Endobronchial Tumors in Children: Institutional Experience and Literature Review

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Background/Purpose: Endobronchial tumors are rare in children and often misdiagnosed as benign conditions resulting in delayed definitive treatment. The authors reviewed their experience to highlight pertinent aspects of diagnosis and treatment.

Methods: A retrospective chart review was conducted of children with endobronchial tumors diagnosed between 1980 and 2002.

Results: Nine patients had endobronchial tumors (5 girls, 4 boys), with average age of 13 years (range, 8.5 to 15 years). There were 5 carcinoid tumors, 3 mucoepidermoid carcinomas, and one pseudotumor. Preoperative bronchoscopic biopsy confirmed the diagnosis in 6 patients, was inconclusive in one, and not done in 2. All except one (pseudotumor) underwent surgical resection. Laser ablation was performed in 2 cases with complete cure in one. All had an uneventful postoperative course except one patient in whom ipsilateral pneumonia developed. Long-term follow-up was obtained with clinical examination, pulmonary x-ray, abdominal ultrasound scan, chest computed tomography scan, and serum 5-HIAA in those with carcinoid tumor. Bronchoscopy was performed twice yearly for the first 2 years, then yearly. No evidence of local or distant recurrence was reported.

Conclusions: Endobronchial lesions should be considered in children with persistent pneumonia despite adequate treatment or with undiagnosed respiratory symptoms. Prognosis is excellent with surgical resection. Specific follow-up protocol is recommended.

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INDEX WORDS: Endobronchial tumors, pediatric tumors, carcinoid, pseudotumor, mucoepidermoid carcinoma.

Endobronchial Tumors are rare in children and often are misdiagnosed resulting in delayed definitive treatment.1-3 Although the true incidence of this entity is unknown, bronchial adenomas alone may account for 5% of all primary pulmonary neoplasms in children.4 Relatively little attention has been given to the diagnosis, treatment, and follow-up of these lesions. Recent advances in diagnostic and therapeutic modalities deserve special attention. We reviewed our experience to highlight the pertinent aspects of diagnosis, treatment, and follow-up of these tumors.

MATERIALS AND METHODS

The medical records of 9 children with endobronchial tumors diagnosed between 1980 and 2002 were reviewed retrospectively. Data pertaining to their presentation, diagnosis, treatment, pathology, and clinical course were documented.

RESULTS

Over the past 22 years, 9 children were found to have endobronchial tumors (5 girls and 4 boys). The mean age at diagnosis was 13 years (ranging from 8.5 to 15 years). Table 1 outlines the clinical details of these cases. All patients had computed tomography (CT) scans and underwent bronchoscopy. Bronchoscopic biopsy confirmed the diagnosis in 6 patients, was inconclusive in one, and was not performed in 2 (one because of the presenting symptoms of hemoptysis and the other was at the discretion of the bronchoscopist). All biopsies were uneventful with no significant bleeding reported. Eight of the patients underwent thoracotomy and resection of the lesion (Table 1). The one patient who did not undergo thoracotomy had pseudotumor diagnosed and underwent successful laser ablation. Laser ablation was performed in one other patient but was unsuccessful, and this patient required thoracotomy (Table 1). All patients had an uneventful postoperative course with the exception of one in whom ipsilateral pneumonia developed. Mean follow-up was 3.3 years and ranged from 6 months to 8 years.

DISCUSSION

Bronchial tumors are extremely rare in children. They include benign tumors such as hamartomas, hemangiomas, papillomas, inflammatory pseudotumors (plasma cell granulomas), leiomyomas, and mucus gland tumors.
The malignant lesions include bronchial adenomas (an obvious misnomer), carcinoids, mucoepidermoid carcinomas, and adenoid cystic carcinomas. Bronchogenic carcinomas are reported rarely in childhood.1-8,11 The most common endobronchial tumor in children and adolescents is carcinoid.2 In this study it accounts for 55% (5 of 9) of all patients. Of the 25 cases reported in the literature, 5 (20%) had local or distant recurrence, with subsequent death in 3 patients 4 to 5 years after diagnosis.4,6 Carcinoid tumors can be divided into typical and atypical forms with the latter exhibiting histologically malignant features and aggressive clinical behavior. This histologic and clinical distinction is not as clear in the pediatric age groups as it is in adults because of the limited pediatric experience with this pathology. All 5 patients in this study with carcinoid displayed typical histologic features and had no local or distant recurrences. Because of the abundant vascularization, endoscopic removal of these lesions can be hazardous and currently is not recommended.9 In adult populations, preoperative laser ablation was found to be safe and helpful in relieving bronchial obstruction and facilitating conservative surgical resection.9-10 We observed transient improvement in one of our patients with carcinoid tumor who underwent laser ablation, but the patient was referred subsequently for formal surgical resection. Because of excellent survival, limited resection is the recommended treatment even with lymph node involvement.4,6,9

Bronchial mucoepidermoid tumor (MET) is rare and accounts for 2.5% to 7.3% of endobronchial adenomas and represents 0.1% to 0.2% of primary lung cancers.11,12 In the current study, MET represents one third of all endobronchial tumors, which is more than what has been reported previously. Including our 3 patients, 55 cases have been described in children.11-15 Despite a 3:2 male to female ratio in recent reviews, all of our 3 cases of MET were girls.11,14 These tumors typically arise from bronchial mucous glands in the main stem bronchus or in the proximal portion of lobar bronchi as an endobronchial polypoid growth that is covered by normal respiratory epithelium. For this reason, bronchial lavage and brushing are seldom diagnostic, and forceps biopsy must be performed.11,13 Endobronchial biopsy was performed in 7 of our patients and was diagnostic in 6 (85%) including the 3 MET.

Histologically, MET are classified as low grade or high grade. In childhood, these lesions usually are low grade.12 All of our 3 patients had low-grade lesions with no evidence of metastases. Lymph node metastases, although historically rare, have been documented in the literature in 3 cases (two high-grade variants and one low-grade variant).11,16,17 Of the 3 patients with metastatic disease described in the literature, one patient with high-grade MET was alive without recurrence 8 years after diagnosis, the second high-grade patient was lost to follow-up, whereas the patient with low-grade MET was alive and disease free at 5 years of follow-up.

In the evaluation of patients with MET, chest radiograph usually is abnormal, showing a central mass or a nodule in 66%.18 CT scan or magnetic resonance imaging (MRI) with 3-dimensional reconstruction may be very helpful in planning definitive surgical treatment. Pulmonary inflammatory pseudotumor is known also as plasma cell granuloma. Among the more than 19

<table>
<thead>
<tr>
<th>Patient No.</th>
<th>Age (yr) at Diagnosis</th>
<th>Sex</th>
<th>Symptoms/Signs</th>
<th>Diagnostic Workup</th>
<th>Location</th>
<th>Diagnosis</th>
<th>Treatment</th>
<th>Follow-Up (mo)/Outcome</th>
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<tr>
<td>1</td>
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<td>F</td>
<td>Cough, weight loss, dyspnea</td>
<td>CT, bronchoscopy, biopsy</td>
<td>LMB</td>
<td>Mucoepidermoid carcinoma</td>
<td>Sleeve resection</td>
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<td>2</td>
<td>13</td>
<td>M</td>
<td>Pneumonia</td>
<td>CT, bronchoscopy, biopsy</td>
<td>RML</td>
<td>Carcinoid</td>
<td>Laser, RML, RLL, lobectomy</td>
<td>48/NED</td>
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<td>CT, bronchoscopy, biopsy</td>
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<td>RLL, RML, lobectomy</td>
<td>96/NED</td>
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<td>CT, bronchoscopy, biopsy</td>
<td>LLL</td>
<td>Carcinoid</td>
<td>LLL, lobectomy, and sleeve resection</td>
<td>36/NED</td>
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<td>Pseudotumor</td>
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<td>10</td>
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<td>6/NED</td>
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Abbreviations: LMB, left main bronchus; RML, right middle lobe; LLL, left lower lobe; RLL, right lower lobe; NED, no evidence of disease.
different names in the literature, inflammatory myofibroblastic tumor (IMT) is the most recent and probably the most descriptive term for this pathological entity.\textsuperscript{18-20} IMT is believed to be a nonneoplastic reactive inflammatory condition. The concept that these are benign lesions has been challenged recently by both clinical demonstration of recurrence and cytogenetic evidence of acquired clonal chromosomal abnormality.\textsuperscript{21} Detailed immunohistochemical studies and flow cytometric analyses of IMT may be helpful to differentiate lesions that will remain localized from the more aggressive forms.\textsuperscript{22} These lesions constitute 20\% of all primary lung tumors and 57\% of all benign lung lesions.\textsuperscript{19} They present typically as a pulmonary nodule within the lung parenchyma and occasionally as an endobronchial lesion. Preoperative diagnosis usually is difficult, and frozen section may be useful to differentiate pseudotumors from neoplastic lesions such as soft tissue sarcomas, but radical resection should not be based on frozen section alone. In our patient, frozen section suggested the diagnosis of pseudotumor, which was confirmed by final histopathologic and immunohistochemical studies.

The recommended treatment for IMT is conservative complete surgical resection. Recurrence has been reported in 3 of 5 patients who underwent incomplete resection.\textsuperscript{23} Other therapeutic modalities include radiation therapy, immunomodulatory therapy, steroids, and combination of chemotherapeutic agents in recurrent cases.\textsuperscript{24,25} We used laser ablation in one of our patients who was cured and found free of the disease 6 years after treatment. Laser ablation was reported also to be successful in treating 2 other cases of tracheal pseudotumors.\textsuperscript{19} This should be considered an excellent alternative treatment in those cases with difficult surgical options. Successful local excision through bronchotomy has been reported also in one case of endobronchial pseudotumor.\textsuperscript{26}

Because of the slow growth of all endobronchial tumors and the potential for local or distant recurrence, a specific follow-up protocol should be considered. We propose the following: clinical examination and chest x-ray every 3 months for 1 year and every 6 months thereafter, abdominal ultrasound scan to rule out hepatic metastases, chest CT scan, and urine and serum 5-HIAA every 6 months for those patients with carcinoid tumor. Bronchoscopy is performed every 6 months for the first year, then yearly, particularly in those patients who have not had formal resection.

Endobronchial tumors should be considered in the differential diagnosis of children with respiratory symptoms who do not respond to standard medical therapy. Selective conservative surgical resection is the treatment of choice. Laser ablation may be considered an alternative therapy with variable efficacy in certain tumors. Considering the fact that recurrences have been reported many years after surgery, long-term follow-up, probably for life, is essential.

REFERENCES