Atypical Bronchial Carcinoid

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ABSTRACT

Carcinoid tumour is a rare entity accounting for less than two percent of bronchial neoplasms. They are rare well-differentiated, neuro-endocrine tumors, with low-grade malignancy. They are distinguished in two different groups: typical and atypical carcinoid. Typical and atypical carcinoids differ in their biological behavior and prognosis. Atypical bronchial carcinoid tumor is still rare. We report one such rare case.

Keywords: Carcinoid, atypical, bronchial

INTRODUCTION

Carcinoid tumors develop from stem cells of the bronchial epithelium known as Kulchitsky cells, which have neuroendocrine activity. Bronchial carcinoids account for 1-2% of all lung tumors[1].

At present, surgery is the gold standard for treatment of this tumor, with a different approach between typical carcinoids, in which a parenchyma-sparing resection is preferred, and atypical carcinoids, in which a limited resection should be obviated[2]. The outcomes of bronchial carcinoid tumors are generally favourable after surgery. However local recurrences and metastases to lymph nodes and lung parenchyma can occur more commonly with atypical carcinoids. High index of suspicion with prompt diagnosis and histopathological examination along with long term follow up is necessary for effective management of these tumors[3]. We present a rare case of atypical carcinoid in an elderly male treated surgically.

CASE REPORT

A 65 year old male presented to our institute with complaints of recurrent breathlessness, hemoptysis and fever since six months. He was under treatment for tuberculosis since two months at a district hospital and had no response to this treatment. Thereby, he was referred to our institute for management. He was admitted under Respiratory medicine department and was thorougly investigated.

His complete blood picture was normal. Montoux test was negative. Chest X-ray revealed consolidation of the left lung. Contrast enhanced computed tomography (CECT) of the chest showed large well defined heterogeneously enhancing mass with few hypodense areas of necrosis and chunky calcifications at the left hilum encasing bronchovascular bundle and seen involving lingula and left lower lobe with few pleural based nodules in the left lung (Figure 1) . CT of the chest was reported as malignancy of the lung with metastases. Sputum smears for acid fast bacilli and fungal stain were negative. Bronchoscopy was performed which revealed an endobronchial mass with cheesy necrotic foci originating at the left main bronchus which was bleeding on touch. Bronchoscopic biopsy of the mass was done for histopathological examination.

Grossly, the specimen was linear, grey white to grey red, measuring 0.7x0.4 cm and friable. Microscopically, it showed cells arranged in solid nesting pattern with nuclear powdery chromatin, increased mitotic activity (6/ 10 HPF) and mild atypia. The tumor cell nests were separated by thin fibrovascular septa. There was a small focus of necrosis. Chromogranin and synaptophysin were positive in tumor cells and TTF-1 was negative. The impression given was atypical carcinoid.

Later, the patient was posted for surgery and surgical resection of the mass in toto was done along with mediastinal lymph nodal resection. During this procedure three satellite nodules were also resected. The subsequent histological examination confirmed the diagnosis of atypical carcinoid with lymphnodal metastasis.

DISCUSSION

Carcinoid bronchopulmonary tumors represent approximately 25% of all carcinoid tumors and 1%–2% of all lung neoplasms[4]. Approximately 70% of these tumors are located centrally in the large bronchial tubes leading to the lung collapse, while 10%–20%, are peripheral carcinoids, which occur in the periphery[4]. World Health Organisation categorized neuroendocrine pulmonary tumors into four types: typical carcinoid; atypical carcinoid (ATC); large cell neuroendocrine

carcinoma; and small cell lung cancer[5].

Typical carcinoid pulmonary tumors manifest nine to ten times more often than ATC tumors. Typical carcinoid tumors are seen in younger patients more often than ATC, which occur in the elderly as was in our case.

Patients with bronchial carcinoid typically presents with hemoptysis, dyspnea on exertion and wheeze because of central obstruction of the airways. Diagnosis can often be delayed and patients may present to physician after prolonged courses of antibiotics, inhaled corticosteroids and bronchodilators in view of suspected asthma or recurrent post obstructive pneumonia[6]. A variety of neoplastic syndromes occur with these tumours including carcinoid syndrome, Cushing's syndrome and Acromegaly. Our case has no such syndromes.

Pathologically, carcinoids are classified as (a) classic carcinoids which have a mosaic or trabecular pattern, and (b) variants which include adenopapillary, clear cell, oncocytic, melanenogenic, spindle cell and atypical carcinoid[1]. Those carcinoid tumours that exhibit not only the overall architectural, ultrastructural and immuno-histochemical features of classic carcinoid tumours but also atypical features in the form of increased mitotic activity (>5/10 HPF), nuclear hyperchromasia and foci of necrosis are referred to as atypical carcinoids[1]. Immuno-histochemistry reveals variable positivity for keratin, serotonin, NSE, chromogranin A and B, synaptophysin, Leu-7 and neuro filaments[1]. In our case, chromogranin and synaptophysin were positive.

Bronchoscopic appearance of carcinoid is classic: pink to red vascular mass attached to the wall of the bronchus by a broad base. HRCT of the chest plays an important role in the diagnosis of bronchial carcinoids as it gives good information regarding tumor extent, size, location and mediastinal lymph nodal involvement.

Typical and atypical carcinoids differ in their biological behavior and prognosis. Atypical carcinoids have a higher recurrence rates and present more often with hilar and mediastinal lymph nodal metastases when compared with typical carcinoids[6] . 5 to 20% of typical carcinoids and 30% to 70% of atypical carcinoids metastasize to lymph nodes[6].

Typical carcinoids usually have a good prognosis with a 5 year survival rate of 87% to 89%. [7]Atypical carcinoids are associated with 5 year survival rate of 44% to 78%[7] and distant metastases can occur when compared to typical carcinoids. A prolonged follow up of 10 years is mandatory and recommended [3].

Treatment for both typical and atypical carcinoids localised to the lung consists of excision of the lesion and mediastinal lymph node resection. Bronchoscopic treatment is considered curative in cases of pure intraluminal typical carcinoids with central airway obstruction[8]. It also helps in good visualization of tumor margins if a surgical resection is opted. Atypical carcinoids and tumors with extraluminal involvement and severe distal parenchymal disease require more extensive surgery along with complete tumor removal and mediastinal lymph nodal dissection[9] .

Our patient had a central endobronchial mass and a large mass in the left lung with surrounding nodules along with mediastinal lymph node involvement. So a more extensive surgical resection of the lung parenchyma was done along with mediastinal lymph nodal dissection.

CONCLUSION

To conclude, possibility of atypical carcinoid should be kept in mind while dealing with lung masses presenting with breathlessness, hemoptysis and various syndromes mentioned earlier. Histopathological examination is diagnostic and its classification is important as the treatment and prognosis will differ according to the type of carcinoid.

Figure 1:

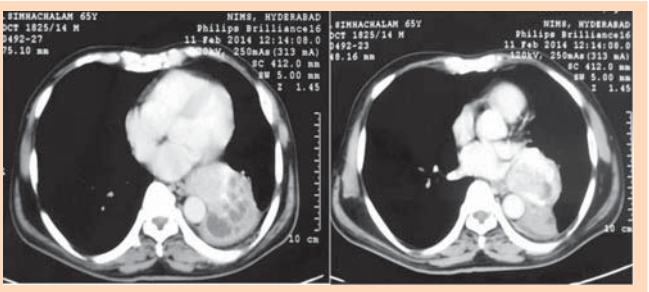

Figure 1: Large well defined heterogeneously enhancing mass with few hypodense areas of necrosis and chunky calcifications seen involving lingula and left lower lobe.

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