Successful rigid bronchoscopic resection of recurrent pulmonary inflammatory myofibroblastic tumor after complete surgical resection

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ABSTRACT

Inflammatory myofibroblastic tumor, previously named inflammatory pseudotumor, is a biologically borderline mesenchymal neoplasm often associated with an inflammatory infiltrate. The incidence of inflammatory myofibroblastic tumor has been found to range from 0.04% to 1.2%, with endobronchial cases being extremely rare. The treatment of choice for pulmonary inflammatory myofibroblastic tumor is complete surgical resection. However, disease recurrence has been reported. Modalities used to treat recurrent and metastatic disease include surgical resection and corticosteroids. We present a case of recurrent endobronchial inflammatory myofibroblastic tumor that was successfully treated endoscopically with rigid bronchoscopy and laser debulking without need for further surgical intervention.

KEYWORDS Bronchoscopy; inflammatory myofibroblastic tumor; inflammatory pseudotumor

CASE DESCRIPTION

A 56-year-old woman presented with chronic cough associated with postnasal drip. She denied fever, chills, weight loss, and hemoptysis. Over-the-counter antihistamines were not helpful and she did not respond to a course of antibiotics. Chest computed tomography (CT) showed a left main stem–obstructing lesion (*Figure 1*). The patient then underwent flexible bronchoscopy, during which a rounded, well-circumscribed mass was seen occluding the left main stem bronchus. Pathological analysis of the biopsy revealed relatively tightly packed spindle cells admixed with scattered mixed inflammatory cells. Staining for anaplastic lymphoma kinase (ALK-1) was positive, consistent with an inflammatory myofibroblastic tumor (IMT). She underwent a left posterolateral thoracotomy and distal left main stem partial bronchotomy with complete local resection of the myeloproliferative tumor at the left main stem bifurcation.

Pathological analysis of the surgical specimen showed a densely cellular spindle cell neoplasm with scattered admixed inflammatory cells, predominantly lymphocytes. The spindle cells showed moderately abundant lightly basophilic cytoplasm and relatively vesicular nuclei with small nucleoli. The margins were not tumor free, which may indicate incomplete surgical resection. No overt anaplasia was evident. Selective immunohistochemical stains showed strong and diffuse ALK-1 positivity, which confirmed the diagnosis of IMT.

The patient recovered without complications and was monitored by her oncologist. Her follow-up CT 6 months after surgery did not show evidence of tumor recurrence but at 18 months the CT showed a 1.6-cm endobronchial lesion in the left main stem bronchus. This time the patient was asymptomatic. She underwent rigid bronchoscopy, with complete resection of the endobronchial mass using argon laser and a cryoprobe (*Figure 2*). Pathology confirmed

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Figure 1. CT showing tumor in left bronchus with near-complete obstruction and postobstructive atelectasis.

Figure 2. (a) Bronchoscopy showing inflammatory myofibroblastic tumor in the left main bronchus. (b) Bronchoscopy image after resection and laser argon probe treatment.

recurrence of disease (*Figure 3*). Eight months after resection of the recurrence, bronchoscopies have shown no tumor recurrence.

DISCUSSION

Inflammatory pseudotumor is a generic term applied to a variety of neoplastic and nonneoplastic entities that share a common histological appearance: a cytological bland spindle cell proliferation, usually with a prominent chronic inflammatory infiltrate.¹ Thoracic tumors typically present as small peripheral tumors, and they constitute 0.7% to 1% of all lung tumors.^{2,3} This tumor has been known to grow slowly and cause local invasion.

Lesion cells are predominantly myofibroblasts in a myxoid collagenous stroma admixed with lymphocytes, eosinophils, and histiocytes.⁴⁻⁶ At the genetic level, approximately 50% of these tumors exhibit rearrangements involving the ALK locus on chromosome 2p23, with reactivity ranging from 36% to 71%.¹

IMT primarily affects younger patients.⁷ The mean age of diagnosis and treatment is 23 to 66 years.⁵⁻⁷ Presenting symptoms include dyspnea, chest pain, hemoptysis, and recurrent cough.⁶ Sometimes, weight loss, fever, and obstructive airway syndrome can also be present.⁵

Figure 3. Pathological appearance of inflammatory myofibroblastic tumor.

The treatment of choice is surgical resection.^{5,6,8} Wedge resection, either video-assisted or with an open technique, is suitable for curative purposes. When wedge resection is not technically feasible, the only other option is major resection. This involves lobectomy or pneumonectomy and in some cases may involve removal of neighboring anatomic structures, including the chest wall and diaphragm.⁸

In one case series, the 5-year and 10-year survival rates after complete resection were 89%.² Tumor size seems to be an important factor in survival. In an analysis of 18 surgical resections, survival was better with tumor-free margins and a tumor size <3 cm.⁸ IMT of the trachea and lung does not usually recur after complete resection. Relapse has been reported; it may occur many years after resection and can cause disease-related mortality.⁹ Distant metastasis is rare, occurring in <5% of cases.¹ Recent studies suggest that ALK-positive tumors have a very low risk of metastasis, and ALK reactivity does not appear to correlate with recurrence.¹

Recurrence of IMT is rare. In a large series of 23 pulmonary parenchymal IMT surgically treated cases over 47 years, only three recurrences were reported, all due to incomplete resection.⁹ There are no specific data on the recurrence rates of endobronchial tumors. No consensus guidelines exist for treatment of a recurrence. Some published data suggest treatment with surgery.⁹ The type of treatment depends on various factors, including site of the recurrence, amenability to complete resection, size of the lesion, number of lesions, and whether the lesion involves the central airway or the lung periphery. A central lesion is generally more amenable to resection than multiple and/or peripheral lesions.⁹

One report noted successful treatment of recurrent parenchymal lung disease with corticosteroid therapy.^{10,11} Nonsteroidal anti-inflammatory drugs and chemotherapy regimens including vincristine and methotrexate have also been used.^{9,12} The role of chemotherapy or radiation is controversial, and they are not considered first-line modalities of

treatment.⁸ When complete resection is not possible, due to anatomic location or comorbidities, adjuvant chemotherapy in conjunction with radiation therapy has been used.⁷

Radiation treatment is also controversial with indeterminate results. Currently, the recommended treatment is 4000 to 4500 rad given in 180- to 200-rad fractions, with carefully tailored fields to tumor volume to minimize the dose to the surrounding normal tissue.² Radiation is typically reserved for palliation, to alleviate the mass effect, or in conjunction with chemotherapy for cure in patients who are not amenable to resection.⁷

Rigid bronchoscopy with laser mechanical debulking has been used primarily as a means to relieve airway obstruction prior to surgical resection.⁶ This case reflects successful use of this treatment modality as a means of treating IMT recurrence, therefore avoiding further additional surgery.

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