CASE REPORT

Rare cause of lung atelectasis in a young woman

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SUMMARY

Pulmonary mucoepidermoid carcinoma is an extremely rare intrathoracic malignancy, comprising less than 1% of all lung tumours. These are very slow growing and are classified into low grade and high grade based on histological features. Surgical resection is the primary treatment with excellent outcomes, while chemotherapy or radiotherapy effectiveness is not known. Preoperative fluorine-18 fluorodeoxyglucose positron emission tomography/CT (18F-FDG PET/CT) is useful for predicting tumour grade and postsurgical prognosis. A clinical case of a 31-year-old woman who presented with dyspnoea on exertion, cough and wheezing is reported. Imaging studies revealed a mass involving the left lower lobe bronchus and atelectasis. 18F-FDG PET/CT showed uptake in the described mass with a maximum standardised uptake value of 9.7. Complete surgical resection was performed, and pathological examination revealed a high-grade mucoepidermoid carcinoma with tumour-free margins. Adjuvant chemotherapy was given and there is no evidence of tumour recurrence.

BACKGROUND

Pulmonary mucoepidermoid carcinoma (PMEC) is an uncommon salivary gland-type tumour and there are few case reports since it was first described by Smetana *et al.*¹ It accounts for 0.1%–0.2% of all lung cancers.^{2,3}

PMEC is slow growing and is usually presented as an intraluminal mass causing luminal occlusion. The symptoms are associated with the airway obstruction.^{2,4}

The tumour is classified into low and high grade. Low-grade tumours are usually confined to the bronchial wall, while high-grade tumours can extend into the adjacent lung parenchyma.^{4–6}

Complete surgical resection is the treatment of choice. The exact role of chemotherapy or radiotherapy is not known, but combination chemotherapy has been reported to be effective when administered as adjuvant treatment after surgery.⁷

CASE PRESENTATION

A 31-year-old Caucasian woman without a significant medical history presented with increasing dyspnoea on exertion, dry cough and occasional wheezing, since June 2014.

She was prescribed by her general physician with inhaled bronchodilators that she rarely used, without any relief of the symptoms.

In May 2016, she got clinically worse and her chest radiograph was compatible with a pneumonia in the left lung. She took 2 weeks of antibiotics with

clinical improvement but only partial resolution of the radiological changes.

It was decided to perform CT of the chest which showed an endobronchial mass with 9.5 mL in the left lower lobe bronchus, associated with consolidation and atelectasis of the lung (figure 1).

She was sent to an appointment in the respiratory department of our hospital in July 2016.

She worked as a business manager and was a non-smoker. There was no significant family history.

INVESTIGATIONS

Flexible fiberoptic bronchoscopy demonstrated a round, smooth, well-circumscribed tumour that almost fully occupied the luminal space of the left lower lobe bronchus orifice.

For maintaining airway patency, endobronchial intervention with rigid bronchoscopy was performed and it was possible to destroy and resect the tumour. The biopsy of the tumour was compatible with low-grade mucoepidermoid carcinoma of the lung.

Fluorine-18 fluorodeoxyglucose positron emission tomography/CT (18F-FDG PET/CT) scan showed tracer uptake in an irregular mass in the left lower lobe with a maximum standardised uptake value (SUVmax) of 9.7 (figure 2).

The lung function tests were normal, and it was decided to proceed with surgery. The patient underwent a thoracotomy with left lower lobectomy and mediastinal lymph node dissection. Pathological examination revealed a 2.5 cm grey-white mass in the lung parenchyma that involved the left lobar bronchus, microscopically compatible with a high-grade mucoepidermoid carcinoma (figures 3 and 4). All resection margins were negative for tumour involvement, and the lymph nodes were free of metastatic disease (pT1cN0).

TREATMENT

As the surgery was after the endoscopic treatment, the thoracic oncology multidisciplinary clinical team proposed that the mass would initially have higher dimensions and, as it caused atelectasis, it was assumed that the initial tumour stage would be IB (T2aN0M0) according to the seventh edition of the tumour, node, metastases (TNM) classification for lung cancer.

Considering the histological grade, tumour stage and age of the patient, the multidisciplinary team decided to perform adjuvant chemotherapy with carboplatin and vinorelbine.

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Figure 1 (A) Visible endobronchial mass on the left lower bronchus; (B) Collapsed left lower lobe.

OUTCOME AND FOLLOW-UP

The patient did four cycles of adjuvant chemotherapy and remains asymptomatic without evidence of tumour recurrence at 22 months of postsurgical follow-up.

DISCUSSION

PMEC is an extremely rare tumour of the lung. There is no gender preference and it can affect any age group, although approximately half of the cases occur under 30 years of age.^{4,8}

Histologically, the tumour arises from the serous and mucous gland of the trachea and bronchi and it is composed by mucin-secreting, squamous and intermediate-type cells.^{2,8,9}

PMEC can be subclassified into low grade and high grade. The high-grade tumour is rarer and consists of atypical squamoid and intermediate cells, with mitosis and necrosis, and a number of mucin-secreting cells that is variable. It is usually more infiltrative and presents an exophytic endobronchial growth.⁸

Because the tumour frequently involves large airways, the clinical symptoms and signs of the disease are related to bronchial obstruction and consequent lung atelectasis, which will eventually cause obstructive pneumonia as occurred with our patient. PMECC patients usually present a cough, wheezing and haemoptysis.^{2,4}

Our patient's diagnosis was delayed for 2 years, which is frequent in this disease due to the slow growth of the tumour with small changes on thoracic imaging during the early stage, and due to the non-specific symptoms.¹⁰

Figure 2 Mass in the left lower lobe showing with an SUVmax of 9.7 using 18F-FDG PET/CT. 18F-FDG PET/CT, fluorine-18 fluorodeoxyglucose positron emission tomography/CT; SUVmax, maximum standardised uptake value.

Figure 3 High-grade MEC showing a solid squamous component, with areas of an oncocytic-like appearance, admixed with cystic spaces, desmoplasia and areas of necrosis and haemorrhage (circle). (H&E). MEC, mucoepidermoid carcinoma.

Although most cases of mucoepidermoid carcinoma can be confidently diagnosed with an endoscopic biopsy, misdiagnoses are a possibility because of tissue fragmentation and stroma predominance.⁴ This is shown in our patient whose initial diagnosis through endoscopic biopsy was low-grade mucoepidermoid carcinoma, but after surgery we concluded that the tumour was actually high grade.

Complete surgical resection with curative intent should be the first treatment option in these patients. The evidence to support the effectiveness of chemotherapy or radiotherapy is scarce.^{2,5,7,11} Combination chemotherapy has been reported to be administered as adjuvant treatment after surgery in patients with high-grade malignancy, and it was shown to be effective. In low-grade malignancy, due to their favourable prognosis, it is not recommended.⁷

In the case of our patient, it was first performed interventional bronchoscopy for tumour debulking, which is known to be effective for maintaining airway patency, but surgical resection

Figure 4 Higher magnification of the squamous component with nuclear pleomorphism and mitotic activity (arrows), typical of a high-grade MEC. (H&E). MEC, mucoepidermoid carcinoma.

is still required because of the remaining residual tumour within the bronchial wall after endobronchial intervention.^{7 12}

Independent factors that influence long-term prognosis are described for PMEC patients. The tumour histological grade, TNM stage and lymph node metastasis seem to be associated with their survival.^{3 6 11 13}

Recently, Park *et al* stated that 18F-FDG PET/CT could be a valuable imaging modality for predicting the tumour grade and stage and the postsurgical prognosis. They concluded that patients showing an SUVmax greater than 6.5, as the case described, would be more likely to have severe disease. They frequently present a high-grade tumour, lymph node metastasis and disease recurrence. Consequently, these patients might need a more aggressive surgical approach like extensive mediastinal lymph node dissection and the use of adjuvant therapies.¹⁴

Learning points

- Pulmonary mucoepidermoid carcinoma (PMEC) usually presents with non-specific signs and symptoms similar to other obstructive diseases.
- Endobronchial obstruction should be considered in case of chronic or recurrent respiratory symptoms, and a chest CT scan ±bronchoscopy should be performed.
- Half of the PMEC cases occur in individuals under 30 years of age.
- PMEC has a favourable prognosis after complete surgical resection.
- PMEC patients with high uptake on fluorine-18 fluorodeoxyglucose positron emission tomography/CT imaging might require aggressive surgical approach and adjuvant therapies.

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involved in patient's follow-up consult and manuscript review. DA performed the histopathology analysis, contributed to the diagnosis of the disease and manuscript review. All authors read and approved the final version submitted.

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