Aggressive Fibromatosis Of The Trachea: A Case Report Of A Primary Tracheal Tumor

ERIC C. VILLALUNA, M.D.; REY A. DESALES, M.D.

Abstract

A rare case of aggressive fibromatosis of the trachea in a 34-year-old male is presented. Sleeve resection and reconstruction was achieved after double lung ventilation was instituted. This procedure followed a preliminary laser "coring out" of the obstructing tracheal lesion.

Key words: primary tracheal tumor, aggressive fibromatosis

PRIMARY tracheal tumors are rare, most of which are malignant.[1,2] The incidence of benign tumors reportedly ranges from 4.5% to 33%.[3,4,5] About two-thirds of primary tracheal tumors are of 2 histologic types: squamous cell carcinoma and adenoid cystic carcinoma (cylindroma).[1,2]

Most of these cases are easily overlooked and misdiagnosed because of their rarity.[6,7] Tracheal tumors may present insidiously. The most common symptoms and signs are cough, hemoptysis, and signs of progressive airway obstruction, wheezing and stridor and less commonly, dysphagia or hoarshness. Wheezing in particular may cause a misdiagnosis of asthma. Primary tracheal tumors that are surgically removable, even by present techniques, are rare in any single institution,[6,7,8,9,10] thus, the accumulation of experience in the treatment of specific lesions has been slow.

It is the objective of this paper to present a report of an obstructing aggressive fibromatosis of the trachea in terms of the:

a. Clinical course
b. Diagnostic modalities
c. Treatment options, including airway control and
d. Review of literature regarding primary tracheal tumors in general

Case Report

A.C., a 34-year-old male, came in with chief complaint of productive cough which started one year prior to admission as intermittent cough productive of scanty phlegm. He self-medicated with antitussives which afforded temporary relief. A month prior to admission, he developed noisy, difficult breathing prompting consultation with a private physician who prescribed salbutamol inhaler and a mucolytic. However symptoms persisted thus the admission.

Physical examination was unremarkable except for occasional inspiratory wheezes on both lung fields. Chest radiograph revealed only right pleural thickening and calcification (Figure 1). An initial fiberoptic bronchoscopy (FOB) under local anesthesia revealed

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Department of Surgery, Lung Center of the Philippines, Quezon Boulevard, QuezonCity.
Reprint requests: Dr. Eric Villaluna, Department of Surgery, Lung Center of the Philippines, Quezon Boulevard, Quezon City.

a mass lesion with smooth surface at the right lateral wall of the trachea with 70% obstruction. A plain chest computed tomography (CT) (Figure 2) revealed an enlarged right paratracheal node measuring 2 cm x 2.5 cm x 3 cm located posterior to the superior vena cava and producing compression of the trachea. Fibrotic densities were noted on the right upper and lower lobes with a moderately thick pleura and calcification on the midposterolateral hemithorax.

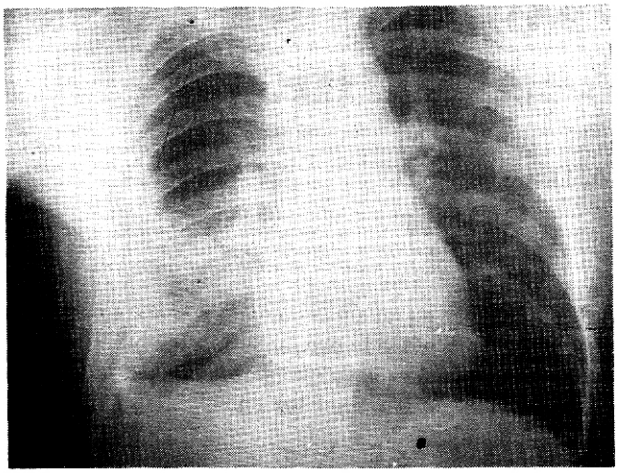

Figure 1. Pre-operative chest x-ray showing right pleural thickening and calcification.

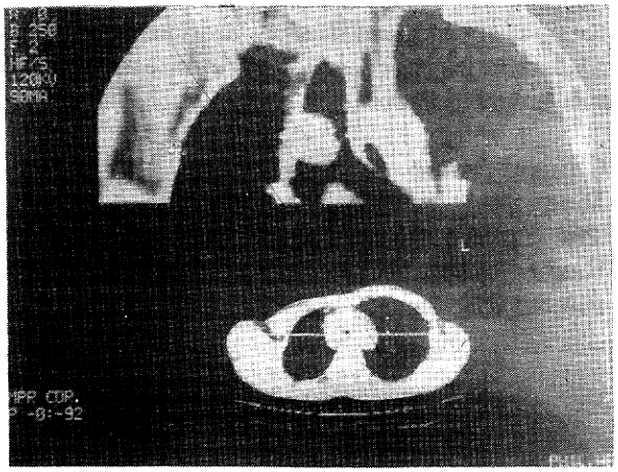

Figure 2. Chest CT scan showing obstruction of tracheal lumen.

The patient underwent rigid bronchoscopy, fiberoptic bronchoscopy and NdYag laser therapy under general anesthesia which showed a sessile, wide-based mass with smooth surface and vascular engorgement attached to the right anteroposterolateral wall of the distal trachea causing 80 to 90% obstruction. The proximal and distal extent was 5 cms and 2 cms from the carina respectively. NdYag laser coagulation of the tumor opened up the tracheal lumen to 20%. The mass was hard with cartilagenous consistency and was difficult to enucleate. Histopathologic report came out as fibromatosis of the trachea.

Chest CT scan with sagittal reconstruction (Figure 3) was done to determine the longitudinal and extratracheal extent of the lesion to plan possible surgical resection. A right posterolateral thoracotomy, decortication, distal tracheal sleeve resection and end-to-end anastomosis was done. Intraoperatively a thickened parietal pleura (Figure 4) 0.5 cm thick over the right middle and right lower lobe and a 3.5 cm x 4 cm mass (Figure 5) about 1.5 cm proximal to the carina encompassing three-fourths of the whole trachea free from the surrounding structures were noted. The trachea was opened distal to the mass and a sterile armored endotracheal tube was inserted (Figure 6) into the left main bronchus for anesthesia and ventilation. Later, another endotracheal tube was placed in the right main bronchus achieving double lung ventilation. The proximal endotracheal tube was retracted superior to the tumor and a tracheal sleeve resection was done. Mobilization of the trachea, carina, right and left main bronchi, and division of the right inferior pulmonary ligament were done together with neck flexion so that both ends of the resected trachea could be approximated without tension. After the sutures were laid down the endobronchial tubes were removed and the knots were tied. Ventilation again resumed through the proximal endotracheal tube.

Postoperatively the endotrachial tube was maintained for 4 days. He was subsequently discharged on the 16th postoperative day after FOB showed an intact anastomosis with minimal granulation tissue.

Histologically the benign submucosal neoplasm in the trachea was composed of interlacing bundles of elongated cells with wavy, dark staining nuclei

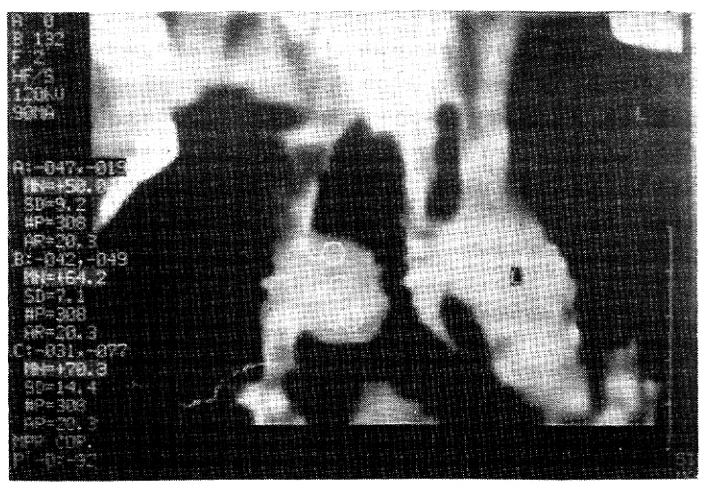

Figure 3. Chest CT scan with sagittal reconstruction showing distal extent of lesion.

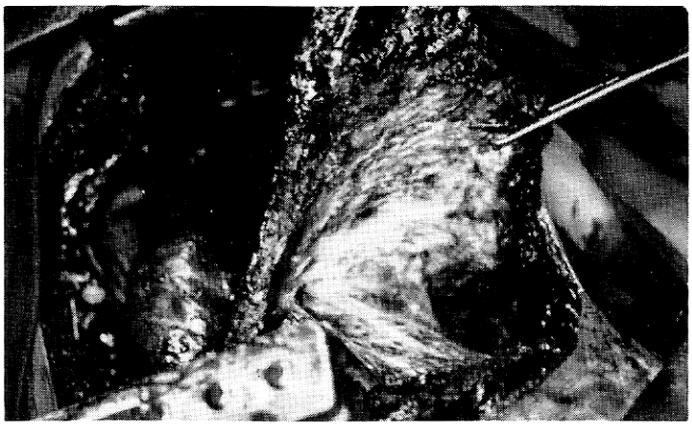

Figure 4. Intraoperative finding of a thickened parietal pleura.

arranged in fascicles or whorls. The stroma was myxoid with a sprinkling of lymphocytes and glandular structures. No abnormal mitosis was seen. Final histopathologic report was reactive hyperplasia, paraesophageal lymph node and aggressive fibromatosis of the distal tracheal mass.

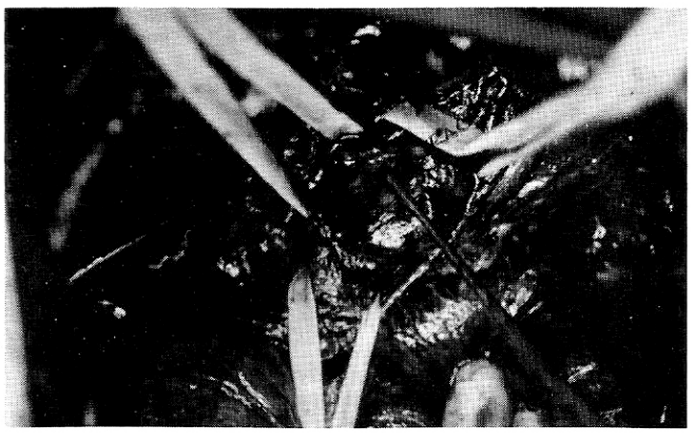

Figure 5. Tracheal mass; its extratracheal component was free from surrounding strictures.

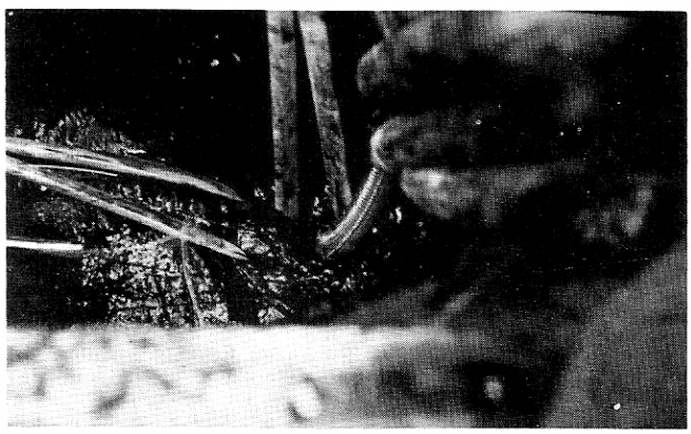

Figure 6. Endobronchial intubation of left main bronchus. Another endotracheal tube was placed into the right main bronchus achieving double lung ventilation.

Discussion

Benign tracheal tumors are rare. There are only a few reported cases of aggressive fibromatosis of the trachea in world literature[3,10,11] and most of them are mentioned as part of a large series.

Fibromatosis are fibroblastic proliferative lesions that fall between a hyperplastic connective tissue scar and a low grade fibrosarcoma. They are not believed to be neoplastic, however, despite their lack of encapsulation and apparent infiltration into surrounding structures; they never display anaplasia nor do they ever metastasize. They are also known as desmoid tumors, which are not muscle cell derivatives from the musculoaponeurotic supporting tissues. The great majority arise in the anterior abdominal wall but origin in the extremities and in relation to muscles within the body has been recorded, e.g. in relation to neck (fibromatosis colli), in the penis (Peyronie's disease), in the feet (plantar fibromatosis).

Morphologically these lesions occur as unicentric, gray-white, firm, unencapsulated poorly demarcated masses that vary in size from small nodules 1 to 2 cm in diameter to large masses up to 15 cm in diameter. The lesion has a rubbery tough consistency and on gross inspection interposes itself between muscles and muscle bundles, separating groups of muscle cells. Histologically they resemble a somewhat cellular scar, having an abundance of collagenous fibrous tissue. The individual fibrocytes are usually uniform in size and shape, and only rarely can large atypical cells or nuclei be identified. Mitoses are infrequent and when found are regular.

In some benign tumors or low grade malignant tumors of the trachea the mean duration of an incorrect diagnosis was four years. Our patient had his disease for one year before it was diagnosed. Too often, a chest radiograph is read as "normal"; however, on closer inspection, abnormality of the tracheal air column may be seen. Computed tomography is helpful to define an extratracheal component as well as to delineate the location of the lesion and its linear extent to determine if resection and reconstruction is possible.

Endoscopy is the most common means by which a tracheal tumor is diagnosed and is usually done for hemoptysis of unknown origin or signs of progressive airway obstruction. Preoperative diagnosis before the planned surgical treatment is viewed differently by different authors. Perelman and associates[5] believe that biopsy should be done as a separate procedure; however, Eschapasse[3] reported that biopsy may be traumatic and potentially hazardous, with risk of hemorrhage and complete tracheal obstruction, and should only be performed when full surgical facilities

should only be performed when full surgical facilities are available. Grillo[14] contends that in radiologically well-defined tumors that appear to be completely resectable biopsy can be deferred until the planned surgical procedure but should be performed as a preliminary procedure if the tumor is extensive. In our patient, an initial diagnostic endoscopy was done to open up the tracheal lumen using NdYag laser in preparation for surgery, since impression on CT Scan was an enlarged paratracheal node.

Surgical resection with primary airway reconstruction is the treatment of choice for benign tracheal tumors. The goal of surgery is two-fold: relief of airway obstruction in the most effective way possible and the achievement of cure.

Before 1962, primary tracheal tumors were managed by endotracheal morcellation, irradiation and conservative lateral resection with patching. The results of earlier treatment, even for adenoid cystic carcinoma with its long clinical course, were poor. Early recurrence was the rule, and in most cases death was due to airway obstruction. After 1962, a technique evolved which permitted resection in most instances of half or more of the trachea with primary reconstruction. One large series is at the Massachusetts General Hospital[15] from November 1962 to July 1987 involving 198 patients with primary tracheal tumors of which 147 underwent surgical resection.

To date, there is no prosthesis that can replace the trachea or the rest of the bronchial tree, nor is there any autogenic material proven to be effective in replacing the respiratory tract. Thus reconstruction is achieved thru primary anastomosis. Various release procedures have been devised to relieve anastomotic tension. The simplest and most commonly used maneuver is neck flexion of 15 to 35 degress which enabled us to obtain a 4.5 cm additional length or approximately 7 tracheal rings. Beyond 35 degree flexion, an additional 1.5 cm length can be achieved. If this is not enough, additional release procedures include: Montgomery suprahyoid release (1.5 cm),[16] hilar and inferior pulmonary ligament release (1.5 cm), and dissection of the pulmonary vein and pulmonary artery intrapericardially (1.5 cm). Our case represented a benign tracheal tumor which involved the distal trachea sparing the carina thus a straightforward resection and end-to-end anastomosis was done. However had the tumor involved the carina a bronchoplastic technique such as described by Grillo[17] would be appropriate.

For patients who may not be candidates for resection either because of too extensive a disease or intolerance of the procedure, various options of palliative airway management are available. McCaughan[18] reported photodynamic destruction of endobronchial tumors using tunable dye argon laser (model 171; Spectra Physics, Mt. View, CA) for adenocarcinoma and squamous cell carcinoma. Mathisen and Grillo[19] used biopsy forceps and a rigid bronchoscope to "core out" obstructing airway neoplasms. Tsang and colleagues[20] used sequential silastic and expandable metal stenting on five patients with recurrent tracheal or bronchial strictures.

Grillo's extensive experience and earlier report made the following recommendations:[15]

First: Benign primary tumors of the trachea and tumors of intermediate aggressiveness are best treated by surgical resection with airway reconstruction.

Second: Primary squamous cell carcinoma and adenoid cystic carcinoma of the trachea are best treated by resection when primary reconstruction may be safely accomplished. Resection should be followed by full dose mediastinal irradiation in most cases to control late recurrence, and

Third: Malignant tracheal tumors of other types should also be resected if safe primary reconstruction can be achieved.

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