Patient Report

Mucoepidermoid carcinoma of the bronchus: A rare entity in childhood

ARIF KUT,1 BULENT KARADAG,1 FAZILET KARAKOC,1 REFIKA ERSU,1 BEDRETTIN YILDIZELI,2 ESIN KOTILOGLU,3 MUSTAFA YUKSEL2 AND ELIF DAGLI1
Departments of 1Pediatric Pulmonology, 2Thoracic Surgery and 3Pathology, Marmara University Hospital, Istanbul, Turkey

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Primary neoplasm of the lung is very rare in childhood.1 Mucoepidermoid cancer (MEC) of the bronchus is composed of an admixture of mucus secreting, intermediate and squamous cell types.2 Although the clinical features of the bronchial MEC are identical to those in adults, such as recurrent pneumonia, cough and/or hemoptysis, the diagnosis is often unsuspected in children because of the rarity of the condition. Here, we report a 13-year-old boy with MEC located at the left lower lobe orifice that presented with recurrent pneumonia.

Case report

A 13-year-old boy was referred to our clinic for the evaluation of the two episodes of pneumonia in the left lower lobe during the preceding 8 months. Each episode was characterized by cough, fever and left lower lobe consolidation on the chest X-ray which improved over a period of 2 weeks of intravenous antibiotic treatment in the hospital.

The patient’s medical history was unremarkable except for these two hospitalizations. His body temperature was 38.5°C, heart rate was 130 beats/min, respiratory rate was 38 breaths/min and blood pressure was 100/60 mmHg. Auscultation of the chest revealed diminished breath sounds in the left lower lung field. The remainder of his physical examination was normal. Laboratory values included a hemoglobin level of 10.4 g/dL, a white blood cell count of 30,400/mm³, a platelet count of 414,000/mm³ and a C-reactive protein concentration of 48.0 mg/dL. The admission chest roentgenogram showed left lower lobe consolidation (Fig. 1). Computed tomography demonstrated left lower lobe atelectasis, consolidation spreading from left main bronchus to the entire left lower lobe. Multiple enlarged mediastinal and hilar lymph nodes were also observed (Fig. 2). After 10 days of intravenous antibiotic treatment, the symptoms resolved and fiberoptic bronchoscopy was performed which revealed a soft polypoid mass at 3 cm distal to the left main bronchus occluding the entire lumen. Preoperative bronchoscopic biopsy diagnosis was squamous cell carcinoma. The patient underwent left thoracotomy and lower lobectomy with removal of the left hilar and peribronchial lymph nodes. A polypoid mass, 1.5 cm at its greatest diameter, was seen partially obliterating the mainstem of the left lower lobe bronchus. Its peduncle had a broad base. The cut section was glistening and bluish-pink. Microscopic study revealed a solid tumor composed of cells with round, hypochromatic nuclei and either clear or eosinophilic and granular cytoplasm. There were slight variations in nuclear size but hyperchromasia, prominent nucleoli or mitoses were not seen. Intracytoplasmic mucin production and small to big glandular spaces filled with mucin were seen in a minor proportion of the tumor (Fig. 3). No single cell keratinization or pearl formation was seen. The overlying surface epithelium showed squamous cell metaplasia with focal erosions. The lesion was accepted as intermediate grade due to the larger proportion of the epidermoid component. Lymph nodes were free of tumor cells. Pathological diagnosis was intermediate grade mucoepidermoid carcinoma.

The postoperative course was uncomplicated. Full expansion of the left upper lobe was achieved and the patient was discharged 9 days after surgery. The patient has been well and free of recurrence for 6 years postoperatively and no sequelae of the lobectomy and removal of lymph nodes were observed.

Discussion

Primary lung tumors in children are very rare.1 A variety of tumors may arise from submucosal glands of the
tracheobronchial tree. They are identical to tumors of the salivary glands and constitutes less than 0.5% of the primary lung tumors. The MEC, accounting for about one-third, is the second most common submucosal gland tumor and is less than 0.2% of all primary tumors. In children under 16 years of age, only 74 cases of tracheobronchial MEC have been reported previously. As in our case, most of the patients were male (44 patients, 60.3%). The symptoms resulted from the associated bronchial obstruction in all patients and typically included pneumonia or recurrent pneumonia (56%), cough (48.7%), fever (17.8%) and less commonly, hemoptysis (16.2%). Our patient could be diagnosed after two lower respiratory tract infections in an 8 month period. Marked delay in the correct diagnosis may be encountered due to the lack of specificity of the symptoms.

Chest radiographs are usually normal in cases of tracheal MEC but most bronchial lesions present with recurrent or lobar infiltrates, atelectasis or bronchiectasis due to partial or complete endobronchial obstruction. Detailed investigations such as computed tomography or bronchoscopic examination are usually required for the diagnosis of the tumor. Fiberoptic bronchoscopic examination and biopsy are helpful in determining the cause of the respiratory symptoms and also unexpected lesions. This approach may prevent the delay in diagnosis.

In our case, computed tomography of the chest failed to visualize the tumor due to diffuse pneumonic consolidation and flexible bronchoscopy was diagnostic. Cytological studies of sputum, bronchial washings and brushings are not helpful in establishing the diagnosis of mucoepidermoid tumor because the lesion is usually covered by normal respiratory epithelium, and forceps biopsies must be performed. Macroscopically, MEC is polypoid covered with respiratory mucosa and is most frequently located in the left main bronchus (17 cases, 23.4%). On microscopic examination, epidermoid areas are made up of cells without marked pleomorphism, with a large amount of cytoplasm and normal nuclei. Mucus producing cells are also present. Similar to MEC elsewhere, they can be divided into high and low grades which closely relates to prognosis. High-grade variant of MEC is rarely seen and may be difficult to differentiate from typical squamous carcinoma of the lung when its bulk is composed of epidermoid cells with true squamous differentiation. In the presented case, bronchoscopic biopsy specimen was initially diagnosed as squamous cell carcinoma because
only the epidermoid cells were represented in the whole sample. The correct histological diagnosis could only be achieved by the examination of the tumor mass, obtained by surgery. As the specimen represented both elements of previously reported low and high grades, the tumor was labeled as intermediate grade, as two cases had reported previously.\textsuperscript{7,8}

Treatment of bronchial MEC requires complete removal of the lesion while sacrificing as little of the normal lung tissue as possible. Although a sleeve resection of the involved bronchus is recommended, in many cases lobectomy will be necessary for the total removal of the lesion because of its location. Endoscopic resection was not recommended due to the risk of incomplete removal of the neoplastic tissue and possibility of recurrence.\textsuperscript{3} However, there was a recent report of successfully performed video-assisted sleeve lobectomy and this may be another treatment option.\textsuperscript{9} The majority of low-grade MEC have a good prognosis. Only one of the patients died during the follow up.\textsuperscript{10} Our case underwent left thoracotomy and lower lobectomy with removal of the left hilar, mediastinal and peribronchial lymph nodes and the postoperative course was uncomplicated. The patient has been well and free of recurrence for 6 years postoperatively as in the majority of cases.

In conclusion, the bronchogenic MEC is very rare in the pediatric age group, but it must be considered in the differential diagnosis of persistent pulmonary symptoms which do not respond to medical treatment. Use of flexible bronchoscopy widely in pediatric respiratory medicine will make the early diagnosis of such cases possible and the conservative surgical excision is the recommended treatment.

References