Sleeve Lobectomy of Mucoepidermoid Carcinoma in a 5-year-old Girl

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A 5-year-old girl presented to our hospital with prolonged pneumatic symptoms over 3 months. After a complete work-up, she was diagnosed with endobronchial mucoepidermoid carcinoma and treated with a left upper sleeve lobectomy. The patient is cured and doing well, 8 months after the surgical resection.

Keywords: endobronchial tumor, mucoepidermoid carcinoma, sleeve resection

Introduction

Primary lung tumors are unusual in childhood, comprising only 0.2% of all malignancies in children. Due to nonspecific symptoms, the endobronchial tumor is challenging to diagnose and manage for pediatric pulmonologists. Children with recurrent pneumonia or prolonged pneumonia should be suspected of having endobronchial lesions. Mucoepidermoid cancer, which is an uncommon endobronchial tumor, comprises less than 10% of all pediatric pulmonary malignancies.1) Although pediatric patients with mucoepidermoid tumors usually have a good prognosis, early diagnosis and complete resection are essential for better outcomes. We report a 5-year-old girl with a recurrent pneumonia due to endobronchial mucoepidermoid cancer.

Case Report

A 5-year-old girl presented with a 1-month history of coughing and recurrent respiratory distress although she had been treated with oral antibiotics before admission. Her breathing sounds were diminished in the left upper lung field. She had no fever, however, laboratory tests revealed a white blood cell count of 20470/mm³. A chest X-ray (CXR) demonstrated atelectasis of the left upper lung field, and a computed tomography (CT) scan and bronchoscopy were then performed. A CT scan showed atelectasis on the left upper lobe and an endobronchial mass on the left main bronchus (Fig. 1A), and bronchoscopy revealed a tumor that totally obstructed the left upper lobar bronchus. The mass was located near the left upper lobar bronchus and protruded into the left main bronchus (Fig. 1B). A biopsy of the tumor suggested mucoepidermoid carcinoma. On the third day after admission, the patient’s respiratory distress was aggravated, and a completely collapsed left lung was identified by CXR (Fig. 1C). The patient experienced a short period of aggravated symptoms, which were immediately relieved, soon after massive physiotherapy. We performed urgent surgical management. The trachea and right bronchus were intubated with a 4.5-mm single-lumen endobronchial tube. Right single-lung ventilation was maintained with FiO₂ at 0.5, tidal volume at 8 ml/kg, respiratory rate at 15 breaths/minute and peak inspiratory pressure at lower than 25 cmH₂O. A left posterolateral thoracotomy was performed through the fifth intercostal space. Operative findings showed a protruding mass, originating from the left upper lobe orifice. A left upper sleeve lobectomy was performed instead of the pneumonectomy, in order to eradicate the tumor. This surgical procedure resulted in margins free of tumor involvement. Histologically, the
tumor was a low-grade mucoepidermoid carcinoma originating from the bronchus composed of predominantly well differentiated mucous cells with occasional intermediate cells (Fig. 2). The patient was discharged 7 days after surgical management with an uneventful postoperative period. Because the tumor was completely resected with clear margins and was proven to be low-grade by the histological examination, a long-term follow-up without chemotherapy or radiotherapy was indicated. At the last visit, 8 months after the resection, the girl remained free of disease.

Discussion and Conclusion

We experienced a rare case of bronchial obstructing mucoepidermoid carcinoma in a 5-year-old child, and here, we have to emphasize two important points in the management of this patient.

First, it is challenging to diagnose an endobronchial tumor in a child. If a child has recurrent pneumonia or prolonged pneumonia, however, an airway obstructing mass must be considered. Therefore, Curtis et al. have suggested that any child who presents with persistent, unexplained consolidation that fails to resolve within 2 weeks of vigorous physiotherapy and antibiotics should be referred for early bronchoscopy.2) Because mucoepidermoid carcinoma, in particular, has a predilection toward appearing in the main stem bronchus or the proximal portion of the lobar bronchus, even an ordinary chest x-ray can be sufficient to identify atelectasis or consolidation due to obstruction.3) In this case, the patient had an unexplained, long-lasting upper respiratory tract infection that was managed conservatively, and then the definitive diagnosis was made, even after she complained of dyspnea on exertion. Because lung tumors occur so rarely in children and adolescents, tumors are not a part of the clinical consideration in general, and this may result in a delayed diagnosis. The mean delay between symptom appearance and diagnosis is around 5 months.4) This fact emphasizes the value of bronchoscopy or computed tomography, which should be promptly performed in all cases with persistent or recurrent respiratory symptoms. Second, it this tumor needs to be resected surgically. Pediatric physicians are often reluctant to perform radical surgical resection for endobronchial tumors in small
children due to sequelae, particularly if mass is diagnosed as benign. Some investigators suggest that bronchoscopic resection should be done for small lesions that are benign, according to the histological examination.\textsuperscript{5)} However, even though primary lung tumors in children are very rare, two-thirds of these tumors are malignant or potentially malignant. Therefore, bronchoscopic removal of these tumors is becoming more attractive along with simpler and less invasive procedures, though endoscopic excision does not guarantee radical excision and the tumor may recur locally or metastasize to a distant site. Surgical resection is thought to be the primary choice of treatment for this tumor. In the past, many patients underwent pneumonectomy for such tumors in the main stem, however, recent parenchymal-preserving methods such as sleeve resection or circumferential resection ensure histologically negative margins without serious complications.\textsuperscript{50) In the current era, video-assisted thoracoscopic surgery is also possible depending on the patient’s age.

In conclusion, differential diagnosis of recurrent or prolonged pneumonia in childhood should include endobronchial tumors, and because endoscopic resection does not ensure a sufficiently radical resection, parenchymal preserving surgical resection is the most preferred treatment.

References