Granular Cell Tumor of the Bronchus

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Summary. Persistent atelectasis and recurrent pneumonia in the same location should raise suspicion of congenital anomalies or obstructing lesions of the bronchus leading to the affected area. We present an 8-year-old black female with a history of recurrent fever, cough, atelectasis of the right middle and lower lobes, and weight loss for several months. Flexible bronchoscopy revealed a polypoid mass obstructing the bronchus intermedius. Biopsy of the neoplasm demonstrated a granular cell tumor (GCT). The patient had a lobectomy of the right lower and middle lobes. She had no recurrence of the tumor after several years of follow-up. Pediatr Pulmonol. 2000; 30:425–428. © 2000 Wiley-Liss, Inc.

Key words: granular cell tumor; myoblastoma; atelectasis; cough; pneumonia; children.

INTRODUCTION

Obstruction of the airways leads to atelectasis and recurrent or persistent lung infiltrates. Extraluminal lesions include enlarged lymph nodes, vascular rings, left atrium, or pulmonary vessels. Intraluminal lesions include foreign body, mucus plug, bronchial stenosis, and tumors.1 Benign primary pulmonary neoplasms are uncommon in pediatric patients.2,3 Endobronchial granular cell tumor (GCT) is an unusual benign tumor that is rarely seen in young children.4 It causes obstruction of the involved bronchi and atelectasis of the lung distal to the tumor.4 Surgical resection of the neoplasm and the damaged distal airways is curative and has an excellent prognosis.5

CASE REPORT

This is an 8-year-old black female who was admitted to the hospital with a history of fever, cough, right chest pain, loss of weight, and loss of appetite. A decreased appetite and weight loss had been present over a 4-month period. She had arthralgia of the right ankle, right shoulder, and right wrist joints. She had a previous history of an infiltrate of the right middle and right lower lobes 9 months prior to this admission. A few months prior to the current admission, she was hospitalized and treated for right lower lobe pneumonia. She was also treated multiple times with oral antibiotics as an outpatient. She had normal IgA, IgM, and IgG immunoglobulin levels, a moderate increase in her IgE level, normal IgG subclasses, a normal total hemolytic complement level, and one normal sweat chloride test. She also had a negative PPD skin test prior to this hospitalization.

During this admission, the patient’s chest x-ray showed atelectasis of the right middle and right lower lobes and flattening of the right hemidiaphragm. Bronchoscopy showed a polypoid tumor-like mass at the entrance of the bronchus intermedius, obliterating the openings of the right lower and right middle lobe bronchi (Fig. 1). The mass was about 1 cm in diameter with a capsule and a marked vascular pattern. A CT scan of the chest was done which showed an intrabronchial lesion that appeared entirely within the right bronchus at the origin of the right upper lobe branch. No lymphadenopathy was seen. There was also atelectasis of the right lower lobe. A bronchogram was done, and this showed extensive bronchiectatic changes in the right lower lobe bronchi. No definite middle lobe bronchus was identified. The patient underwent right middle and right lower lobectomies with bronchoplastic reconstruction of the right upper lobe bronchus.

The tumor was a polypoid mass measuring 2.5 × 2 cm and was filling the main bronchial branch to the right

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lower and middle lobes of the lung (Fig. 2). Microscopically, the lesion was made up of clumps and sheets of round to polyhedral pale cells with marked cytoplasmic granularity (Fig. 3). The granules stained positively with PAS and S100 stains. The overlying respiratory epithelium was focally ulcerated. Chronic inflammatory cells, including lymphocytes and plasma cells, were noted throughout. Trapped submucous glands were also present.

The patient was evaluated by the immunologist for her weight loss and arthralgia. Her hand and wrist X-rays showed generalized bone demineralization. There was diffuse juxta-articular osteoporosis with focal erosions, and soft-tissue swelling of interphalangeal and wrist joints bilaterally. She had positive ANA and rheumatoid factor tests. She was diagnosed with juvenile rheumatoid arthritis (JRA) and was treated with naprosyn, prednisone, and methotrexate. No recurrence of the tumor was seen after 6 years of follow-up.

DISCUSSION

Recurrent pneumonia can be caused by various disorders of local or systemic defense mechanisms. Persistent atelectasis or infiltrates in the same area should raise suspicion of aspiration syndromes or structural abnormalities of the respiratory tract. Several underlying factors, including anatomic, neuromuscular, and functional disorders of the respiratory and gastrointestinal tracts, can predispose the patient to aspiration injury and recurrent pulmonary infiltrates. Aspiration disorders were excluded in this patient on the basis of a negative history and examination. From the history and previous negative workup, there was also no evidence of systemic diseases such as cystic fibrosis, immune deficiencies, or immotile cilia syndrome.

Primary pulmonary neoplasms are unusual in children. In a review of 230 primary tumors of the lung in children 16 years of age or younger, two thirds of the tumors were malignant or potentially malignant. The most common primary pulmonary tumors were bronchial adenomas, which are considered malignant in nature. Benign primary pulmonary neoplasms are uncommon in

**Abbreviations**

ANA Antinuclear antibody
CT Computed tomography scan
GCT Granular cell tumor
HIV Human immunodeficiency virus
JRA Juvenile rheumatoid arthritis
PAS Periodic acid-Schiff
PPD Purified-protein derivative
Granular Cell Tumor

pediatrics. These include hamartomas, sclerosing hemangiomas, inflammatory pseudotumors, chondromas, neurogenic tumors, leiomyomas, mucous gland adenomas, and granular cell tumors.\(^2\,^3\) Hamartomas and inflammatory pseudotumors are the most common benign tumors of the lung in children.\(^2\,^3\)

Granular cell tumors (GCTs) are usually tumors of the skin and tongue.\(^7\) They were first described by Abni Kossoff in 1926.\(^4\) They are uncommon in the respiratory tract.\(^5\,^8\,^9\) Pulmonary GCTs constitute 6–10% of all GCT.\(^4\,^10\,^11\) These tumors tend to occur classically in middle-aged patients.\(^4\) The mean age of patients with GCT was reportedly in the third decade of life (36–37 years).\(^11\,^12\)

In two recent series of 28 cases, the patients’ ages ranged from 18–57 years, with a median age of 45 years.\(^5\,^13\) The first pediatric patient with bronchial GCT was reported in 1939 by Kramer in a 15 year old female.\(^14\) Oparah and Subramanian reported a GCT in a 10-year-old black boy that obstructed the left upper lobe bronchus.\(^11\) The youngest pediatric patients reported, however, were 5- and 8-year-old girls.\(^15\,^16\) Both patients had the tumor in the right main bronchus. The 5-year-old had the tumor in the right main bronchus extending to the lower trachea. This patient needed a right pneumonectomy, external radiotherapy, and interstitial brachytherapy.\(^1\) Our patient was later found to have juvenile rheumatoid arthritis (JRA). This is the first case of GCT described with JRA. No association between JRA and neoplasms has been described previously in the literature. Juvenile rheumatoid arthritis is probably the cause of this patient’s weight loss and recurrent febrile episodes. It is possible that the presence of JRA is an incidental finding in this patient. However, GCTs were reported in a few patients with other diseases.\(^4\,^20\,^21\) Multiple endobronchial GCTs were described in an HIV-positive patient.\(^20\) GCTs were also reported in 2 patients with sarcoidosis.\(^4\,^21\) Additional non-neoplasia-associated conditions included cerebral arteriovenous malformation and pulmonary chondroid hamartoma.\(^4\)

The majority of GCTs cause obstruction of the involved bronchi resulting in pneumonias, atelectasis, cough, and hemoptysis.\(^4\,^13\) In one series, 50% of patients had radiological findings consistent with bronchial obstruction.\(^4\) Because of the proximal airway location of these lesions, bronchoscopy and biopsy can provide a definitive diagnosis. CT scan of the chest may be helpful in further localization of the tumor, and in excluding lymphadenopathy and peribronchial extension of the neoplasm. Bronchial GCTs can infiltrate locally into the peribronchial tissue, which makes it difficult to excise them endoscopically.\(^4\,^10\,^11\,^22\) There is a high recurrence rate when these tumors are removed either endoscopically or by laser therapy.\(^9\,^23\,^24\) However, a recent article reported successful use of an Nd:YAG laser to remove a small GCT in an elderly patient, with no recurrence.\(^25\) It is suggested that lesions of 8–10 mm or larger should be surgically removed.\(^23\,^24\) Pneumonectomy and lobectomy should also be done for tumors associated with profound destruction of the distal lung tissue.\(^5\,^23\) In our patient, there was complete obstruction of the bronchus intermedius, and atelectasis of both right middle and right lower

### TABLE 1—List of Pediatric Patients Younger Than 18 Years of Age With Bronchial Granular Cell Tumors

<table>
<thead>
<tr>
<th>Reference</th>
<th>No. of patients</th>
<th>Age (in years)/sex</th>
<th>Location</th>
<th>Treatment</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Kramer(^14) (1939)</td>
<td>1</td>
<td>15/F</td>
<td>RLLB</td>
<td>Excision by endoscopy</td>
<td>Patent bronchus for 5 months</td>
</tr>
<tr>
<td>Kommel and Bernstein(^16) (1960)</td>
<td>1</td>
<td>8/F</td>
<td>RMB</td>
<td>Excision by endoscopy</td>
<td>Asymptomatic for 18 months</td>
</tr>
<tr>
<td>Oparah and Subramanian(^11) (1976)</td>
<td>1</td>
<td>19/M</td>
<td>LUL</td>
<td>Lobectomy of LUL</td>
<td>Asymptomatic after 3 yrs</td>
</tr>
<tr>
<td>Sawada et al.(^15) (1981)</td>
<td>1</td>
<td>5/F</td>
<td>RMB</td>
<td>Right pneumonectomy, external radiotherapy,</td>
<td>Asymptomatic after 12 yrs</td>
</tr>
</tbody>
</table>

\(^1\) yrs; years; RMB, right main bronchus; RLLB, right lower lobe bronchus; LUL, left upper lobe bronchus; M, male; F, female.
lobes. Removal of these two lobes was undertaken because of the bronchiectatic changes distal to the obstruction. Since her surgery, the patient has been followed in the immunology clinic and has been given multiple courses of anti-inflammatory and cytotoxic drugs for her JRA. No recurrence of the tumor has been reported in 6 years.

In summary, we have described an 8-year-old black female with JRA, a history of recurrent pneumonitis, and persistent right lower lobe atelectasis caused by obstruction of the bronchus intermedius by a GCT. This is one of the youngest patients described with this primary benign tumor. Surgical excision is recommended for this type of tumor, especially for large lesions.

REFERENCES