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# Typical Carcinoid of Lung – Rare Case with Review of Literature

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***Authors' contributions***

*This work was carried out in collaboration between both authors. Author AD performed surgery, collected clinical data, searched literature and reviewed the manuscript. Author DK completed histopathology processing, searched literature and prepared manuscript. Both authors read and approved the final manuscript.*

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***Case Report***

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## ABSTRACT

Neuroendocrine tumors represent 25% of primary lung neoplasms, of which only 2% are typical carcinoid. 75% of lung carcinoids originate from Kulchitsky cells in bronchus. Typical carcinoid tumors are well differentiated with low grade malignancy. Strong and diffuse positivity for neuroendocrine markers CD 56, synaptophysin and chromogranin is exhibited by these tumor cells. Carcinoid tumors in the past were excluded from TNM staging but 7$^{th}$ edition of TNM staging system includes carcinoid. The patients are usually younger than for usual lung cancers. Carcinoid syndrome is usually absent when the tumor is confined to lung. Markedly different prognosis and treatment underlies the importance of accurate pathologic diagnosis of neuroendocrine tumors of lung. Typical carcinoid has a five year survival up to 95% when there is complete surgical excision.

*Keywords: Neuroendocrine tumor; typical carcinoid; lung.*

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## 1. INTRODUCTION

Neuroendocrine tumors of respiratory tract develop from stem cells of bronchial epithelium, Kulchitsky cells, which have neuroendocrine activity. These arise most of the times within bronchi (central) in about 70 % patients and sometimes in lung parenchyma (peripheral) in 10-20%. The 2004 World Health Organization (WHO) classification recognizes four major types of lung endocrine tumors ; Typical Carcinoid, Atypical carcinoid, Large cell neuroendocrine carcinoma and Small cell lung carcinoma. They are further grouped as Low grade (Typical carcinoid), Intermediate grade (Atypical carcinoid) and High grade (Large cell neuroendocrine tumor and Small cell lung carcinoma). Lung is a second common site for carcinoid tumors after gastrointestinal tract and accounts for 30% of neuroendocrine tumors in the body. Low grade Carcinoid tumors of lung are common in 40-50 years age group and are not associated with history of smoking. These tumors are strongly and diffusely positive for neuroendocrine markers CD 56, synaptophysin and chromogranin. Less than 2% are associated with carcinoid syndromes, but are found in 5% patients of Multiple Neuroendocrine Neoplasia1 (MEN 1) [1].

At present, surgery is the gold standard for treatment of this tumor. Parenchyma sparing resection is usually preferred in typical carcinoids with favorable outcome. [2] However, rare local recurrences and nodal metastasis are reported. With high index of suspicion and prompt diagnosis by histopathological examination and long term follow up is necessary for effective management of this tumor [3].

## 2. CASE REPORT

A sixty year old male clinically presented with complaints of recurrent cough, intermittent chest pain and breathlessness for last three months. His High Resolution Computerised Tomography (HRCT) showed a single well defined 22x27 mm heterogeneously enhancing nodule in left paracardiac region, suggesting it to be metastatic deposit. This was followed by PET scan which revealed a single nodule in lingular lobe of (Lt) lung. Surgical excision of the mass was planned. His laboratory investigations like complete blood count, urine analysis, liver and kidney function tests, etc… were within normal limits. ECG and 2D Echocardiography were also normal.

Under general anesthesia, (Lt) lateral muscle sparing thoracotomy was performed through 5$^{th}$ intercostal space. (Lt) upper lobe arteries were dissected and doubly ligated. (Lt) pulmonary vein was dissected and ligated. Bronchial stumps were closed with staplers. There were no pleural adhesions. There was a large well circumscribed mass in the lingular lobe of (Lt) lung. No other mass lesion or lymphadenopathy was seen. (Lt) upper lobectomy was performed.

## 3. MORPHOLOGY

Surgically resected specimen of (Lt) lung upper lobe was received in histopathology section. It was grayish 8 x5x 3 cm, covered by glistening white pleura. Externally no mass or cavity was seen. Hilum showed intact bronchus. Cut surface had a single rounded whitish 3x2.5 cm firm mass (Fig.1). Rest of the lung tissue showed diffuse areas of hemorrhage.

**Fig. 1. (Lt) lung apical lobectomy specimen showing well circumscribed single nodule within lung parenchyma**

Multiple sections from various areas showed tumor tissue arranged in nests and trabecular pattern separated by delicate fibrous septae consisting of monotonous appearing cells with rounded nuclei and granular cytoplasm (Fig 2a Low power view and 2b High power view) and highly vascularized stroma with bronchial epithelium and anthracotic pigment in Fig 2a. No necrosis or hemorrhage was seen. The mitotic figures were 0-1/ten hpf. The tumor cells have not extended into capsule or surrounding lung parenchyma which consisted of alveoli, cartilage, bronchial tissue and blood vessels. A diagnosis of Typical Carcinoid (Low grade) was conveyed to the surgeon with a request for oncopathologist's opinion. IHC was carried on paraffin blocks for Neuron specific enolase, Synaptophysin (Fig 3a) and Chromogranin (Fig 3b) which were positive while Pancytokeratin showed focal weak positivity, Ki-67 was positive (< 1%) and Vimentin was negative and thus confirming our diagnosis of Low Grade Typical Carcinoid.

## 4. DISCUSSION

Carcinoid tumor is a rare entity accounting for less than 2% of pulmonary carcinoids [4]. Typical pulmonary carcinoids manifest nine to ten times more often than atypical carcinoid and seen in younger patients. A variety of paraneoplastic syndromes are known to occur with this tumor. Our patient didn't have any such syndromes. HRCT of chest plays an important role in the diagnosis as it gives good information regarding tumor extent, size, location and mediastinal lymphnode involvement. Most of the carcinoid tumors arise in the main segmental bronchi (central), also known to arise in peripheral part where these are solid, nodular, well defined with smooth lobulated ivory to pink glistening cut surface with rare necrotic foci. The lung distal to tumor may show atelectasis, obstructive pneumonia or bronchiectasis [5]. Our patient had atelectasis. Pathologically carcinoids are classified as (a) classic carcinoids which have mosaic or trabecular pattern and (b) variants

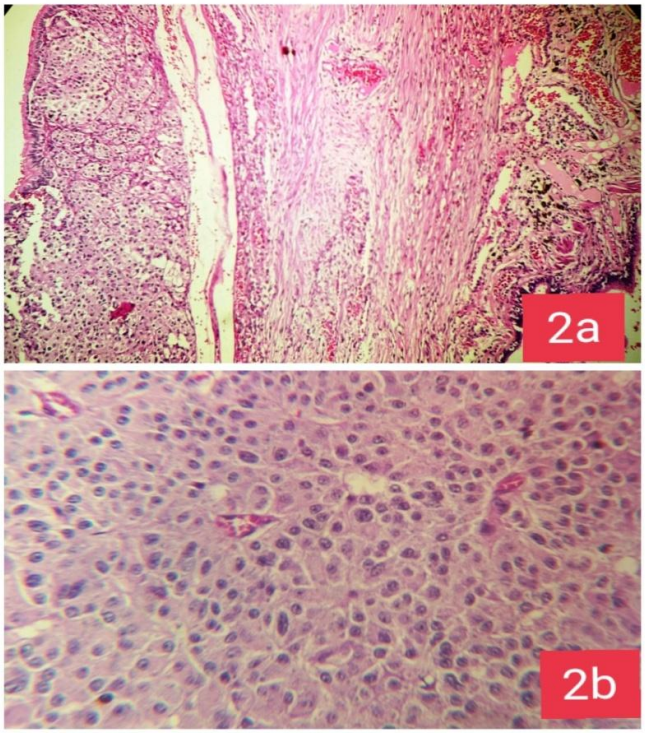


**Fig. 2.(a). Section from mass with lung parenchyma showing pseudostratified columnar ciliated bronchial epithelium and beneath tumor tissue arranged in nests. Surrounding area shows lung alveoli with anthracotic pigment and bronchiole. (10x X 10x H&E); (b)Section from mass showing tumor tissue arranged in nests separated by fibrous septae with round to oval monotonous cells containing rounded nuclei and granular cytoplasm in vascularized stroma. (40x X 10x H & E)**

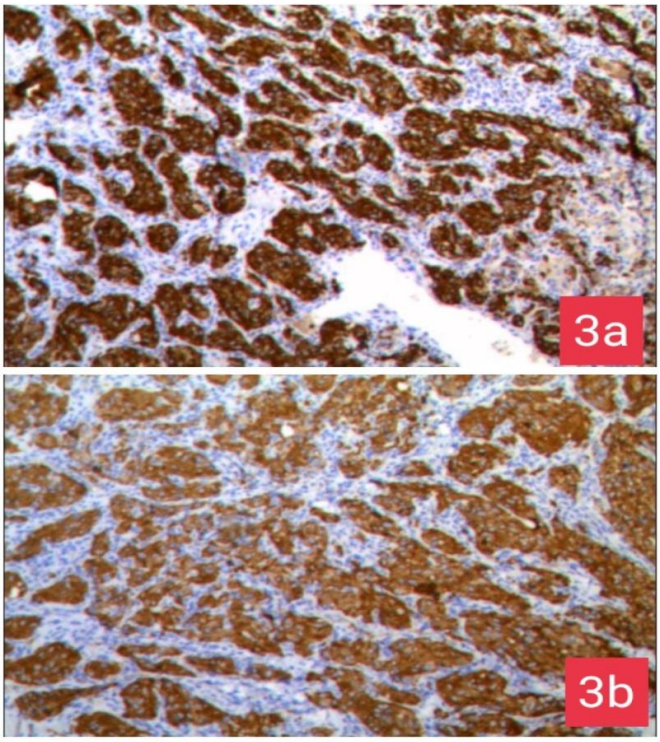


**Fig. 3. Immunohistochemistry showing positivity to Synaptophysin (3a) and Chromogranin (3b)**

which include adenopapilary, clear cell, oncocytic, melanogenic, spindle cell [6]. Histologically tumor is made up of nests, trabeculae or mosaic pattern of medium sized polygonal cells with oval to spherical nuclei and eosinophilic granular cytoplasm. Typical carcinoids are low grade, well differentiated tumors with less than two mitoses per 10 high power fields (< 2 mitoses/10 hpf) and without foci of necrosis. Immunohistochemistry reveals positivity for CD 56, synaptophysin, chromogranin, neuron specific enolase and focal positivity for, pancytokeratin and Ki 67 and immunonegative for vimentin. In our patient also same immunoreactivity was observed and that confirmed the diagnosis.

The treatment for both typical and atypical carcinoid consists of excision of the lesion and mediastinal lymphnode resection [7]. With complete removal of tumor, typical carcinoid has excellent prognosis with 85-95% five year survival [8]. Typical carcinoid has 3 – 5% recurrence [9]. It has also been observed that typical carcinoid are capable of regional lymph node metastasis, so a prolonged 10 year follow up is recommended [5]. Carcinoid syndrome is usually absent when the tumor is confined to lung, but can develop years later with metastasis [5]. Patients presenting with carcinoid syndrome have a shorter survival [10]. Little can be offered to patients with metastasis [6].

The tumor responded very well to complete surgical excision in our patient and he is on regular follow up since four years.

## 5. CONCLUSION

Carcinoid tumors are a relatively uncommon subset of pulmonary tumors which most often present with nonspecific pulmonary symptoms like cough, hemoptysis, recurrent pneumonia, etc. and very rarely with carcinoid syndrome. Lung sparing resections as a primary treatment is adequate and complete resection yields long term survival. The role of adjuvant chemotherapy or radiotherapy has not been well documented.

## CONSENT

As per international standard or university standard, patient's consent has been collected and preserved by the authors.

## ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

## ACKNOWLEDGEMENT

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## COMPETING INTERESTS

Authors have declared that no competing interests exist.

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