CASE REPORT

Huge carcinoid tumour causing complete obstruction of the left main bronchus and destruction of left lung

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Abstract

Carcinoid tumours arise from the neuroendocrine cells of the bronchial epithelium known as Kulchitsky cells and comprise 4% of all lung malignancies. We report a case of a 16- year-old male who presented with a 1-month history of fever, cough, left-side chest pain and shortness of breath. Chest X -ray showed loss of lung volume on the left side with cystic spaces and air fluid levels. Computed tomography scan chest demonstrated a homogenous mass extending into the left main bronchus causing its complete obstruction along with extensive bronchiectatic changes in the left lung. Bronchoscopy and bronchial biopsy confirmed the diagnosis of typical carcinoid tumour. A pneumonectomy was performed. This case is unusual due to the large size of the tumour, its location and associated destruction of the entire left lung.

Keywords: Carcinoid tumour, Lung tumour, Bronchiectasis, Bronchial obstruction

Introduction

Carcinoid tumours arise from the neuroendocrine cells of the bronchial epithelium known as Kulchitsky cells.[1] They comprise 4% of all lung malignancies,[2] with studies indicating an incidence of 1 to 2/100,000 in various populations.[3]

The lung is considered to be the 2nd most common site of occurrence for carcinoid tumours. Within the lungs, the tumours are most commonly found on the right side in the lobar bronchus.[2]

We report a case of a male patient who presented with loss of lung volume on the left side with cystic spaces and air fluid levels. The case was unusual due to the large size of the tumour, its location and associated destruction of the entire left lung.

Case Report

A 16-year-old male presented with a history of fever, cough, left-sided chest pain and shortness of breath. General physical examination was unremarkable. Chest examination revealed decreased chest movements on the left side. Percussion note was dull with increased vocal fremitus. There was no air entry in the left lung. Baseline work-up for tuberculosis was negative. Chest X-ray showed loss of lung volume on the left side with cystic spaces and air fluid levels (Figure-1 and 2). Mediastinum was shifted to the left. Computed tomography (CT) scan demonstrated a fairly defined homogenously enhancing mass approximately 6x5cm seen in the left lung predominantly in the left hilar region. Medially it was extending in to the left main bronchus causing its complete obstruction. Left lung showed extensive cystic bronchiectatic changes (Figure-3). Bronchoscopy revealed a growth in the left main bronchus, completely obstructing the lung, >2cm from the carina. Bronchial biopsy of the tissue showed that the mass was arranged in small clusters and aggregates of cells with pink eosinophilic cytoplasm, uniformity of nuclei and mild hyperchromasia. No mitosis was discernable. Specimen was positive for cytokeratin CAM 5.2, AE/A3, chromogranin A, CD56 and synaptophysin. Features favoured carcinoid tumour of the lung. After pre-operative work-up, left-sided thoracotomy and pneumonectomy was performed. The whole left lung was destroyed secondary to persistent obstruction at the level of the left main stem bronchus due to the tumour. The left lung was resected entirely along with the tumour (Figure-4). During surgery, a nodular fleshy grayish white large lesion was found in the left main bronchus. Surrounding

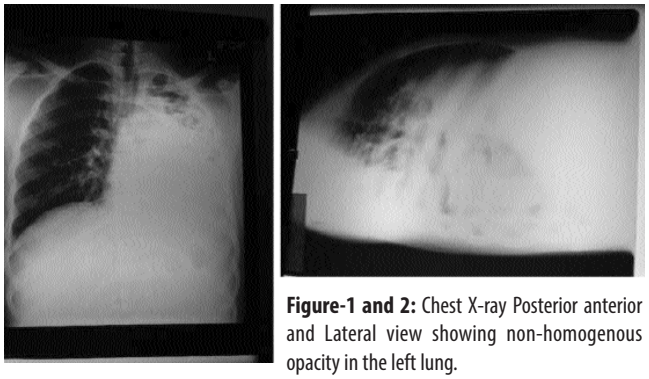

Figure-1 and 2: Chest X-ray Posterior anterior and Lateral view showing non-homogenous opacity in the left lung.

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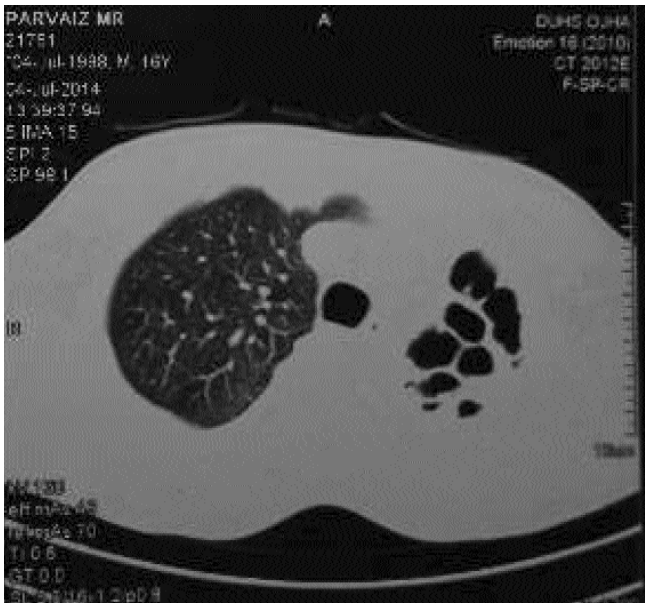

Figure-3: Computed Tomography scan of lung window showing extensive bronchiectatic changes in left lung.

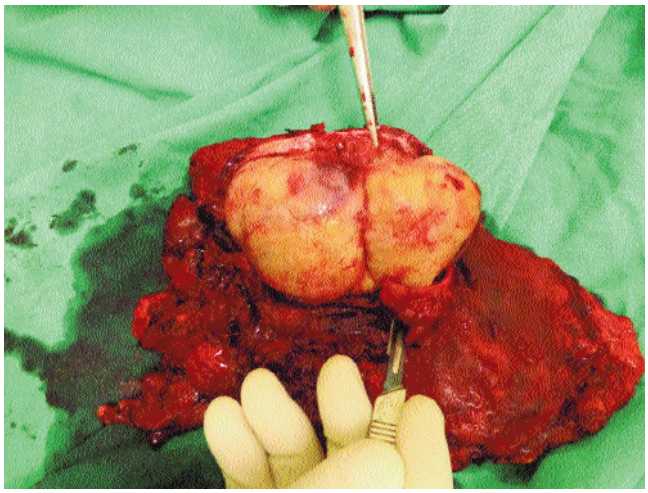

Figure-4: Post-pneumonectomy view. Huge bulky tumour removed from left main bronchus.

lung parenchyma showed dilated air spaces filled with pus and mucoid secretions. Left lung was resected entirely. Histopathology, later on, revealed a neoplastic lesion composed of small monomorphic cells in an organised growth pattern exhibiting focal perivascular rossetting. Individual cells showed moderate amounts of eosinophilic cytoplasm and round to oval nuclei with stripped chromatin. Approximately 1 mitosis/10 high-power field (HPF) was noted. There was no evidence of granuloma formation or malignancy. Final diagnosis of typical carcinoid tumour arising from left main bronchus was made. Patient's recovery was smooth and he is doing well at follow up. He is now status post-10 months after surgery and remains healthy. After surgery, the patient was seen 1 week later with the biopsy report. After that he remained on weekly follow-up for a month, then fortnightly follow-up for another month and since then he is on regular monthly visits. The patient's progress has been monitored via symptom history, physical exam and serial chest X-rays.Up till his last follow-up in February 2015, the patient was symptom free and without any complaints with satisfactory chest X-ray.

Discussion

The lung is considered to be the second most common site of occurrence for carcinoid tumours. Within the lungs, the tumours are most commonly found on the right side in the lobar bronchus.[2] A study on 79 cases of bronchial carcinoids showed that 19 tumours were located in the upper lobe, 10 in the middle and 26 in the lower robe. The right to left side ratio was roughly equal with 40 tumours arising in the right lung and 39 in the left.[2] In 9 out of 79 cases, the tumour was found in the main stem bronchus, a relatively rare location for carcinoids.[2] In our case, the tumour was found in the left main bronchus, protruding into the carina. Pulmonary carcinoid tumours may be central or peripheral in origin with a peak incidence between 4th and 5th decades of life[4] although it may affect almost any age group. Our patient was 16-years-old. While peripheral tumours are more commonly asymptomatic, central tumours may present with chest pain, persistent cough, asthma-like wheezing, dyspnoea, haemoptysis, lymphadenopathy and obstructive pneumonitis. Some of these symptoms may be mistaken for tuberculosis and in a region like ours, makes the diagnosis difficult, requiring a high degree of suspicion.

According to World Health Organisation (WHO), carcinoid tumours have been divided into typical and atypical.[5] Atypical carcinoidis based upon four elements: a morphological base, number of mitoses <2 per 10HPFs, absence of necrosis and dimension ≤0.5 cm; while an atypical carcinoid shows carcinoid morphology with number of mitoses ≥2 and <10 per 10 HPFs and areas of coagulative necrosis.[3] The histopathology report of our patient showed approximately 1 mitosis/HPF which favoured a diagnosis of typical carcinoid. A study showed that the mean size of a bronchial carcinoid was 2.4±1.5 cm. Lesions ranged in size from 0.6 to 6 cm, with 6cm being the largest. The mean dimension of typical carcinoids was 2.1±1.2 cm, while that of atypical carcinoids was 3.6±2.1 cm.[5] In our case, the tumour was unusually large measuring 9x7x6cm. Our literature search failed to find a

tumour this large having been reported before.

CT scan, bronchoscopy, and transbronchial biopsy may aid in diagnosis.[6] Final diagnosis is based on histology and confirmed with immune histochemical staining for neuroendocrine markers. Carcinoid tumours secrete a variety of tumour markers, including 5HIAA, Chromogranin A, cytokeratins, CD56 and Ki67. Immunohistochemistry of tissue specimen of our patient was positive for Chromogranin A, Cytokeratin AE1/AE3, Cytokeratin CAM 5.2, CD-56, Synaptophysin and Ki-67 Mib. Chromogranin A is a secretory protein found in elevated concentrations in neuroendrocrine tumours like carcinoid tumours, pheochromocytoma or medullary thyroid cancer. It can be measured in either the serum or detected by immunohistochemistry in a biopsy specimen. Chromogranin A levels were tested via immunohistochemistry and were positive in the tissue specimen of our patient. We did not check serum levels of Chromogranin A.

Goal of treatment is to remove the tumour completely and preservation of as much unaffected lung tissue as possible.[7] Our patient presented with complete destruction of the left lung and, as a result, underwent complete removal of left lung along with the tumour. Although the 5-year survival for typical bronchial carcinoid tumours is over 80%,[3] a delay in diagnosis is undesirable and may be associated with adverse outcomes. Distant metastases are seen in 1.5% cases.[3] Recurrence is very low, about 3-5%, and is usually due to incomplete resection.[5]

Conclusion

Typical bronchial carcinoid tumour is a rare malignancy of the lung. If timely diagnosed, a sleeve resection may be all that is needed for complete cure. Delay in treatment can result in potential complications from increased tumour size causing obstruction of the bronchus and destructive changes in the lung, as was the case in our patient.

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