of tumor often have a cribriform pattern. Individually infiltrating cells often occur. Although flagrant cytologic features of malignancy do not occur, the malignant behavior of the tumor is manifest by its insidious growth pattern, with nests of cells exceeding grossly suspicious areas and nests of cells growing within a preexistent template of mucinous glands within bronchial or tracheal mucosa (Fig 6). Growth along nerve sheaths may be extensive (Fig 7). The differential diagnosis most commonly includes mucoepidermoid carcinoma and, on a small biopsy, carcinoid tumor.

Treatment

Local treatment options consist of endoscopic resection, segmental airway resection, and radiotherapy. Of these techniques, only segmental tracheal or bronchial resection permits both complete removal of the tumor and pathologic confirmation of completeness of resection.

Bronchoscopic Resection

Endoscopic techniques are attractive as a less invasive approach. However, benign salivary gland tumors originate in the submucosal plane and may extend between cartilages. Malignant tumors invade into and beyond the airway wall. Successful treatment of luminal tumor by laser or forceps extraction may restore the airway lumen but does not provide confirmation of resection margins and can leave tumor behind. The primary role of bronchoscopic resection is therefore palliative (in patients who are not candidates for surgical procedures) or temporizing (to reopen the airway for acute obstruction prior to definitive therapy). The published reports of laser resection raise but do not answer the question of long-term outcome.²⁷⁻³³ Complete endoscopic resection would hypothetically require full-thickness resection of intramural and transmural tumors and is therefore not feasible in most instances.

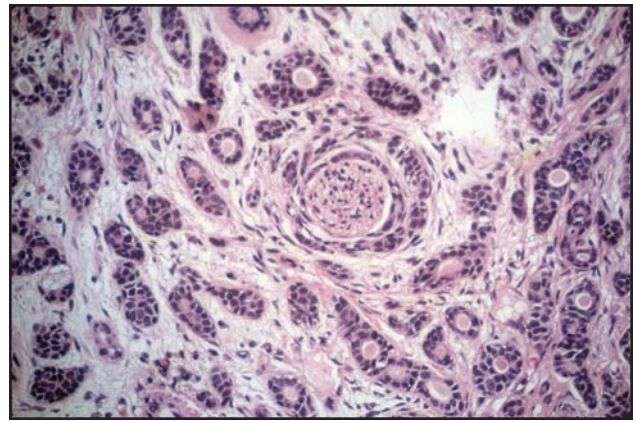


Fig 7. — ACC with small glands and perineural invasion.

Surgical Therapy

The estimation of tumor size, length, and depth influences the assessment of resectability. These parameters, in turn, are related to airway dimensions and individual growth characteristics of the tumor. The location of tracheal tumors in the mediastinum preempts conventional radical resection with wide margins. The maximal resectable tumor length in the trachea is also constrained by tension at the anastomosis. ACCs often involve a longer tracheal segment before airway obstruction occurs; these tumors expand by infiltrative growth along the airway and are thus more likely to involve the peritracheal planes or lymph nodes. In contrast, benign seromucous gland tumors grow by expansion and involve a short length of bronchus or trachea; their polypoid growth is more likely to obstruct the lumen.

Airway tumors may appear unresectable because of their central location. Compared to tumors in the periphery of the lung, however, the lumen size of large airways favors earlier detection. The diameter of the adult trachea measures between 1.4 and 1.75 cm,³⁴ while the average left main bronchus has a diameter of 1.18 to 1.3 cm.³⁵ A neoplasm expanding as a sphere may therefore be expected to cause symptoms at a diameter of less than 2 cm. At this size, even malignant tumors have progressed less commonly to invasion into adjacent vital structures or to regional lymph nodes. The average length of most resected tracheal tumors measures less than 3 cm, with the exception of ACC (Table 4).

Table 4. — Mean Length of the Two Most Common Tracheal Gland Tumors Compared to Other Tracheal Tumors Treated With Resection

Tumor Type	Length (cm)
Mucoepidermoid carcinoma	1.5
Adenoid cystic carcinoma	3.2
All benign	2.2
Squamous cell carcinoma	2.4
Data from references 10, 12.	

The resectability of tracheobronchial tumors is further determined by patient-related factors. Old age, short neck, stocky body habitus, and thoracic kyphosis all limit resectable tracheal length, while young age, thin body habitus, and long neck favor resection of even long tumors. The longest tracheal tumor resected successfully at MGH with primary tracheal anastomosis measured 6.5 cm (unpublished observation). The critical distance in carinal involvement refers to the combined length of resected trachea and main bronchus; a gap of more than 4-cm long between trachea and left main bronchus is associated with excessive tension and may prevent primary anastomosis. Even a surgical connection of the left main bronchus to the right bronchial tree to the bronchus intermedius, for example, would be associated with excessive tension and a greater risk of dehiscence. If the main carina is preserved, the entire length of either mainstem bronchus may be resected, with reconstruction between main and lobar carina.

The classic concept of complete resection is violated in the mediastinum due to the immediate vicinity of vital structures and the limited length of the airway. In recognition of close surgical margins in malignant tumors and after early anecdotal observation of local recurrence, adjuvant radiotherapy is now standard therapy in ACC and other high-grade bronchial gland tumors. Since tracheal anastomoses heal under tension (up to 1 kg), bronchoscopic assessment of the anastomosis is important to ascertain healing before proceeding with radiation, usually 2 months after the operation. There are no studies examining dose or field variations due to the rarity of these tumors.

A defined set of operative procedures has evolved. For tracheal tumors, these are laryngotracheal, tracheal, and carinal resection; for bronchial tumors, they are main bronchial resections with parenchymal sparing and sleeve lobectomy. The concept of surgical reconstruction of the tracheobronchial tree originated in 1947 with resection of the main bronchus³⁶ and sleeve lobectomy shortly thereafter.³⁷ The technical principle common to these procedures is segmental airway resection — the excision of a length of bronchus or trachea with end-to-end restoration of airway continuity by primary anastomosis. Due to its greater reliability and uncompromising patency, segmental resection superseded earlier surgical attempts at lateral, window, or wedge resection of trachea and bronchus. We comment on the different types of resection, with particular emphasis on ACC because of the larger clinical experience with this tumor.

Tracheal Resection

Primary tumors in the subglottic larynx and those extending above the cricoid from the cervical trachea can be removed by laryngotracheal resection. The operation consists of immediate reconstruction of the subglottic airway with stable tracheal wall, thereby avoiding tracheostomy

or tracheal stents in most patients. Close tumor margins are accepted in an attempt to preserve the larynx and vocal cord function. The extent of resection is tailored to the particular lesion, creating a beveled anastomosis of larynx and trachea and accepting sacrifice of one recurrent nerve if involved by tumor as long as the contralateral vocal cord retains function. In malignant tumors, resection is followed by postoperative adjuvant radiotherapy. Using this approach, the observed rate of local recurrence is low, less than 5%,^{38,39} and laryngectomy for local disease is not required. Of 25 patients with primary tumors at MGH, 11 had tracheal gland tumors: ACC in 9 and mucoepidermoid carcinoma in 2.³⁹

Tracheal resection with end-to-end anastomosis was performed in 29 of 38 patients with ACC of the upper airway at Toronto General Hospital⁴⁰ and in 44 of 101 resections for the same diagnosis at MGH.¹⁰ Because of infiltrative growth, 59% of patients at MGH had positive airway margins after tracheal or carinal resection. This circumstance meant that the surgeon had removed the maximal amount of airway and that further resection would have placed excessive tension on the anastomosis. In most cases, tracheal margins were positive due to microscopic rather than gross disease. Why pursue resection when microscopically incomplete resection is so common? While negative margins were associated with better long-term survival, resection and reconstruction with tumor-bearing airway margins provided a survival advantage compared to unresectable tumors.¹⁰ ACC is radiosensitive, and most patients with transmural tumors have undergone postoperative radiotherapy. Tracheal resection is also suitable for any of the other tracheal gland tumors that are usually of limited length.^{7,12,41} The operative mortality for this procedure is below 5% at centers with experience in these tumors.^{10,40}

Resection of the carina is the most complex bronchoplastic procedure. In the total experience of carinal resection, salivary gland tumors represent a minority. In a report by de Perrot et al,⁴² 12 of 119 carinal tumors (8 adenoid cystic, 4 mucoepidermoid) were salivary gland tumors, and Mitchell et al⁴³ found 44 of these tumors (37 adenoid cystic, 7 mucoepidermoid) among 118 carinal resections for tumor. The greater complexity is explained by the need for two bronchial anastomoses, the greater susceptibility of the bronchus to anastomotic tension, and, when carinal resection and pneumonectomy are combined, by the greater physiologic insult of the operation. The operative mortality reaches 16% in series that included early surgical results of this procedure but has fallen below 10% in the recent experience.^{10,40}

Bronchial Resection

Sleeve lobectomy allows resection of a tumor that involves the lobar bronchus and has grown into the mainstem bronchus, while preserving the remaining

lobes of the ipsilateral lung. By avoiding pneumonectomy, sleeve lobectomy lowers the operative risk close to that of standard lobectomy and preserves postoperative exercise capacity. This operation is often the only surgical alternative in patients who would not tolerate pneumonectomy because of poor cardiopulmonary reserve; however, salivary gland tumors usually occur in younger patients not impaired by emphysema associated with long-term smoking. The technique may be applied to any pulmonary lobe, but resections of the right upper lobe are most common.^{11,44,45}

The technique of bronchial sleeve resection is used to remove tumors located in the mainstem bronchus or bronchus intermedius, while resection of lung parenchyma is altogether avoided. The main advantage consists of evading pneumonectomy in the case of mainstem lesions or resection of the middle and lower lobes in the case of bronchus intermedius tumors. Anastomotic tension is reduced by hilar mobilization.

Unresectable Disease

In the MGH experience, 25% of patients with ACC had unresectable disease. Tumor length as determined by bronchoscopy in 68% of these patients was the most common reason resection was declined. Regional disease precluded resection in 23%, whereas distant metastatic disease at presentation occurred in only 6%.¹⁰ The goals of treatment in unresectable malignant tumors are to restore a patent airway and to slow progression of disease. Effective local therapy other than resection may provide meaningful palliation, either by regional radiation or bronchoscopic destruction. Mediastinal radiation with doses ranging from 54 to 60 Gy is administered as curative therapy to patients with good performance status. The role of preoperative, neoadjuvant radiotherapy has not been explored. The reluctance to use preoperative radiotherapy in tracheal tumors is based on two reasons. Every tracheal resection results in tension at the anastomosis, and longer, unresectable tumors are not resected because of excessive tension. If resection was pursued after radiation of tracheal tumors, the problem of excessive tension would be potentiated by delayed healing of radiated tissue. There is no defined role for chemotherapy in glandular tumors. Local destruction of tumor alone may be accomplished by methods other than bronchoscopic coring-out: the laser, cryosurgery, brachytherapy, photodynamic therapy, or argon beam coagulation. Malignant strictures may be stented using T tubes^{46,47} or a tracheostomy tube. Because self-expanding stents are almost impossible to remove once placed, life expectancy should be limited to a few months at the time of insertion so that secondary complications of stenting do not have an impact on quality of life.

Results

In these uncommon tumors, long-term results of therapy range from the representative in ACC to the anecdotal in pleomorphic adenoma. Benign bronchial gland tumors have an excellent long-term prognosis and no recurrences were reported when the resection was complete. Reports of endoscopic resection for tracheal tumors have not provided long-term follow-up.^{48,49}

The disease-free survival after resection of malignant tumors is limited by distant metastasis and regional disease, while local recurrence is uncommon. Among 18 mucoepidermoid tumors of the lung, most in trachea and large bronchi, none of 15 low-grade tumors, but all 3 high-grade carcinomas, proved fatal.²⁵ Perelman et al⁸ reported the survival rate of 66 ACCs at 5 years as 35.9% and at 10 years as 27.1%. In reporting their experience with ACCs, Maziak et al⁴⁰ of the University of Toronto noted an actuarial survival rate of 79% at 5 and 51% at 10 years in 32 patients treated with primary resection and adjuvant radiotherapy. There was a trend toward longer survival, but no significant difference, between patients undergoing complete vs incomplete resection. Six of their patients with ACC treated with radiotherapy alone had a mean survival of 6.2 years. The experience at MGH demonstrates a significant survival advantage for patients with complete resection compared to unresectable patients¹⁰; long-term survival beyond 13 years was observed after incomplete resection but not in unresectable patients. The 5-year survival rate was 52.4% for resected and 33.3% for unresectable ACCs. A collective French study found 5- and 10-year survival rates of 73% and 57%, respectively, in ACC, with longer survival after complete compared to incomplete resection.⁷

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