International Journal of Research in Medical Sciences
Khan NA et al. Int J Res Med Sci. 2020 Dec;8(12):4493-4496
www.msjonline.org
pISSN 2320-6071 | eISSN 2320-6012

DOI: https://dx.doi.org/10.18203/2320-6012.ijrms20205330

Case Report

An unusual 7 cm large typical carcinoid managed by surgery: a rare case report

Nafees Ahmad Khan[1], Huma Firdaus[1]*, Ajay Lall[2], Jaya Kumar[3]

[1]Department of TB and Respiratory Diseases, Jawaharlal Nehru Medical College AMU, Aligarh, Uttar Pradesh, India
[2]Max Super speciality Hospital, Saket, New Delhi, India
[3]Department of Respiratory Medicine, Max Super speciality Hospital, Saket, New Delhi, India

Received: 28 September 2020
Revised: 05 November 2020
Accepted: 06 November 2020

***Correspondence:**
Dr. Huma Firdaus,
E-mail: Huma2170@gmail.com

ABSTRACT

Carcinoid tumors of the lung are uncommon group of pulmonary neoplasms. Most common site is gastrointestinal tract followed by lungs. Typical pulmonary carcinoids are usually small as described in various case series size of a typical carcinoid may ranges from 0.5-2 cm and are managed surgically. Here we present a case of unusually large typical carcinoid measuring up to 7 cm which was managed surgically.

Keywords: Typical carcinoid tumor, Uncommon large size, Surgery

INTRODUCTION

Lung carcinoids are a group of neuroendocrine tumor which can be typical carcinoid or atypical carcinoid. Typical carcinoids are much more common than atypical carcinoids.[1] And rarely spread beyond lungs as opposed to atypical carcinoids. Risk factors for carcinoid include age <60 years, female sex, Caucasians, family history of carcinoid and Multiple endocrine neoplasia type 1 (MEN-1) gene mutations. However typical carcinoids are not associated with smoking. Central typical carcinoids usually present with features of cough, hemoptysis, wheezing and pneumonia.[2,3] Peripheral carcinoids may be asymptomatic. Typical carcinoids range from 0.5-2 cm and surgical approach is the primary therapy for localized and locoregional resectable disease.[4] In the present case an usually large typical carcinoid of 7 cm was managed surgically.

CASE REPORT

46 years old female patient presented with complains of non-specific chest pain on the right side since 4 weeks which was mainly present in infra-scapular region without any association with respiration. She had 3-4 episodes of hemoptysis in last 10 days which was fresh, bright red in color around 50-60 ml. No history of fever, cough, no past history of hemoptysis or pulmonary tuberculosis. There was no associated co morbidities like hypertension, diabetes or any other chronic illness or previous history of hospitalization. Patient was admitted for further evaluation of hemoptysis and chest pain.

Chest x-ray was done which showed a well-defined homogenous opacity in right lower zone with silhouetting of right cardiac border (figure 1). A Computed tomography (CT) scan of thorax was done which showed a large spherical heterogeneous strongly enhancing mass lesion in right lower para-cardiac region and fine soft tissue infiltrates seen in right postero-basal region (figure 2).

Bronchoscopy was done which showed big blood clot coming out from the right middle and lower lobe and bronchus intermedius which was removed by biopsy forceps and suction after removing clots there was a endobronchial growth seen in right middle lobe ½ cm

distal to right middle lobe (RML) orifice. Endobronchial biopsy (2 small) sample was sent for histopathology which was suggestive of malignancy, but tissue typing could not be done. Further Positron emission tomography CT (PET-CT) scan was done which showed large fluorodeoxyglucose (FDG) avid well defined, heterogeneously enhancing soft tissue mass lesion measuring 6.7×6.1×5.6 cm in right middle and lower lobe, no other significant FDG avid lesion seen. As there was no tissue diagnosis but radiologically it was suspected to be malignancy and endobronchial biopsy also proved that it's a malignant lesion. After discussion with onco-surgery team it was planned to do lobectomy as it was a solitary mass with no distant metastasis or mediastinal lymph node. So, right middle lobe and lower lobe lobectomy was done and around 7×7×7 cm grayish tan solid homogenous mass was removed along circumferential tumor free lung margin (figure 3A) and with interlobar, hilar and subcarinal nodal tissue.

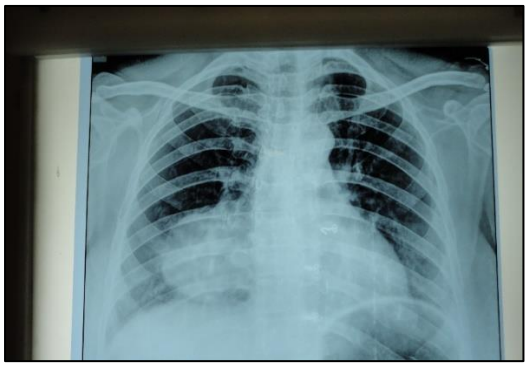

Figure 1: Chest x-ray well defined homogenous opacity in right lower zone with silhouetting of right cardiac border.

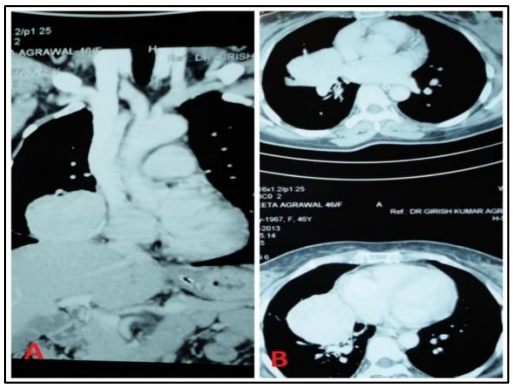

Figure 2: CT scan of the patient: 2A: coronal section 2B: sagittal section, showing a well defined enhancing lesion in right para-cardiac area.

On microscopic examination there were circumscribed growth composed of monomorphic round cells with small uniform centrally placed nuclei with coarse granular chromatin, small nucleoli, mitosis <1/10 high power field, and no evidence of necrosis was seen microscopically (figure 4 A and B), hilar, subcarinal, and lobar lymph nodes were free of metastasis. On immunohistochemistry it was positive for- cytokeratin, chromogranin A, CD56 (figure 4C), neuron specific enolase (NSE) (figure 4D) and negative for synaptophysin. Based on these finding it was labeled as typical carcinoid tumor of right lung with stage-pt2bn0.

Post-surgery course showed excellent recovery of the patient and chest x-ray was done post operatively which showed significant improvement (figure 3B).

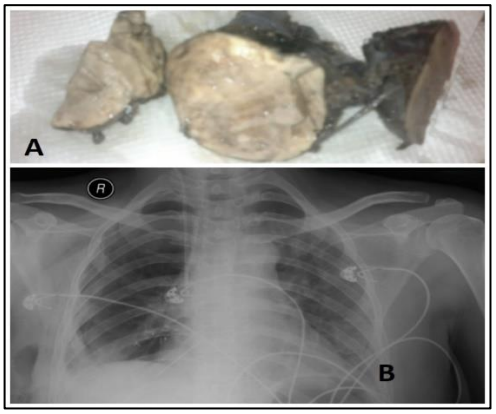

Figure 3: 3A: Grayish tan solid homogenous mass removed; 3B: post-operative chest x-ray showing significant improvement.

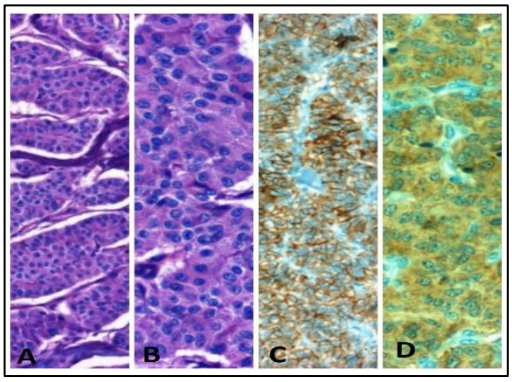

Figure 4: A: intermediate power H and E staining, 4B: high power H and E staining showing round cells with small uniform centrally placed nuclei and no evidence of necrosis; 4C: CD56 staining; 4D: NSE staining.

DISCUSSION

Carcinoid lung tumors are an uncommon group of lung tumors, with an incidence of approximately 0.5-5% of all diagnosed lung cancers develop from neuro-endocrine cells (Kulchitsky cells that are normally present in the bronchial mucosa) and are malignant tumors with the potential to metastasize.[2,5,6,7] Most lung carcinoids are small. They vary from 0.5-2 cm at the time of diagnosis. Patients with carcinoids larger than 3 cm, atypical carcinoids, or carcinoids that have spread to lymph nodes

have a worse prognosis. Most carcinoid tumors originate in the gastrointestinal tract (58%), but lung carcinoid tumors represent about (27%) of all carcinoid tumors.[5]

The 2004 World Health Organization (WHO) classification recognizes 4 major types of lung NETs. They are as follows: atypical carcinoid (AC), (0.2%)- intermediate grade tumor; typical carcinoid (TC), (2%)- low grade tumor; large cell neuroendocrine carcinoma (LCNEC), (3%)- high grade tumor; and small cell lung cancer (SCLC), (20%)- high grade tumor.

Typical carcinoid lung tumors are about nine times more common than atypical carcinoid and characteristically grow slowly and only rarely metastasize beyond the lungs. Carcinoids are commonly found within the cartilaginous portion of the tracheobronchial tree, are usually soft masses covered with intact bronchial epithelium, and very vascular.

Photomicrograph of typical carcinoid shows small nests of uniform cells and atypical carcinoid shows nuclear pleomorphism and mitosis. It has been shown that the number of mitoses is a predictor for prognosis. Typical carcinoid usually have ≤2 mitoses/ 2 mm^2 of viable tumor and atypical carcinoid have 2 to 10 mitoses, necrosis, or architectural disruption.[6]

Clinical presentation

Bronchopulmonary carcinoids have a wide age distribution (4 to 86 years) with an equal gender distribution. In younger patients, more than 90% of carcinoid are typical carcinoid. Atypical carcinoids have been reported to have an association with tobacco use however typical carcinoids have no such association with tobacco smoking or other carcinogenic exposure.[2] The chance of atypical carcinoids is about 25% in patients aged older than 50 years and less than 10% in patients younger than 30.[3] Most common presenting feature is cough not responding to treatment followed by hemoptysis. Patients can also present with non-specific symptoms like chest pain, wheezing, dyspnea, fever due to repeated bronchopulmonary infections.[2,3] Some carcinoid tumours release hormones including serotonin and bradykinin, gastrin, somatostatin, corticotrophin and neuron-specific enolase into the peripheral circulation that cause particular symptoms called as carcinoid syndrome, includes bronchospasm, diarrhoea, skin flushing, and low blood pressure etc. Consistently, about 85% of central tumors are TC, compared with 50% to 80% among peripheral tumors.[2,3]

Management

No medical therapy exists for the primary treatment of carcinoid tumor of the lung. Chemotherapeutic agents and radiation therapy have been used in the treatment of metastatic disease but have met with virtually no success. A response rate of 30-35% has been reported using a combination of 5-fluorouracil and streptozotocin. For symptomatic relief of carcinoid syndrome from metastatic disease administration of radiolabeled octreotide (example- Lutetium (177Lu) DOTA- octreotate) or the radiopharmaceutical 131I-mIBG (meta iodo benzyl guanidine) for arresting the growth of the tumors and prolonging survival in patients with liver metastases have been tried, though these are currently experimental.[8]

The most effective treatment of carcinoid lung tumor is surgical resection of the tumor.[4] Most tumors follow a benign course and are amenable to surgery. Surgical options range from radical resection (the tumor with a good margin of normal tissue is removed) to minimally invasive surgery.

Sleeve resection

Section of the airway containing the tumor is removed.

Segmental resection

Segment of the lung containing the tumor is removed.

Wedge resection

Small wedge of the lung containing the tumor is removed.

Lobectomty

Involved lobe is removed.

Pneumonectomy

Entire lung involving the tumor is removed.

Endoscopic tumor ablation using laser

Nd:YAG laser photoresection of intrabronchial carcinoid tumors also has been proposed for a selected group of patients with low grade (typical), polypoidand central lung NETs. Complete tumor removal is extremely unlikely using these methods because these obstructing intrabronchial tumors usually have penetrated the bronchus and invaded the local pulmonary parenchyma by the time they are discovered, so laser resection is reserved for patients who are poor surgical candidates with central occlusion for palliation.[10,11]

CONCLUSION

This was a rare case of large typical carcinoid measuring 7 cm and was free of any metastasis despite large size so was managed surgically. Lung sparing resection as a primary therapy seems to be adequate for most carcinoid tumors, with complete resection yielding excellent local control and long-term survival. For atypical carcinoids with concerning pathologic features or disease with metastasis adjuvant radiotherapy or chemoradiation must be studied

well in larger and randomized studies in the future to determine the most optimal management of patients with pulmonary carcinoid tumor.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Khan NA, Firdaus H, Ajay Lall A, Kumar. An unusual 7 cm large typical carcinoid managed by surgery: a rare case report. J Int J Res Med Sci 2020;8:4493-6.