

CASE REPORT

A Case of Bronchial Carcinoid Presenting With Collapse of Lung, Initially Treated For Wheeze

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ABSTRACT:

A 60 year male patient, smoker since last 30 years, who had been unsuccessfully treated with inhalational β -2 agonist, corticosteroid and anticholinergic for obstructive airway disease by local physician over last three years, presented with gradually progressive breathlessness over one month and mild dull aching left sided chest pain since six months. On clinical examination, patient was found to have features of loss of lung volume on left side and diminished breath sound over entire left hemithorax. Chest X-ray revealed collapse of left lung. CT scan of thorax showed complete left lung collapse with a mass in left main bronchus. Metastatic nodules were found in liver in CT scan of abdomen. Subsequent fiberoptic bronchoscopy demonstrated a polypoid mass in left main bronchus partially occluding the lumen and biopsy from the mass revealed features of carcinoid tumour. Bronchial carcinoid can sometimes present with features of airway obstruction which leads to a misdiagnosis of the case as asthma or COPD which responds poorly to conventional therapy. So, CT thorax and bronchoscopy are recommended in patients with wheeze who cannot be managed adequately with inhalational therapy to exclude other underlying pathology.

Key words: wheeze, collapse, bronchial carcinoid, asthma, neuroendocrine tumour.

INTRODUCTION:

Bronchial carcinoid is an uncommon tumour of respiratory system although it is the commonest benign tumour of tracheobronchial tree. This neuroendocrine tumour usually manifests with haemoptysis and cough; but can present sometimes in an unusual manner with refractory wheeze and breathlessness. Such patients are often mistakenly treated for bronchial asthma or COPD for a long duration before the actual diagnosis is established. We report a case of bronchial carcinoid who presented to us in a very advanced stage of the disease with collapse of left lung and liver metastasis, after being misdiagnosed and treated for obstructive airway disease since three years.

CASE REPORT

A 60 year male patient, farmer by occupation, presented to our OPD with complaints of increased dyspnoea since one month and mild dull aching left sided chest pain since six months. Dyspnoea was of mMRC grade III during presentation, occurring after walking for 20-30 meter at his own pace. Chest pain was poorly localised without any radiation to other site and having no association with respiration or coughing. He also complained of occasional cough with minimal mucoid expectoration. There were loss of appetite and generalised weakness since six months but he denied any history of fever or haemoptysis. Patient was being treated with inhalational formoterol, budesonide and tiotropium since last three years for COPD as there was history of breathlessness and wheezing. He was on regular treatment and never discontinued therapy.

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There was no history of sneezing, rhinorrhoea or atopy. He was non-alcoholic but a smoker since nearly 30 years with 2-3 cigarettes per day and stopped since last three years. There was no other co-morbid illness. Bowel and bladder habit were normal.

On clinical examination, he was of average built. Pallor, cyanosis, icterus, oedema, clubbing and lymphadenopathy were all absent. Respiratory rate was 20/min. Pulse rate, blood pressure, temperature and SpO₂ were within normal limit. There was mild scoliosis with concavity towards left side. Chest examination revealed reduced volume of left hemithorax with shifting of mediastinum to left side and diminished vesicular breath sound over entire left chest. There was no added sound on auscultation.

Initial chest X-ray in postero-anterior view (Figure I) showed collapsed left lung with mediastinum shifted to left along with rib crowding and elevated left hemidiaphragm. Scoliosis of thoracic spine was also evident in chest radiogram. A complete blood count (CBC) showed Haemoglobin 12.0 gram/dl, total white cell count 11,000/mm³ with neutrophil 68%. Other routine blood examinations were within normal limit.

Contrast enhanced CT scan of thorax (Figure II) revealed collapsed left lung with compensatory hyperinflation of right lung plus an intraluminal mass in left main bronchus. Borderline right upper paratracheal node and minimal left pleural effusion were also noted. CT scan of abdomen demonstrated enhancing metastatic nodules in segment II and segment V of liver (Figure III).

Subsequently fiberoptic bronchoscopy was done and a smooth yellowish polypoidal mass (Figure IV) was found in left main bronchus, partially occluding the lumen. A biopsy was taken from the mass which on histopathological examination showed uniform polygonal cells with round nuclei (Figure V) suggestive of carcinoid tumour with features of nuclear atypia and increased mitosis. Immunohistochemistry further confirmed neuroendocrine differentiation of tumour cells with cytoplasmic positivity of cytokeratin and chromogranin A.

Therefore, a diagnosis of atypical carcinoid tumour with mediastinal lymphadenopathy and hepatic metastasis was established and the patient was referred to thoracic surgeon and oncologist for further evaluation and management. Surgical treatment was not considered as an option due to presence of metastatic disease. Patient denied further management with chemotherapy due to financial constraint. He was counselled about poor prognosis of metastatic atypical carcinoid tumour and subsequently he expired after nine months since diagnosis was made.

DISCUSSION:

Pulmonary carcinoid is the most frequently encountered benign tumour of tracheobronchial tree and constitutes 2–5% of all lung cancers¹. Although carcinoid tumour is most commonly located at gastrointestinal system, 25% cases are located within respiratory tract². Carcinoids belong to the neuroendocrine tumour group arising from the APUD (amine precursor uptake decarboxylation) system and share the same neuroectodermal cell of origin with small cell carcinoma of lung.

Typical and atypical carcinoids are subgroups of neuroendocrine tumours that are determined as low-grade and intermediate-grade tumours according to biological aggressiveness respectively. About 90% of carcinoid tumours are well differentiated with rare mitoses, pleomorphism, and necrosis³. These are referred to as “typical” carcinoid tumours. The remaining 10% are characterized by increased mitotic activity, nuclear pleomorphism, and disorganization; these lesions are designated as “atypical” carcinoids. Atypical varieties are more associated with metastasis and poorer prognosis.

Aetiologically, no association was found with cigarette smoking, ambient radiation or exposure to other known carcinogens³ although a recent study described a possible association between atypical carcinoid and smoking⁴. Mean age at presentation of patients with typical carcinoid is in the fifth decade, while atypical carcinoid presents a decade later in life. Carcinoid is commoner in women although our patient was a male in his sixties.

Central tumours (located commonly in right lung) mostly present with haemoptysis (caused by high vascularisation), recurrent lower respiratory tract infections, cough although it can sometimes present with dyspnoea, wheezing or chest pain. Typical carcinoid syndrome, characterized by flushing, palpitations, wheezing and right-sided valvular heart disease, is extremely uncommon with pulmonary carcinoid. But usual symptomatology was not described in our patient who presented with wheezing and there was no haemoptysis in spite of having a centrally located tumour. It delayed the diagnosis by several years and the tumour was eventually diagnosed at an advanced stage with distant metastasis.

So, when a patient comes with refractory asthma or COPD, other causes of non-response to therapy like upper respiratory tract causes (rhinitis, nasal polyp, sinusitis, post-nasal drip), gastro-oesophageal reflux disease, ABPA, airway malignancy, chronic pulmonary infection (specially tuberculosis) and cardiac disorders must be considered.

Radiographic findings depend largely on tumour location⁵. Most carcinoids appear as circumscribed, centrally located lesions with a diameter of 2-5cm^{2,6}. Given the central location of the lesions, associated radiographic findings of post-obstructive pneumonia or mucus plugging are common. Peripherally located lesions occur in approximately one third of cases, and generally are of less than 3 cm in diameter⁶. Endobronchial lesions may also cause distal air-trapping, unilateral hyperlucent lung although X-ray may sometimes be normal. Atelectasis may occur in some patients as happened in our case. Computerised tomography or magnetic resonance imaging is more sensitive, especially in detecting lymph node metastases.

Bronchoscopy can detect centrally located tumour and biopsy can be obtained from it. Although some physicians do not support taking bronchoscopic biopsy due to chance of bleeding, but most of the recent studies as well as British Thoracic Society refer to it as a safe procedure. Endobronchial brushings and washings are typically non-diagnostic because the bronchial epithelium overlying the tumour is typically normal⁷.

Carcinoids may express the neuroendocrine markers by immuno-histochemistry (chromogranin A, synaptophysin etc). Plasma chromogranin A was the most frequently elevated hormone (94%) and thus appears to be the most sensitive tumour marker in patients with metastatic bronchial carcinoids which corroborates with the finding in our patient.

Surgery is the therapy of choice for carcinoid tumours, with parenchyma-sparing procedures are recommended for typical carcinoids². Surgical procedures include pneumonectomy, bilobectomy, lobectomy, segmentectomy, sleeve resection, and wedge resection. The aim is to remove the primary tumour and affected lymph nodes radically, keeping as much of the lung parenchyma as possible. If complete surgical resection cannot be accomplished, or if metastatic disease is present, chemotherapy and radiation therapy provide little hope for cure. Metastatic or locally advanced carcinoids display very poor responsiveness to a variety of chemotherapeutic regimens with a median overall survival of 25 months since treatment initiation⁸. So, early diagnosis is the only key to avoid such a poor prognosis which unfortunately, did not happen to our case. Biotherapy with α -Interferon or somatostatin analogue can be used, especially for progressive disease although their role on tumour growth is limited⁸.

In conclusion, aetiology should be reviewed in patients of obstructive airway disease who are not responding to conventional management. Especially, an endobronchial growth must be suspected if patient presents with unilateral wheeze or features of lung collapse, to avoid unnecessary delay in diagnosis and management.

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Fig.I: Chest X-ray (PA) showing collapsed left lung with mediastinum shifted to left side. Left sided rib crowding, elevated hemidiaphragm and thoracic scoliosis with concavity to left were also noted.



Fig.II: CT scan thorax (lung window) showing left lung collapse with hyperinflated right lung. Mediastinum shifted to left. An intraluminal mass noted within left main bronchus (indicated by arrow).

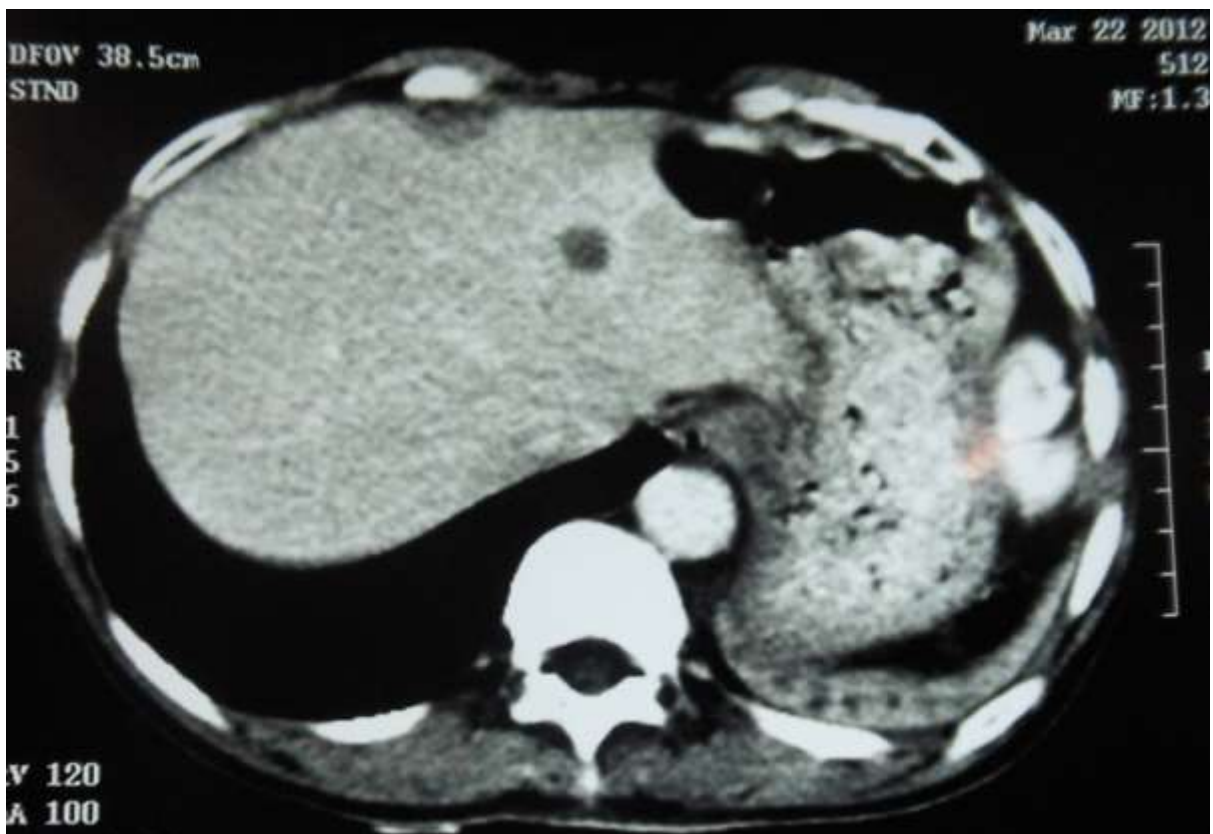


Fig.III: CECT scan of abdomen showing hypodense lesion in liver suggestive of metastasis



Fig IV: Fibreoptic bronchoscopy showing intraluminal mass in left main bronchus

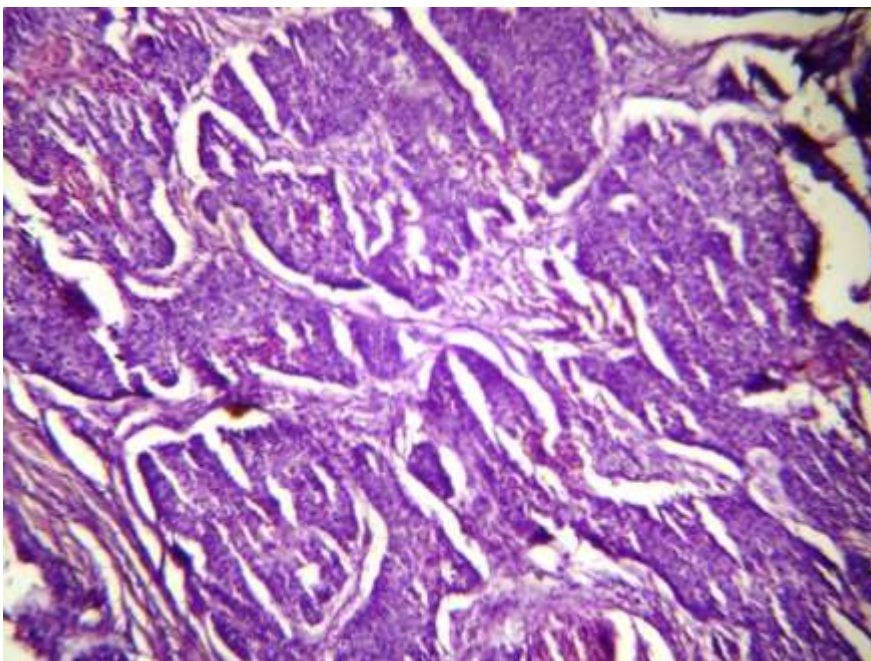


Fig V: Histology showing features of carcinoid tumour. Photomicrograph demonstrating solid nests of monotonous appearing cells with small, round nuclei; moderate amount of finely granular cytoplasm & fine nucleoli.(Hematoxylin& eosin;100X)