

# Atypical Presentation of Typical Carcinoid of Lung with Therapeutic Cryoextraction –A Case Report

Isha Garg<sup>1</sup>, Srikant Agarwal<sup>2</sup>, Jitender Kumar Saini<sup>3</sup>, Lokender Kumar<sup>4</sup>

## ABSTRACT

**Introduction:** Neuroendocrine tumors (NETs) of the lung account for 5% of all cases of lung cancer, which itself is the leading cause of cancer-related death worldwide. In view of its rarity and atypical presentation, delay in diagnosis and treatment are common.

**Case report:** The patient reported in this case is a young 19-years old male, who never smoked and exhibit atypical presentation. Chest CT-scan showed intraluminal non enhancing soft tissue density in left main bronchus extending to left upper lobe bronchus. The detected soft tissue endobronchial mass was cryobiopsied and bronchoscopically resected via electrocautery and cryoextraction in multiple sittings. Histopathological diagnosis identified well-differentiated typical carcinoid (TC), which represents one of the four subgroups of pulmonary NETs. Additionally, it expressed neuroendocrine marker synaptophysin and chromogranin, that further verified their neuroendocrine origin. In line with the importance of cancer treatment, early identification of neuroendocrine neoplasms is highly warranted.

**Conclusion:** The unusual presentation of the young patient described here mimicked Bronchial asthma or foreign body but found to have carcinoid which is being extracted and successfully treated by therapeutic bronchoscopy without need of any radical surgery.

**Keywords:** Neuroendocrine, Carcinoid, Immunohistochemistry, Cryoextraction.

## INTRODUCTION

Carcinoid tumors are neuroendocrine tumors derived from enterochromaffin or Kulchitsky cells, which are widely distributed in the body.<sup>1</sup> Carcinoid tumors may develop in many locations in the body, but most often they are found in small intestine (26%), respiratory system (25%) and appendix (19%).<sup>2</sup> They are characterized histologically by positive reaction to silver stains and markers of neuroendocrine tissue, including neuron specific enolase (NSE), synaptophysin and chromogranin.<sup>3</sup> Carcinoid tumors termed (incorrectly) as bronchial adenomas in the past are uncommon pulmonary neoplasms making 1-2% of all lung tumors.<sup>4</sup> They often arise in persons who are younger than is usual for lung cancers and male to female ratio is 1:1.

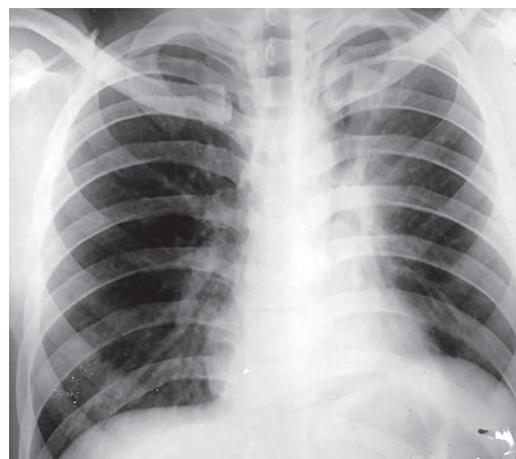
## CASE REPORT

A young 19 years old student who never smoked presented with breathlessness on minimal exertion since 4 months, intermittent left side chest pain since 3 months. General examination was within normal limits. Systemic examination

revealed decreased air entry at base of left lung. Chest X Ray done subsequently showed left upper lobe collapse [Fig 1]. High-resolution computed tomography (CT) thorax showed intraluminal non enhancing soft tissue structure (17.1x5.8 mm) in left main bronchus extending into left upper lobe bronchus [Fig 2].

Bronchoscopy showed smooth well circumscribed polypoidal growth in upper division of left upper lobe bronchus extending into left main bronchus. Cryobiopsy was taken. Histopathology revealed bronchial wall tissue infiltrated by tumor cells arranged in nests exhibiting mild anisonucleosis and coarse chromatin with moderate amount of eosinophilic cytoplasm. <1/10 high power field mitotic figures was evident. Diagnosis of typical carcinoid was given which was confirmed by positivity for chromogranin, synaptophysin, CD56 and Ki-67 proliferation index of <1% on immunohistochemistry (IHC). It was negative for cytokeratin-7 (CK-7), thyroid transcription factor-1, and CK-5/6.

Electrocautery (power setting of 35volts for 5 seconds



**Figure-1:** Showing left upper lobe collapse

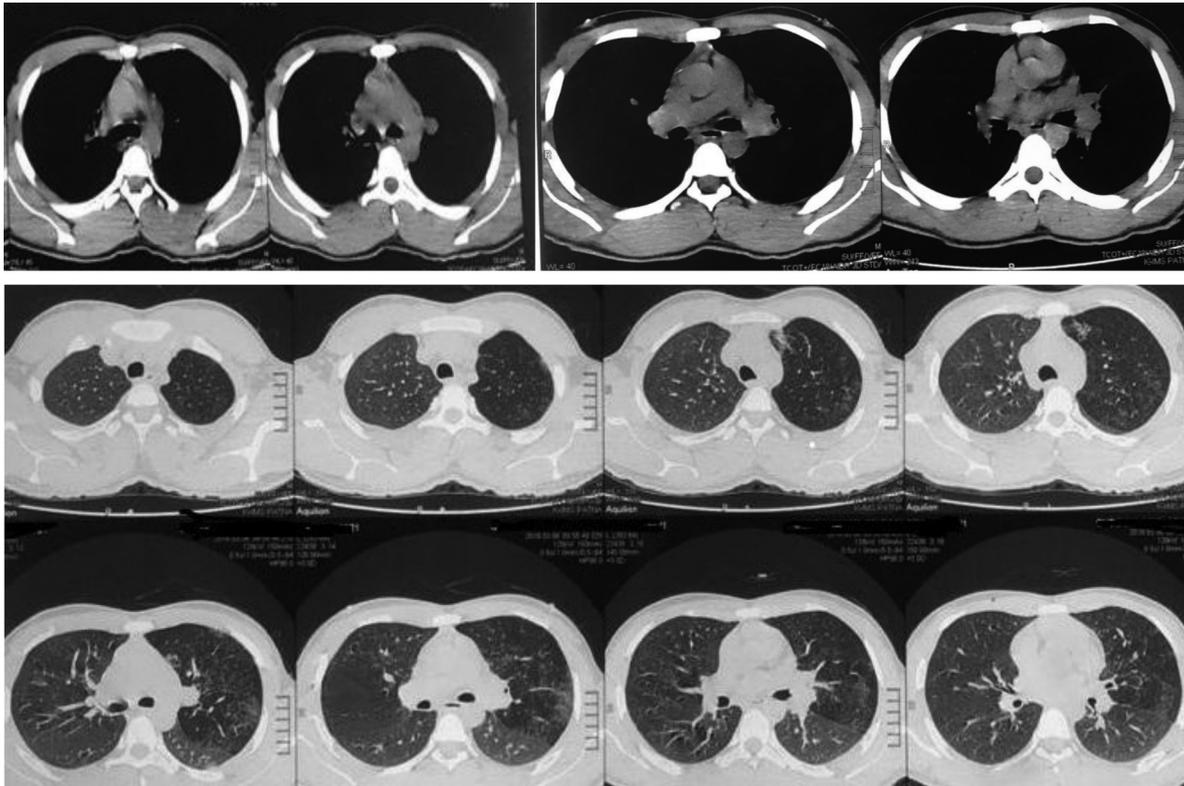
<sup>1</sup>Senior Resident, Department of Respiratory Medicine, NITRD, New Delhi, <sup>2</sup>Senior Resident, Department of Respiratory Medicine, NITRD, New Delhi, <sup>3</sup>Senior Chest Specialist, Department of Respiratory Medicine, NITRD, New Delhi, <sup>4</sup>Senior Chest Specialist, Department of Respiratory Medicine, NITRD, New Delhi, India

**Corresponding author:** Dr Isha Garg, Department of Respiratory Medicine, NITRD, New Delhi, India

**How to cite this article:** Isha Garg, Srikant Agarwal, Jitender Kumar Saini, Lokender Kumar. Atypical presentation of typical carcinoid of lung with therapeutic cryoextraction –a case report. International Journal of Contemporary Medical Research 2020;7(4):D1-D3.

**DOI:** <http://dx.doi.org/10.21276/ijcmr.2020.7.4.5>





**Figure-2:** Showing intraluminal non enhancing soft tissue structure in left main bronchus extending into left upper lobe bronchus



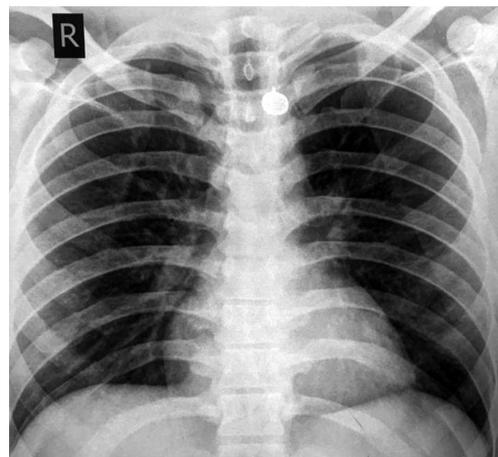
**Figure-3:** Showing bronchoscopic view of polypoidal growth in left upper lobe bronchus undergoing cryoextraction



**Figure-4:** Showing complete clearance of left upper lobe bronchus after cryoextraction followed by electrocautery

done twice,blend mode,normal coagulation, electrosurgical monopolar unit, with foot switch, flexible monopolar electrocautery blunt probe Olympus CD-6C-1, loop snare, [Olympus, Tokyo, Japan] was performed on the polypoidal tumor tissue followed by cryoextraction.[Fig 3]

This intervention fully re-opened the obstructed left main



**Figure-5:** Showing expansion of left lower lobe collapse after debulking

bronchus and the upper division of the left upper lobe that was confirmed during follow up bronchoscopy as a area of raw mucosa [Fig 4]. Follow up was done with regular monitoring and imaging studies (i.e, chest x-ray, chest computed tomography). Subsequent bronchoscopies did not show any recurrence during follow up of 1 year and the patient was asymptomatic with normal chest radiographs as shown below [Fig 5].

## DISCUSSION

In neuroendocrine tumors, three grades based on histologic features and biologic behavior are currently recognized-Grade I or typical carcinoid, Grade II or atypical carcinoid and Grade III or small cell carcinoma/ large cell carcinoma.<sup>5</sup> Typical carcinoids occur in both sexes with equal frequency

and the age at onset ranges from childhood to 9th decade showing no association with smoking. However, atypical carcinoids tends to occur in older patients with smoking as a risk factor. Many patients with typical carcinoid are asymptomatic, but dyspnea, cough and hemoptysis may occur particularly in central lesions.<sup>5,6</sup> The differentials of a patient with symptoms of bronchial obstruction, bronchospasm, and hemoptysis includes an obstructing bronchial carcinoma, endobronchial metastasis, hamartomas, aspirated foreign body, asthma, and chronic obstructive pulmonary disease. Variability in clinical presentation may lead to delay in diagnosis or even misdiagnosis. Our patient was a 19-years-old male, non-smoker presented with breathlessness and intermittent chest pain simulating foreign body aspiration. The bronchial mucosa overlying carcinoid tumors is frequently intact or may show squamous metaplasia. Thus, cytological examination of sputum is frequently negative and only tissue biopsy would be highly successful in harvesting large number of malignant cells.<sup>5</sup> Mostly, these tumors arise in main to the segmental bronchus, but tumors of peripheral origin are occasionally seen.<sup>8</sup> Grossly, the tumors are polypoid, tan to yellow, 0.5-8 cm in diameter and covered with intact bronchial mucosa.<sup>8,6</sup> Histologically typical carcinoid exhibit an organoid pattern and the nuclear chromatin of tumor cells shows "salt and pepper" appearance. According to recent WHO classification, atypical carcinoid differs from typical carcinoid by the presence of punctuate coagulative necrosis and or 2 to 10 mitosis/10 high-power fields mitotic indices. Carcinoid tumors whether typical or atypical, stains positively for neuroendocrine markers ie. chromogranin, synaptophysin, and NSE.<sup>5</sup> In the present case, the polypoidal tumor was located in upper division of left upper lobe bronchus. Histologically features of carcinoid tumor with occasional mitotic figure were seen without areas of necrosis. Furthermore, IHC was positive for chromogranin and synaptophysin. Hence, the diagnosis of typical carcinoid was confirmed. Treatment of typical carcinoid is surgical and usually involves lobectomy or pneumonectomy with lymphadenectomy.<sup>6,7</sup> Metastases occurs usually to regional lymph nodes however distant metastases to bone and liver may be associated with carcinoid syndrome.<sup>5</sup> At the time of diagnosis 10-15% of typical carcinoid and 40-50% of atypical carcinoid present with lymph node metastasis. Typical carcinoids have an excellent prognosis, and overall 5 and 10-year survival rate are 90-98% and 82-95% in typical carcinoid and only 61-72% and 35-39% in atypical carcinoid. With metastatic disease chemotherapy can be given with cisplatin based or streptozocin based regimen with moderate effectiveness.<sup>6</sup> In the present case, the typical carcinoid tumor was successfully resected with cryoextraction and electrocautery and patient was followed 3 months later with marked improvement.

## CONCLUSION

Diagnosis of carcinoid tumors is often challenging demanding separation from other neuroendocrine tumors and a wide variety of other tumors like sclerosing hemangioma,

paraganglioma, glomus tumor and adenocarcinoma. IHC is the most remarkable investigation in making final diagnosis. Distinction of typical carcinoid from atypical carcinoid is essential since staging is an important prognostic factor. However, even with lymph node metastasis typical carcinoid carries an excellent prognosis. In the present case, the diagnosis of typical carcinoid was made. The case is presented for its rarity and unusual course of events and patient was successfully cured in the form of debulking of the bronchial mass. However the diagnosis of Carcinoid should be kept in mind whenever looking at a mass especially in young patients where suspicion of malignancy appears low to offer various newer bronchoscopic methods of treatment available. Our case encourage further case reports where endobronchial growth be successfully resected via bronchoscopic intervention in a minimal invasive way thereby avoiding radical surgery.

## REFERENCES

1. Zuetenhorst JM, Taal BG. Metastatic carcinoid tumors: A clinical review. *Oncologist* 2005;10:123-31.
2. Emeryk J, Czekajaska-Chehab E, Korobowicz E, Korbel M, Wegrzyn-Szkutnik I, Milanowski J. Bronchial carcinoid in a 39-year-old man treated for bronchial asthma: A case report. *Cases J* 2008;2:7414.
3. Kulke MH, Mayer RJ. Carcinoid tumors. *N Engl J Med* 1999;340:858-68.
4. Hage R, de la Rivière AB, Seldenrijk CA, van den Bosch JM. Update in pulmonary carcinoid tumors: A review article. *Ann Surg Oncol* 2003;10:697-704.
5. Sadana MJ, Mones JM, Garcia-Molinear M, Frable WJ. Localized disorders of the bronchi & lung. In: Silverberg SG, editor. *Principles & Practice of Surgical Pathology & Cytopathology*. 3rd ed. New York, NY: Churchill Livingstone Inc.; 1997. p. 1189-297.
6. Brambilla E, Lantuejoul S. Neuroendocrine neoplasms. In: Zander DS, Farver CF, editors. *Pulmonary Pathology*. 1st ed. Philadelphia, PA: Churchill Livingstone, Elsevier; 2008. p.563-77.
7. Lips CJ, Lentjes EG, Höppener JW. The spectrum of carcinoid tumours and carcinoid syndromes. *Ann Clin Biochem* 2003;40:612-27.
8. Shimoso Y, Noguchi M. Pulmonary neoplasms. In: Mills SE, editor. *Sternberg's Diagnostic Surgical Pathology*. 4th ed. India: Jaypee Brothers Medical Publishers Ltd.; 2004. p. 1174-216.

**Source of Support:** Nil; **Conflict of Interest:** None

**Submitted:** 17-02-2020; **Accepted:** 16-03-2020; **Published:** 14-04-2020