Tracheal Myxoma: A Rare Benign Tracheal Tumor

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Benign tracheal tumors are rare. We describe a 39-year-old man who underwent resection of a tracheal myxoma, a previously unrecognized benign tracheal neoplasm. He presented with a 9-month history of wheezing, cough, and dyspnea on exertion. Treatment with bronchodilators and corticosteroids administered by inhalation and systemically did not diminish his symptoms. Pulmonary function tests showed a pattern of airflow limitation consistent with variable extrathoracic obstruction. Chest radiography and computed tomography revealed a tracheal mass. Tracheal resection of the tumor with reconstruction was curative. The patient is free of disease 7 years after surgery.

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Tracheal tumors are rare among patients in all age groups, and most occur in adults. Most of the primary tumors of the tracheal and proximal bronchi are malignant in adults, the majority being squamous cell or adenoid cystic carcinoma.[1] Benign tumors occur predominantly in males, with a peak incidence in the fifth and sixth decades of life.[1] Myxomas are benign tumors of connective tissue derived from early primitive mesenchyma.[2-4] The most frequently involved site is the heart, followed by the jaws and muscles of the extremities.[4] To our knowledge, this is the first published report of a primary tracheal myxoma.

REPORT OF A CASE

A 39-year-old man was referred to our institution for evaluation of a tracheal mass. He had had progressive wheezing, cough, and dyspnea on exertion for 9 months before our evaluation. Initially, his symptoms were attributed to asthma, and he was treated with a combination of inhaled bronchodilators as well as inhaled and systemic corticosteroids without improvement. Chest radiography performed before referral showed a round mass in the upper trachea (Figure 1). Chest computed tomography (CT) performed before referral revealed a round and smooth mass, without evidence of ulceration, local invasion, or nodal or distant metastases. The mass was obstructing more than 80% of the tracheal lumen. Pulmonary function testing at our institution yielded a normal forced vital capacity of 4.75 L (99% of predicted), forced expiratory volume in 1 second of 3.63 L (93% of predicted), and moderately decreased maximum voluntary ventilation of 90 L (56% of predicted). The patient had no immediate response to bronchodilators; however, flow-volume curves showed an inspiratory plateau compatible with extrathoracic obstruction (Figure 2). Bronchoscopy performed in the operating room showed a smooth polypoid mass arising from the left posterior wall of the trachea, 3 cm below the cricoid cartilage, at the thoracic inlet. Open segmental resection was performed followed by primary end-to-end anastomosis. The patient recovered without complications and was discharged 7 days postoperatively. Bronchoscopy performed before discharge from the hospital revealed a healed tracheal anastomosis. Seven years after the surgery, the patient is asymptomatic and has had no recurrence of disease.

PATHOLOGIC FINDINGS

The resected specimen consisted of a short tracheal segment containing a well-circumscribed exophytic red-brown polypoid mass, 2 cm in greatest dimension, arising from the posterior aspect of the trachea (Figure 3). All tumor margins were free of involvement. Microscopically, the tumor was nonencapsulated and comprised cytologically bland spindle and occasionally stellate cells arranged randomly within a pale-staining basophilic myxoid matrix (Figure 4). Immunoperoxidase stains performed on paraffin sections of the tumor using a standard technique were negative for CD34, S-100 protein, desmin, and muscle-specific actin (HHF35).

DISCUSSION

To our knowledge, this is the first published report of a tracheal myxoma in a patient whose symptoms were initially attributed to asthma. Chest radiography and chest CT revealed a round and smooth mass obstructing more than 80% of the tracheal lumen. Pulmonary function testing was helpful in showing bronchodilator-insensitive airflow obstruction and inspiratory flow limitation, consistent with an extrathoracic airway obstruction. Tracheal resection and reconstruction were curative.

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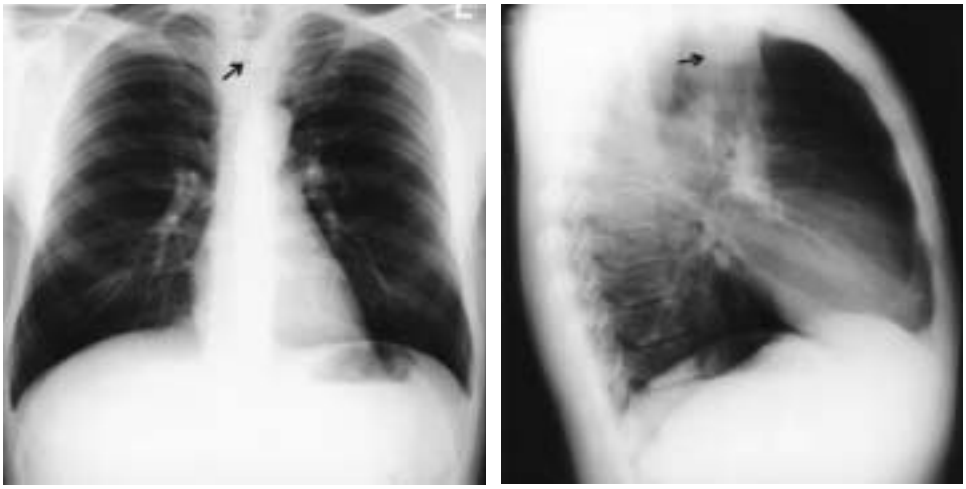

FIGURE 1. Plain posteroanterior (left) and lateral (right) chest radiographs showing round mass (arrow) in the upper trachea.

Because tracheal tumors are rare, the initial presentation is often confused with other diagnoses.[1,5] Wheezing is a common presenting complaint, occurring in as many as 56% of all patients with tracheal neoplasms.[6] Not surprisingly, wheezing in such patients is often mistakenly attributed to chronic asthma.[7] Wheezing is not prominent until the tumor obstructs at least 25% of the normal luminal diameter,[8] although tracheal tumors typically obstruct 50% of the tracheal lumen by the time of diagnosis.[6] Respiratory flow-volume–loop analysis is extremely useful in distinguishing extrathoracic tracheal obstruction from airflow limitation resulting from intrinsic disease of smaller airways.[7]

Radiological differentiation between malignant and benign tracheal tumors is difficult. Features more commonly seen in benign tumors include a round and smooth localized mass less than 2 cm in diameter, without evidence of ulceration, local invasion, or nodal or distant metastases.[9,10] Chest CT is important in assessing tracheal abnormalities in general and tracheal tumors in particular. Computed tomography is not only superior to chest radiography in showing tracheal tumors but also helpful in determining the exact location of the tumor, in determining whether the tumor penetrates the tracheal wall and its relationship to the surrounding structures that may affect resectability, and in determining its ability to disclose nodal or pulmonary metastases.[10] Thin-section multidetector CT with 3-dimensional reconstruction on virtual bronchoscopic models allows more accurate assessment of the lesion's surface features, the degree of airway obstruction, and the extraluminal extent of an airway lesion. It also helps to locate the position of the tumor and the amount of the trachea that will need to be resected for complete excision. Moreover, patency of airways distal to an obstruction can be assessed better than with fiberoptic bronchoscopy.[10,11] Fiberoptic bronchoscopy remains the gold standard to confirm a specific pathologic diagnosis.[5,12]

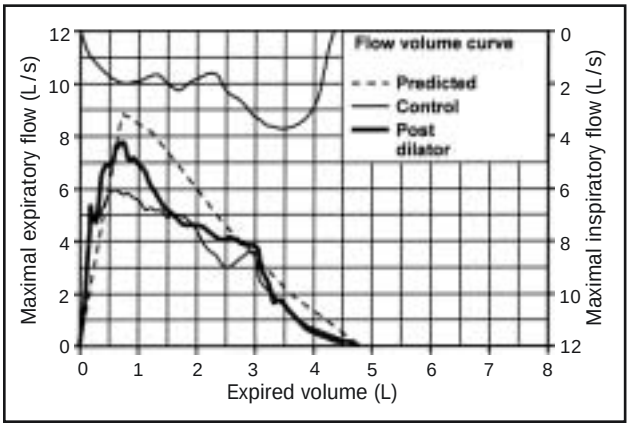

FIGURE 2. Flow-volume–loop analysis before surgery. The inspiratory flow curve shows marked reduction of maximal forced inspiratory flow.

Myxomas are benign tumors of connective tissue derived from fibroblasts and contain a stroma rich in connective tissue mucin.[2-4] In a review of extracardiac myxomas, Stout[4] reported that the most frequently involved sites are the jaws and skeletal muscles of the extremities. Myxomas have been reported less commonly in the orbit, eyelid, joints,[4] larynx,[13] and oral soft tissue.[14] Only 1 endobronchial myxoma[2] and 1 myxoma of the lung[3] have been reported in the English literature. In all these sites, complete surgical excision was curative.

The differential diagnosis of histologically benign spindle cell endobronchial tumors is limited. Hamartomas

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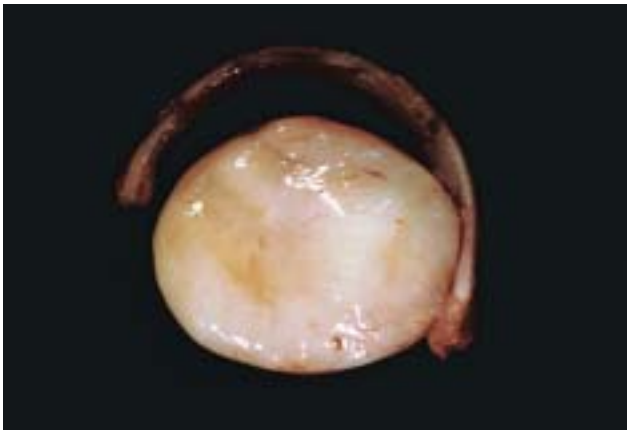

FIGURE 3. Resected specimen showing the cut surface of a polypoid intratracheal myxoma. The well-circumscribed tumor has a slender attachment to the tracheal ring.

are the most common benign lung neoplasms and nearly all are peripheral; endobronchial hamartomas account for less than 2% of reported tumors.[15] A cartilaginous component is present in most cases, and S-100 staining reveals both cartilaginous and noncartilaginous stromal elements, features absent in our case. Intrapulmonary solitary fibrous tumors are less common and usually lack the myxoid stroma that represented a distinctive histological feature of the tumor described in our patient.[16] Immunohistochemical staining positivity for CD34 is also typical of solitary fibrous tumors, including uncommon myxoid variants,[17] and stains of our patient's tumor were negative for CD34. Carcinoid tumors are commonly endobronchial and can have spindle cell morphology but have different cytologic characteristics, including larger nuclei with occasionally conspicuous small nucleoli and more abundant cytoplasm. Neoplastic cells in carcinoid tumors are organized in distinctive growth patterns, characteristic of neuroendocrine neoplasms, that differ substantially from the random and haphazard arrangement of spindle cells of myxoma.

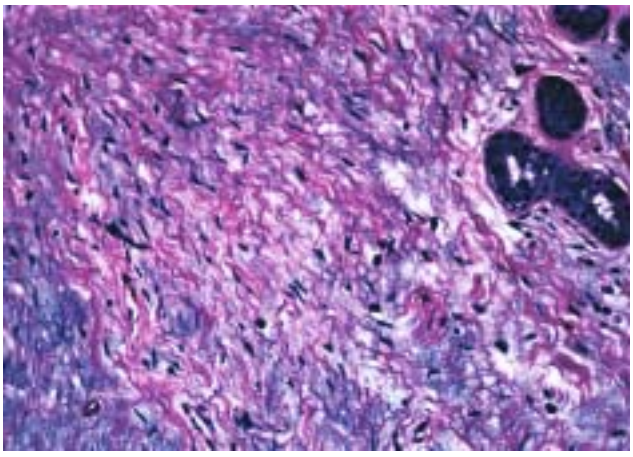

FIGURE 4. Photomicrograph of tracheal myxoma showing bland neoplastic spindle cells arranged haphazardly in a pale-staining myxoid matrix. Neoplastic spindle cells surround nonneoplastic submucosal glands (upper right corner) (hematoxylin-eosin, original magnification ×100).

CONCLUSION

To our knowledge, this is the first published report of a patient with a primary tracheal myxoma. Surgical resection is the treatment of choice for such tumors. Although tracheal neoplasms are rare, their presence should be considered in all cases of atypical wheezing, especially when asthma symptoms are difficult to control.

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