CASE REPORT

Atypical presentation of typical carcinoid

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SUMMARY

Bronchial carcinoid tumours can present with symptoms of distal airway obstruction, local airway irritation or bleeding due to hypervascularity. Presentation with a spontaneous pneumothorax (SP) is rare. We present a case of typical endobronchial carcinoid tumour that presented with a SP. We explored the possible mechanisms of this rare presentation, especially the check-valve mechanism of airway obstruction, which was evident during the fiberoptic bronchoscopic examination.

BACKGROUND

In 1950, Chevalier Jackson explained the check-valve mechanism responsible for pulmonary hyperinflation in the setting of endobronchial tumour.¹ Although pulmonary hyperinflation can result from incomplete endobronchial obstruction, extreme hyperinflation secondary to endobronchial tumour leading to a pneumothorax is rare. In 1990, Wagner *et al* described the first reported case of this phenomenon subsequent to an endobronchial carcinoid tumour that presented with a spontaneous pneumothorax (SP).² Our case supports this rare presentation of carcinoid tumour and provides bronchoscopic evidence of the underlying check-valve mechanism.

CASE PRESENTATION

A 65-year-old man with a remote 10-pack-year smoking history presented with symptoms of mild dyspnoea on exertion and left-sided chest pain. His chest pain was sharp, of sudden onset and

increased with deep inspiration with no relieving factors. The patient had occasional dry cough for few months prior to his presentation with no relief with over the counter medications. He had no family history of lung disease, no pets and no history of recent travels. He denied fever, chills, weight loss or haemoptysis. There was no symptoms of flushing or diarrhoea. On presentation, his blood pressure was 130/77 mm Hg, heart rate was 90 bpm and oxygen saturation was 96% on 2 L of oxygen by nasal cannula. His physical examination was significant for decreased air entry on auscultation over the left upper lobe. Cardiac and abdominal examinations were within normal limits with no murmurs or organomegaly, respectively. He had no palpable lymphadenopathy or clubbing.

INVESTIGATIONS

An ECG, complete blood count, complete metabolic panel including electrolytes, renal and liver function testing were all within normal limits. Chest X-ray showed left apical pneumothorax (on posteroanterior view; figure 1A) and volume loss of the left hemithorax (on lateral view; figure 1B).

Chest CT showed the left apical pneumothorax and an endobronchial lesion within the left upper lobe bronchus with distal collapse of the left upper lobe (figure 2A, B). The positron emission tomography (PET) scan showed the endobronchial lesion to be hypermetabolic with a maximum SUV of 3.6 with no evidence of distant metastasis (figure 3).

The patient underwent flexible fiberoptic bronchoscopy that revealed a pedunculated, reddish,

Figure 1 (A) Chest X-ray (posteroanterior view) showing left apical pneumothorax (arrows). (B) Chest X-ray (lateral view) showing volume loss with elevated left hemidiaphragm and shift of the major fissure anteriorly.

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Figure 2 Chest CT showing left apical pneumothorax (A) and endobronchial lesion in the left upper lobe bronchus (arrow) (B).

rounded, smooth lesion almost obstructing the left upper lobe and the lingula. The endobronchial lesion showed some mobility with a variable degree of bronchial obstruction throughout the respiratory cycle. During inspiration, the tumour would move to allow air entry into the distal airways (figure 4A) and during expiration, the tumour would bulge in the opposite direction leading to complete airway obstruction preventing air exit (figure 4B), also refer to (figure 5 A, B). After injecting epinephrine (1:10 000 dilution) locally in the body of the tumour, multiple endobronchial biopsies were taken.

Figures 3 Positron emission tomography scan showing soft tissue mass extending into the left upper lobe bronchial tree (arrow) with a maximum standard uptake value of 3.6.

DIFFERENTIAL DIAGNOSIS

Endobronchial tumours can be malignant or benign in nature (table 1). The primary malignant tumours can arise from surface epithelium (most common), salivary glands or mesenchyme. The secondary malignant tumours can result either from direct invasion (more common) or from haematogenous metastasis (less common). Most of the benign airway tumours arise from the mesenchymal tissue while a small percentage can rarely arise from surface epithelium. Some CT features of endobronchial lesions can predict the benign nature of the tumour such as the presence of a high fat content in case of endobronchial hamartomas and lipomas.³

Our patient had an endobronchial lesion with an increased uptake on PET scan with no evidence of metastasis. His tumour most likely represents a primary malignant endobronchial tumour. The most common primary malignant airway tumours are those arising from the surface epithelium, which include squamous cell carcinoma, adenocarcinoma and neuroendocrine tumours including carcinoid tumours. The tumour morphology (pedunculated, reddish, rounded, smooth and mobile) favoured carcinoid tumour over bronchogenic carcinoma (non-pedunculated, irregular, fungating with possible ulcerations). The endobronchial biopsies showed uniform cells with granular cytoplasm and stippled nuclear chromatin growing in an organoid nesting pattern (figure 6). Immunohistochemical stains were positive for CD56, synaptophysin and chromogranin. These findings were consistent with typical carcinoid tumour.

TREATMENT

As the patient had no comorbidities, the PET scan showed no distant metastasis and the pulmonary function tests showed forced expiratory volume in the first second of 2.87 L (78% of the predicted) with a normal diffusion capacity, the decision was made to undergo surgical treatment. The patient underwent left upper lobe sleeve resection with bronchial reanastomosis.

OUTCOME AND FOLLOW-UP

The patient recovered from the surgery with no complications and he was discharged home.

DISCUSSION

The 2004 WHO classification recognised four main types of lung neuroendocrine tumours: typical carcinoid, atypical carcinoid, large cell neuroendocrine carcinoma and small cell lung

Figure 4 Bronchoscopic images showing movements of the endobronchial carcinoid tumour allowing air entry during inspiration (A) and preventing air exit during expiration due to complete obstruction of the lumen (B) (check-valve mechanism).

Figure 5 Diagram illustrating the check-valve mechanism due to flapping endobronchial tumour allowing air entry during inspiration (A) and causing air trapping distal to the obstruction during expiration (B).

cancer. This classification depends on the tumour morphological features, mitotic rate and the presence or absence of necrosis.⁴

Bronchial carcinoid tumours account for 1% to 2% of all lung malignancies. They can be centrally located in 85% of cases (10% in main-stem bronchi and 75% in lobar bronchi) or less commonly peripherally located.^{4 5} Almost 80% to 90% of pulmonary carcinoids are typical and they are thought to arise from Kulchitsky cells located within the bronchial and bronchiolar epithelium.⁴

The clinical presentation depends on the site and the size of the tumour. Peripheral tumours are often asymptomatic and they are incidentally discovered. Centrally located tumours usually present with symptoms of airway irritation or obstruction ranging from cough to haemoptysis and recurrent respiratory tract infections distal to the site of obstruction. Less commonly, carcinoid tumours can present with different syndromes such as carcinoid syndrome due to secretion of vaso-active substances, Cushing's syndrome due to secretion of

Table 1 Classification of endobronchial tumours

Malignant tumour					Benign tumours	
Primary			Secondary			
Surface epithelium	Salivary glands	Mesenchyme	Direct invasion	Haematogenous metastasis	Surface epithelium	Mesenchyme
SCC Adenocarcinoma Neuroendocrine Tumours including carcinoids	ACC MEC	Sarcoma Malignant lymphoma	Thyroid cancer Laryngeal cancer Lung cancer Esophageal cancer	Melanoma Renal cell carcinoma Breast cancer Colon cancer	Squamous cell papilloma Papillomatosis Pleomorphic adenoma Mucous gland adenoma Oncocytoma	Hamartoma Lipoma Leiomyoma Fibroma Neurogenic tumour

ACC, adenoid cystic carcinoma; MEC, mucoepidermoid carcinoma; SCC, squamous cell carcinoma.

Figure 6 (A) Typical carcinoid tumour with the arrow pointing to the endobronchial epithelium (H&E, $\times 10$ magnification). (B) Typical carcinoid tumour demonstrates uniform cells growing in a nesting pattern (H&E, $\times 10$ magnification). (C) Carcinoid tumour cells with round to oval nuclei and coarsely granular stippled salt and pepper chromatin (H&E, $\times 20$ magnification).

adrenocorticotrophic hormone or Acromegaly due to growth hormone-releasing hormone secretion.^{5,6}

Pneumothorax is a rare presentation of endobronchial carcinoid tumours. Generally, the incidence of SP due to lung cancer is 0.003 to 0.005. Squamous cell carcinoma is the most common primary lung cancer that can present with SP, and osteogenic sarcoma is the classic pulmonary metastatic tumour that can present with SP especially in children and adolescents.⁷

Possible mechanisms of pneumothorax in lung cancer:

1. Rupture of a necrotic tumour into the pleural space.
2. Subpleural metastasis with subsequent rupture of the necrotic metastatic foci.
3. Check-valve mechanism (important mechanism especially in setting of endobronchial tumours).
4. Fortuitous association due to rupture of underlying emphysematous bullae due to disturbance of lung architecture with bronchial cancer.

The bronchial obstruction resulting from an endobronchial tumour can be classified clinically into three types depending on the mechanism. The bypass-valve mechanism, the check-valve mechanism and the stop-valve mechanism which can result clinically in wheezing, emphysema and atelectasis, respectively (table 2).¹

In our patient, the most likely mechanism for his pneumothorax is the check-valve mechanism. Chevalier Jackson fully explained the mechanism of pedunculated flapping endobronchial tumour causing the phenomenon of alternating emphysema and atelectasis through check-valve and reverse check-valve mechanisms, respectively.¹ The movements of the flapping tumour during bronchoscopy can explain the presentation with pneumothorax due to check-valve action with excessive hyperinflation distal to the site of obstruction.

On chest CT, endobronchial carcinoid tumour appears as a well-defined ovoid lesion with slight lobulation. Occasionally

tumours with a dominant extraluminal component are referred to as 'iceberg tumours'. Carcinoid tumours may demonstrate intense enhancement due to hypervascularity. On PET scan they show variable degree of uptake depending on the tumour proliferation and mitosis.³

In patients with good pulmonary reserve, surgery is the treatment of choice for carcinoid tumours. If the patient cannot tolerate surgical resection or in highly selected cases where the lesion is entirely intraluminal, bronchoscopic removal may be an alternative.⁸

Typical bronchial carcinoid tumours have an excellent prognosis after surgical resection with a 5-year survival rate ranging from 87% to 99% and a 10-year survival rate between 82% and 87%.^{9–11}

Learning points

- Pneumothorax is a rare presentation of endobronchial carcinoid tumours.
- Bronchial obstruction can result clinically in wheezing, emphysema or atelectasis depending on the underlying mechanism which can be bypass-valve, check-valve or stop-valve, respectively.
- Surgery is the treatment of choice for carcinoid tumours.
- Typical bronchial carcinoid tumours have an excellent prognosis after surgical treatment.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

Table 2 Types of endobronchial obstruction

Type of obstruction	Air entry	Air exit	Result
Type I (bypass-valve)	Yes	Yes	Wheezing
Type II (check-valve)*	Yes	No	Emphysema
Type III (stop-valve)	No	No	Atelectasis

*Reverse check-valve may allow air exit with no air entry leading to atelectasis and soon merge into type 3.

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