Case Report

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Mucoepidermoid carcinoma of the lung in a 6-year-old boy

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ABSTRACT

Primary malignant lung tumours, especially the mucoepidermoid cancer of the bronchus, are very uncommon in childhood. Obtaining the diagnosis might be difficult due to unspecific initial symptoms but early detection and treatment is crucial for a good long-term survival. Bronchoscopy is considered the "gold standard" for making the diagnosis. The recommended therapy for a mucoepidermoid lung cancer is sleeve lobectomy with favourable overall survival after complete resection. We report the case of a 6-year-old boy with a right-upper-lobe bronchus tumour. The histological examination revealed a low-grade mucoepidermoid carcinoma.

Key words: Endobronchial tumour, mucoepidermoid carcinoma, sleeve resection

INTRODUCTION

Lung tumours are extremely rare paediatric diseases, comprising only 0.2% of all malignancies in children.[1] The main pathological spectrum includes endocrine tumours (51.6%), sarcomas (11%) and mucoepidermoid carcinomas (9%) and therefore it is completely different to that of adults.[2,3]

Mucoepidermoid carcinoma, defined by the World Health Organisation as a malignancy consisting of mucous-secreting, squamous and intermediate cell types, usually arises in the salivary glands but can also be found in other organs as lacrimal sac, sinonasal tract, larynx, thyroid and bronchi.[4]

The mucoepidermoid carcinoma of the bronchus has an endobronchial and polypoidal growth pattern. It originates from mucous glands of the main or lobar bronchi as a soft, fragile mass that may induce recurrent respiratory tract infection due to bronchial obstruction.[5]

The vast majority of mucoepidermoid lung carcinomas are low-grade malignancies with an excellent prognosis after surgery.[3] However, early recognition of the disease is essential for a good outcome, but might be difficult because of unspecific initial symptoms and the rarity of the tumour.

We present the case of a 6-year-old boy with a partial lobe atelectasis caused by an endobronchial mucoepidermoid carcinoma. The patient was successfully treated by lobectomy with sleeve resection of the bronchus.

CASE REPORT

A 6-year-old boy was admitted to the hospital for further diagnostic workup and treatment because of a partial atelectasis of the right upper lobe.

The patient was in good state of health until 7 weeks earlier, when acute stridor and dyspnoea developed. Within the next month he had recurrent episodes of febrile bronchitis treated on an outpatient basis with antibiotics. He was referred to a pulmonologist and started on a fluticasone/salmeterol oral inhaler and betamethasone oral. Clinical symptoms improved but auscultation still revealed wheeze-like sounds. The chest X-ray done 2 weeks before admission showed a partial right-upper-lobe atelectasis and hyper-aerated right middle and inferior lobes with a mediastinal shift to the left [Figure 1].

The patient lives in a non-smoking family. There is a positive history of asthma and multiple allergies for his father but no other diseases, especially cancer, are known. The patient has an uneventful medical history.

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Physical examination at the time of admission showed a normal age-related developed, healthy-appearing boy (weight: 22 kg, height: 122 cm). No rash, petechiae, splinter haemorrhages or lymphadenopathy was found. The head and neck were normal. Diffuse inspiratory wheezing and decreased breath sounds were heard over the right lung. The heart was normal. On abdominal examination, the liver and splenic tip were not felt; no costo-vertebral-angle tenderness was detected. No peripheral oedema, cyanosis or digital clubbing was found. The genitalia were normal. Rectal examination was negative. Neurological examination was negative. The temperature was 36.5°C and the pulse rate was 112 beats/min. The blood pressure was 100/60 mmHg.

Urine examination was normal. The haematocrit was 39.7%, the white cell count was 10,000 and the platelet count was 363,000. Electrolytes, creatinine, urea, transaminases, total bilirubin and C-reactive protein were within the normal limits. Blood tests to rule out a primary immunodeficiency (immunoglobulin G, -A, -M, -E; IgG1-4 and alpha-1 antitrypsin) were normal. Sweat test and Mantoux test were negative. A throat swab obtained for culture was negative.

Radiograph of the chest taken at the day of admission was similar to the one done 2 weeks earlier [Figure 1]. The patient underwent a rigid bronchoscopy on the first hospital day. It showed a whitish, rounded tumour located in the upper part of the right bronchus [Figure 2]. Biopsy specimens were taken. On the 2nd day a computed tomography (CT) scan of the chest was performed [Figure 3]. It confirmed the known findings and no other lesions were found.

The biopsy specimens were highly suspicious for a malignant tumour. A second rigid bronchoscopy was performed and most of the tumour was removed. The origin of this lesion was the right-superior-lobe bronchus. A control bronchoscopy 2 days later showed both main bronchi free but the rest of the tumour tissue located at the beginning of the right-superior-lobe bronchus. Histology revealed a low-grade bronchial mucoepidermoid carcinoma.

A combined positron emission tomography and computed tomography (PET-CT) scan showed no 18F-fluorodeoxyglucose (^{18}FDG) uptake at the side of the incompletely removed tumour. No positive lymph nodes or further lesions were found. A magnetic resonance imaging of the abdomen and pelvis and another CT scan of the chest were performed to complete the tumour staging. Again, no metastases or suspicious lymph nodes were found. The chest CT showed the known remaining tumour with its origin at the right-superior-lobe bronchus. Again, this was confirmed by a bronchoscopy [Figure 4]. Further pre-operative workup included a body plethysmography, which showed normal lung function.

A right-upper-lobe sleeve resection was performed to eradicate the tumour. Intraoperative frozen section analysis revealed tumour-free margins. Pathological examination of the lobectomy specimen showed a relarively well differentiated mucoepidermoid carcinoma with infiltration of the bronchial cartilage without reaching the outer bronchial margin [Figure 5a, b]. The pTNM tumour classification was pT1a, pN0 (0/2), pMx, pR0, pL0 and pV0. Grading: G1.

The post-operative course was uneventful. The chest drain was removed on the 6th post-operative day. Control bronchoscopy 2 weeks after operation showed



Figure 1: Chest radiography done 2 weeks before admission with partial right-upper-lobe atelectasis and hyper-aerated right middle and inferior lobes with mediastinal shift to the left

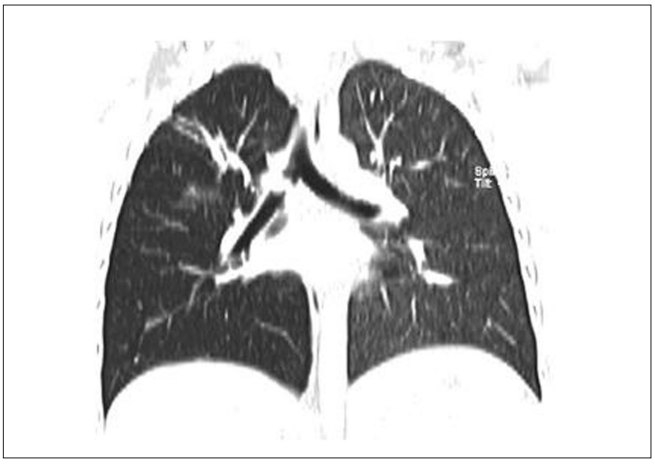


Figure 2: Initial computed tomography scan: tumourous lesion (diameter 1.6 cm) located in the upper part of the right bronchus

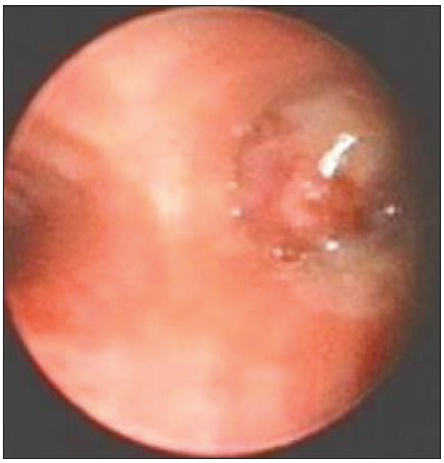


Figure 3: Initial bronchoscopy: whitish, rounded tumour located in the right main bronchus just below the tracheal bifurcation

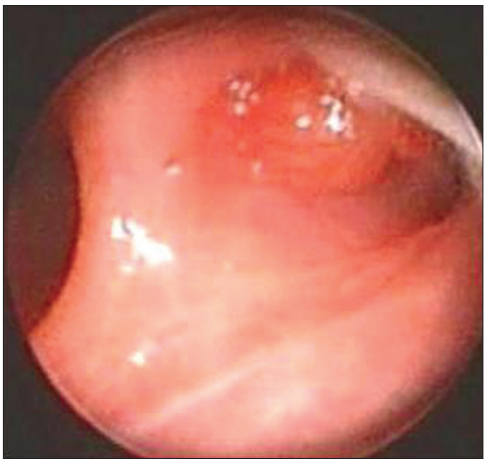


Figure 4: Bronchoscopy prior to sleeve resection: remaining tumour tissue at the beginning of the right-upper-lobe bronchus

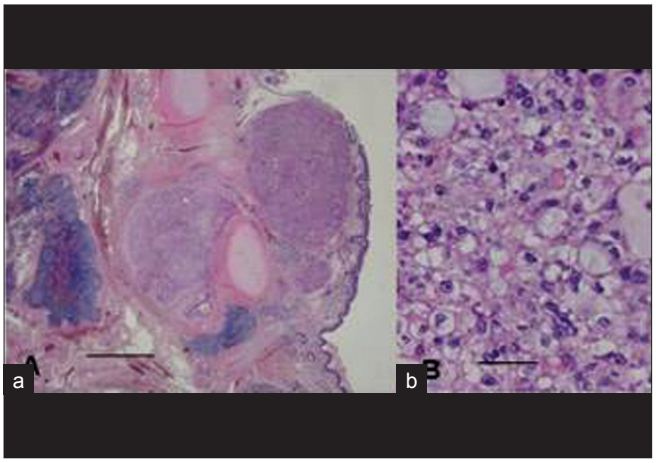


Figure 5: (a) Relarively well differentiated mucoepidermoid carcinoma with infiltration of the bronchial cartilage (HE, bar 1mm), (b) Mucoid differentiated part of the tumor (HE, bar 25 µ)

minimal granulation tissue at the bronchial anastomotic sites. The patient was discharged home on the 15th post-operative day.

Two follow-up bronchoscopies 4 and 6 months after surgery and 2 follow-up CT scans 2 and 6 months after surgery showed no evidence of recurrence.

DISCUSSION

Mucoepidermoid carcinoma of the lung was reported in patients with a wide range of age from 3 to 78 years, but it appears that most of the cases are generated from the paediatric population.[6] In this age group it accounts for approximately 10% of all primary lung tumours.[3] Because of the typical endobronchial growth in large airways, the clinical symptoms may include upper respiratory tract infection or pneumonia, cough, fever, haemoptysis and wheezing.[7] Especially in patients with recurrent airway infections in the same lung region, suspicion of an endobronchial lesion should be raised. Therefore, it was suggested that any child with persistent, unexplained pulmonary consolidation that fails to resolve within 2 weeks of therapy should undergo an early bronchoscopy.[8] An ordinary chest radiography is usually sufficient to identify atelectasis, consolidation or a large pulmonary lesion, but in case of a small endobronchial tumour without airway obstruction no significant information might be provided by this imaging.[9] In our case the primary lesion and the remaining tumour after initial bronchoscopic resection could be clearly detected by CT scan but again a small size lesion might be missed by this imaging. The PET-CT scan showed no ^{18}FDG uptake of the rest of the tumour, but this was after bronchoscopic resection of most of the malignant tissue. There is evidence of increased ^{18}FDG uptake in mucoepidermoid lung carcinoma as reported in an adolescent girl.[10] However, CT and/or PET-CT are helpful imaging tools for tumour staging, but bronchoscopy with biopsy seems to be the method of choice to make the primary diagnosis.

The recommended treatment of a low-grade mucoepidermoid carcinoma with localized disease is complete surgical resection with the sacrifice of as little normal lung as possible.[7] In our case a right-upper-sleeve lobectomy was performed. Ten out of 10 lymph nodes were free of tumour. For a low-grade mucoepidermoid carcinoma no adjuvant therapy is necessary. The 10-year survival after surgery for this type of cancer was reported with 100% (14 patients).[3] Never the less, there is evidence that even low-grade mucoepidermoid carcinomas can be associated with lymph node metastasis, and therefore a close follow-up is recommended.[11]

Our patient follow-up protocol includes physical examinations every 3 months for the first 2 years post-operatively and every 6 months for post-operative years 3 and 4. Five years after a surgery, annual follow-ups will be scheduled. In addition, post-operative surveillance includes CT scans and bronchoscopies 2, 6 and 12 months after surgery.

Now 8 months after tumour excision, our patient is doing well with no evidence of recurrence.

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