systemic disease, the parenchyma sparing is advantageous from a prognostic view, especially because a function restriction of the lung in the progressive course is conceivable.

We described a patient and his surgical treatment, in which we used the lifesaving parenchyma-sparing operation. We believe that replacement of the pulmonary artery for aneurysms of the left pulmonary artery, as well as for aneurysms of the right pulmonary artery, in the intraparenchymal and intersegmental portion over the fissure of the lung is possible. This operation must be performed during extracorporeal circulation because there is excessive back bleeding from the bronchial arteries. To our knowledge, we present the first successful interposition of the left pulmonary artery with implantation of the segmental arteries as a lung parenchyma-sparing procedure for pulmonary aneurysms.

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Chondrosarcoma arising in the trachea: A case report and review of the literature

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PPrimary malignant tracheal tumors are rare, accounting for only 0.2% of all malignancies of the respiratory tract. We present a case of tracheal chondrosarcoma in a 78-year-old man and review the literature.

Clinical Summary

A 78-year-old white man had a 3-month history of progressive shortness of breath with exertion. He did not have fever, cough, chills, night sweats, or hemoptysis. His primary care physician initially attributed his symptoms to worsening chronic obstructive pulmonary disease and treated him with inhalers and oral steroids. As his symptoms progressed, the patient became increasingly

bedridden, with dyspnea at rest, and he developed significant weight loss. He also began to experience episodic hoarseness. This symptom progression prompted further evaluation and he had a chest radiograph and then computed tomography (CT), which revealed a noncalcified mass in the lower third of the trachea with near complete obstruction of the lumen. An extratracheal extension of the mass was also present. The patient was transferred to our institution.

The patient was taken to the operating room where flexible and rigid bronchoscopy demonstrated a firm, white, nonfriable endotracheal mass emanating from the right side of the membranous trachea and obstructing 80% of the tracheal lumen. Endotracheal resection was performed via the rigid bronchoscope to provide an adequate airway. A postoperative CT scan confirmed the improved airway (Figure 1). Pathologic examination of the surgical specimen revealed a type I chondrosarcoma. The patient recovered uneventfully and was sent home for 3 weeks of intensive physical and nutritional therapy. He then underwent definitive resection.

The surgical approach utilized a right posterolateral thoracotomy, and the thorax was entered through the fourth intercostal space. A fourth intercostal muscle flap was harvested. The trachea was exposed anteriorly and posteriorly, and the mass was circumferentially dissected, with 5-mm margins proximally and distally. The tumor was well encapsulated, and both the right vagus and the left recurrent laryngeal nerve were identified and spared. A right

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Figure 1. Computed tomography images demonstrating the tracheal lesion at presentation (A) and after endoscopic resection of the endoluminal disease (B).

TABLE 1. Characteristics of 15 reported cases with tracheal chondrosarcoma

First author (year)	Patient age, years/sex	Symptoms	Calcification	Duration of symptoms to Dx (months)	Treatment	No. of recurrences	Follow-up (years)	
Jackson (1959)	32/M	Cough	NR	72	Endoscopic resection	0	6	NED
Daniels (1997)	73/M	Dyspnea, cough, wheeze	NR	7	Endoscopic resection	5	3	AWD
Farrell (1998)	87/M	Dyspnea	NR	12	Refused resection endoscopic resection 2, radiotherapy	2	1	AWD
Salminen (1990)	57/M	Dyspnea, cough	Absent	3	Incomplete resection	5	14	DOD
Matsuo (1988)	72/M	Dyspnea, hemoptysis	Absent	7	Incomplete resection, laser resection, resection	1	0.5	NED
Moersch (1954)	NR	NR	NR	NR	Resection	NR	NR	NR
Arevalo (1986)	74/M	Dyspnea, pneumonia	Present	0.25	Resection	0	1	NED
Fallahnejad (1973)	48/F	Dyspnea, cough, wheeze	Absent	16	Resection	0	5	NED
Leach (1994)	72/M	Dyspnea	Present	36	Resection	NR	NR	NR
Kaneda (1993)	64/M	Dyspnea	NR	NR	Resection	0	1	NED
Slasky (1985)	58/M	Dyspnea	Present	24	Resection	0	2.5	NED
Weber (1978)	71/M	Dyspnea, cough, hoarse	Present	4	Resection	0	5	NED
Mine (1990)	74/M	Dyspnea, cough	NR	NR	Laser debulking/resection	0	2.8	NED
Kiriyama (1997)	54/M	Dyspnea, wheeze	Present	1	Laser debulking/resection	0	3.5	NED
Maish (2003)	78/M	Dyspnea	Absent	3	Laser debulking/resection	0	0.5	NED

Dx, Diagnosis; M, male; NR, not reported; NED, no evidence of disease; AWD, alive with disease; DOD, dead of disease; F, female.

hilar release was performed, and the trachea was then divided at approximately 1 tracheal ring above and below the tumor. Frozen sections confirmed microscopically negative margins. The anasto-

mosis was performed using interrupted 3-0 and 4-0 polydioxanone sutures for the cartilaginous and membranous portions of the trachea, respectively. Because the patient was elderly and had been

treated with steroids, the fourth intercostal muscle flap was used to cover the anastomosis. Prior to extubation in the operating room, a bronchoscopy was performed, demonstrating a patent anastomosis. The patient had an unremarkable recovery and was discharged 7 days after surgery.

Discussion

Squamous cell and adenoid cystic carcinoma account for more than 80% of malignant tracheal tumors in adults.¹ Squamous cell carcinomas seem to be more common in Europe, as indicated by a comparison of the study by Grillo and Mathisen² with that of Gelder and Hetzel.³ The next most common tracheal malignancies, in order of decreasing incidence, are carcinoid tumor, adenocarcinoma, and small cell carcinoma. The remaining tracheal malignancies are very rare and include chondroma, chondrosarcoma, leiomyosarcoma, fibrosarcoma, chemodectoma, hemangiopericytoma, spindle cell sarcoma, synovial cell sarcoma, and melanoma of the trachea.

A review of the literature revealed only 14 reported cases of tracheal chondrosarcoma.^{1,4-15} Patient and clinical characteristics for those 14 patients and our patients are summarized in Table 1. The mean age of all 15 patients at presentation was 65 years (range 32-87 years). Fourteen of the 15 patients were men. The reported symptoms were dyspnea (n = 13; 93%), cough (n = 6; 43%), wheezing (n = 3; 21%), hemoptysis (n = 1; 7%), hoarseness (n = 1; 7%), and pneumonia (n = 1; 7%) and were present for a mean of 15 months (range 1 week to 6 years) prior to diagnosis. The frequent combination of dyspnea, cough, and wheezing without hemoptysis often leads to a misdiagnosis of the condition as asthma or an exacerbation of chronic obstructive pulmonary disease, as had occurred with our patient.

The initial evaluation of the symptoms commonly involves chest radiography. Infrequently, an hourglass-shaped tracheal stripe may be noted, or if significant air has been trapped distal to the obstructing mass, the lung fields may be overinflated bilaterally. Frequently, however, chest radiographs are normal. CT provides the best visualization of the lesion, permitting accurate localization of the lesion and providing information about tumor length and extratracheal extension. Our review of the literature identified 5 cases, with various degrees of calcification, whereas 4 cases, including ours, had no evidence of calcification. The majority of cases also reported extratracheal extension of the mass. Magnetic resonance imaging has no added value compared with CT.

Bronchoscopy is the best method for the diagnosis and initial treatment of tracheal obstruction. When the airway is severely compromised, the patient should undergo a conscious evaluation in the operating room where the airway can be quickly controlled if necessary. Both flexible and rigid bronchoscopy should be utilized to evaluate the movement of the vocal cords, to identify the proximal and distal extent of the lesion, and to perform biopsies. Concomitant endoscopic resection and laser ablation can be performed to restore an adequate airway. Care must be taken not to dislodge a portion of the mass into a main stem bronchus. Once lodged distally, these masses can be difficult to retrieve and will embolize into the distal airway. Tattooing the airway with India ink at the most proximal and distal extents of the tumor can aid in defining the margins for surgical resection, especially if the tumor

Figure 2. Cross section of resected specimen. The encapsulated nature of the tumor is readily apparent.

is initially removed by bronchoscopy and is primarily endoluminal.

Grossly, tracheal chondrosarcoma is well circumscribed and commonly covered with normal mucosa. It has been uniformly described as a firm or hard smooth mass and is commonly white or tan (Figure 2). Histopathologically, tracheal chondrosarcoma closely resembles the classic chondrosarcoma arising in bone rather than the mesenchymal chondrosarcoma variant that occurs in bone or soft tissue.¹⁶ The cytologic findings include round or polygonal cells with occasional binucleation, round hyperchromatic nuclei, and prominent nucleoli, all present in an amorphous pink-violet or light-blue background containing fragments of chondroid tissue. Distinguishing tracheal chondrosarcoma from benign chondroma can be difficult; however, low-grade chondrosarcomas and chondromas have similar natural histories and should be approached in similar manners. As with bony chondrosarcomas, the malignant transformation of a chondroma or a low-grade chondrosarcoma to a high-grade (ie, “dedifferentiated”) chondrosarcoma with hematogenous metastasis has been reported.⁷

Complete surgical resection of the tumor, with primary reconstruction of the trachea, is the optimal treatment. Of the 15 patients reported in the literature (including the patient in our case), 5 underwent either incomplete surgical resection (positive surgical margins or a “debulking” procedure) or endoscopic resection only as the initial definitive treatment. The remaining 10 patients underwent complete surgical resection (removal of all gross disease with negative surgical margins) with or without initial laser debulking to restore a patent airway. Follow-up in our case and in 13 of the 14 reported cases, including all cases of complete resection, ranged from 3 months to 14 years (follow-up duration was not reported in the remaining case). Overall, there were 13 recurrences in 4 patients, all of whom had undergone incomplete resection or endoscopic resection. None of the patients who underwent complete surgical resection had a recurrence.

Tumors arising in the lower third of the trachea are best approached through a right thoracotomy, whereas tumors in the middle and upper third can be reached with a collar incision. Every effort should be made to achieve negative microscopic margins while still allowing reconstruction with an end-to-end anastomosis. Although up to half of the trachea can be resected in younger patients, elderly patients have less tracheal mobility and must be approached more conservatively. Coverage of the tracheal anastomosis with a muscle flap or omentum is recommended, especially in deconditioned elderly patients and patients who have received steroids.

Chondrosarcoma, especially a lower-grade tumor, is not responsive to chemotherapy. Although chondrosarcoma has been considered radioresistant, there are reports of chondrosarcomas arising in bone being successfully treated with radiotherapy.¹⁶ In addition, 1 report of the use of radiotherapy to treat laryngeal chondrosarcomas also exists.¹⁷ Therefore, although it is not considered to be a first-line treatment, radiotherapy may have a role in cases of surgically unresectable chondrosarcomas.

In conclusion, tracheal chondrosarcomas remain a rare but surgically treatable tracheal tumor. Their nondescript presenting symptoms often delay diagnosis. Aggressive complete surgical resection, with or without initial endoscopic airway restoration, is the treatment of choice; incomplete resections are complicated by high rates of local recurrence and the potential for malignant degeneration. For patients whose disease recurs or is deemed to be unresectable, radiotherapy may provide local control.

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