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Dysphagia due to secondary achalasia as an early manifestation of squamous cell carcinoma

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Summary

A 59-year-old man, a smoker, presented with features of airflow obstruction due to squamous cell carcinoma of central airways mimicking chronic obstructive airways disease. He also had pronounced dysphagia. Computed tomographic and magnetic resonance imaging scans showed mediastinal tumour invasion but no direct oesophageal involvement. Oesophageal manometry studies revealed that dysphagia was due to the oesophageal motility disorder, secondary achalasia.

Keywords: lung cancer, airflow obstruction, dysphagia, achalasia

Early central airway tumours are frequently missed as their presentation often mimics airflow obstruction due to either chronic obstructive airways disease or asthma. A diagnosis is made when the tumour is advanced and associated with other symptoms such as haemoptysis. Dysphagia is usually a late symptom in patients with lung cancer. It is caused by mechanical obstruction of the oesophagus either by extrinsic compression due to mediastinal lymphadenopathy or direct tumour invasion. We describe a patient with central airway lung cancer in whom dysphagia was an early manifestation of the disease caused by a motility disorder.

Case report

A previously healthy 59-year-old male smoker presented with a three-month history of dyspnoea on exertion associated with wheeze and

nocturnal cough. He also complained of dysphagia for solids of three weeks duration and a weight loss of 4 kg. Examination revealed no clubbing of finger nails or enlargement of cervical lymph nodes. He was not dyspnoeic and examination of respiratory system, cardiovascular system and abdomen revealed no abnormal findings. Chest X-ray was normal except for hyperinflated lung fields. Pulmonary function test showed mild airflow obstruction with PEFR of 415 l/min and FEV₁/FVC 2.59/3.67 l (predicted 504 l/min, 3.24/4.03 l). Other tests of pulmonary function such as flow volume loop studies, lung volumes and transfer factor were normal. Oesophago-gastroscopy and barium swallow examination for dysphagia revealed no obstructive lesion or mucosal abnormality.

Airflow obstruction associated with symptoms of exertional dyspnoea, wheeze and cough was suggestive of either chronic obstructive airways disease or late onset asthma and the patient was treated with inhaled beta-2 agonist and corticosteroids and oral corticosteroids. However there was no improvement in the symptoms and two months later the patient presented with further symptoms of hoarseness of voice and haemoptysis. Fibre-optic bronchoscopy revealed squamous cell carcinoma involving airway mucosa of the lower third of the trachea and proximal part of both major bronchi, and paralysis of the left vocal cord. Airway narrowing due to the tumour was confirmed on computed tomography (CT) and magnetic resonance imaging (MRI) scans of the chest, with a cuff of neoplastic tissue seen around the lower part of the trachea and proximal major airways (figure). There was no evidence of local lymph node enlargement or direct oesophageal involvement but tumour

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Accepted 15 February 1995

Figure Sagittal (A) and coronal (B) T1-weighted MRI of the thorax. The medium signal intensity tumour (T) is seen infiltrating around the distal trachea and proximal main bronchi. The surrounding mediastinal fat is of a higher signal intensity (ie, white). A, ascending aorta; AA, aortic arch; L, lung; RA, right atrium; R, right main pulmonary artery; LA, left atrium; IVC, inferior vena cava

A

B

invasion of the mediastinum was noted. However, further investigation of the dysphagia by oesophageal manometry revealed features of achalasia, namely aperistalsis in the oesophageal body, incomplete relaxation of the lower oesophageal sphincter and hypertonic lower oesophageal sphincter. A 24-hour ambulatory pH measurement was within normal range.

Discussion

In the present case, squamous cell carcinoma of the central airways presented with the features of airflow obstruction and mimicked late onset asthma/chronic obstructive airways disease. The diagnosis was made when the tumour caused haemoptysis and hoarseness of voice. However, dysphagia was an early associated

symptom. Dysphagia and hoarseness of voice in patients with lung cancer are usually suggestive of mediastinal spread of the tumour. Symptoms due to mediastinal spread are particularly common in patients with small cell carcinoma because of the high frequency with which this histologic type spreads to the mediastinum and because involved nodes tend to be bulky in comparison with mediastinal involvement by other histological types.¹

We found no evidence of mediastinal lymph node enlargement or direct tumour invasion of the oesophagus and dysphagia was found to be due to oesophageal achalasia. Clinical features of idiopathic achalasia closely mimic secondary achalasia, and can have the same manometric features.² However, it is likely that dysphagia in the present case was due to secondary achalasia as patients with secondary achalasia tend to be older (> 50 years), to have shorter duration of dysphagia (< 1 year) and to have greater weight loss when compared to those with idiopathic achalasia. Furthermore, barium swallow and endoscopic examinations reveal no diagnostic information in dysphagia due to secondary achalasia, as in the present case, when these investigation show features typical of idiopathic achalasia.

Secondary achalasia has been reported with wide variety of tumours such as adenocarcinoma of the stomach and pancreas, hepatocellular carcinoma as well as adenocar-

Features of secondary (carcinoma-induced) achalasia
<ul style="list-style-type: none">• associated with adenocarcinoma of the gastric fundus, carcinoma of pancreas, small cell carcinoma and lymphoma• clinical features of dysphagia of short duration (< 1 year) in patients > 50 years old, associated with marked weight loss• CT/MRI findings of thickening or tumour nodularity of distal oesophageal wall

Causes of secondary achalasia
<ul style="list-style-type: none">• circumferential tumour mass involving the cardia of stomach• disruption of myenteric plexus of oesophagus due to submucosal tumour infiltration• damage to the myenteric plexus due to deposition of eosinophilic cationic protein

Causes of dysphagia in lung cancer
<ul style="list-style-type: none">• mediastinal lymphadenopathy• direct tumour invasion of mediastinum• radiotherapy• secondary achalasia

Learning points
<ul style="list-style-type: none">• dysphasia due to secondary achalasia can be an early manifestation of lung cancer• airflow obstruction due to central airway tumours can mimic chronic obstructive airways disease/late onset asthma

cinoma and oat cell carcinoma of lung.²⁻⁴ In most cases secondary achalasia has been reported due to oesophageal obstruction caused by tumour infiltration or encasement of the oesophagogastric junction. Other suggested explanations include physical disruption of the myenteric plexus by the tumour and deposition of eosinophilic cationic protein associated with the tumour causing damage to the myenteric plexus.^{5,6} Intestinal motility disorders in small cell lung cancer have been reported due to paraneoplastic enteric neuropathy linked to the presence of

antineuronal antibody (also known as anti-Hu antibody).⁷ To our knowledge there are no previous reports of dysphagia caused by secondary achalasia due to squamous cell carcinoma of the lung. In the present case dysphagia due to secondary achalasia was an early feature of central airway squamous cell carcinoma, probably caused by infiltration of the myenteric plexus by the tumour. Secondary achalasia due to squamous cell carcinoma of central airways should be considered in patients with dysphagia without an obvious cause.

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