University Journal of Medicine and Medical Specialities

ISSN 2455- 2852 2019, Vol. 5(1)

Bronchial carcinoid- An unusual cause of hemoptysis in a young woman.

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Abstract : Bronchial neuroendocrine tumors(NET) are slow growing rare pulmonary tumors arising from neuroendocrine cells. Bronchial NETs account for 1 to 2 percent of all lung malignancies in adults. These tumors are usually sporadic, although 10 percent have features suggesting hereditary origin either multiplicity or association with MEN I. Well differentiated NETs are also known as carcinoid tumors which includes low and intermediate grade groups. Poorly differentiated NETs are high grade tumors which include small cell carcinoma and large cell neuroendocrine carcinoma. Here we present a 32 year old female patient with no comorbid illness who came with complaints of hemoptysis. Subsequent evaluation revealed intrabronchial carcinoid as the cause of hemoptysis. Thus in young patients with no known risk factors, presenting with hemoptysis, bronchial carcinoid should be considered in the differential diagnosis.
Keyword : Hemoptysis, Bronchiectasis, Carcinoid

FIG 1: Chest Xray

Carcinoid tumors are rare pulmonary neoplasms arising from Kulchitsky cells accounting for 1-2% of all primary lung neoplasms. They are divided into typical(80%) and atypical (20%) types. In most of the cases carcinoids arise from the airways, within major to subsegmantal bronchi. Rarely it can have tracheal origin. In 15% of the cases it arises in the lung periphery3. Most endobronchial carcinoids have a large extraluminal component and for this reason they are termed as “ice berg” lesions. This case is presented in order to highlight the need to consider bronchial carcinoids as a differential diagnosis in young patients, with no known risk factors, presenting with hemoptysis . Also, in patients who have localised bronchiectasis, diagnostic bronchoscopy is essential in order to rule out intrabronchial lesions. Our patient, a 32 year old female, non smoker, with no known co morbid illness presented with complaints of frank hemoptysis about 80-100 ml, two episodes over two days. She gave history of dry cough for two days. There was no history of breathlessness, chest pain, wheeze, fever, loss of weight or appetite. There was no prior history of pulmonary tuberculosis. Family history was negative for tuberculosis or malignancy. On physical examination vitals were stable with blood pressure-110/80 mm Hg, heart rate-78/min and SPO2- 98%. There was no pallor, clubbing, cyanosis or lymphadenopathy. Respiratory system examination revealed occasional crackles in the left lower interscapular area. Examination of other systems was normal. Routine haematological and biochemical parameters were within normal limits. ECG showed normal sinus rhythm. Chest X-ray PA view showed a well circumscribed heterogenous opacity in the left lower zone . USG abdomen was normal. Patient was further evaluated with contrast enhanced CT chest. It showed a well defined round contrast enhancing soft tissue density lesion of size 2 X 2 cm, with attenuation value equal to aorta (HU-140) in the left lower lobe basal segmental bronchus causing luminal obstruction and distal bronchiectasis with air fluid level. The patient underwent bronchoscopy. A pink, vascular intraluminal mass lesion was seen in left lower lobe. Endobronchal biopsy of the mass was suggestive of central typical carcinoid tumor. Cardio thoracic surgeon opinion sought. Patient underwent left lower lobectomy with regional lymph node dissection. Histopathology of the resected specimen showed uniform cells with scant cytoplasm and uniform round stippled nuclei were seen. No significant pleomorphism, necrosis or mitosis was present. The findings were consistent with typical carcinoid of lung. Nodes were negative for malignancy. She was discharged one week later in good general health. Patient is under follow up.

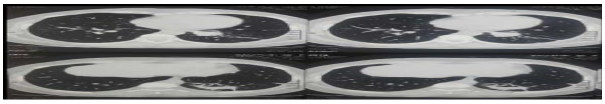

FIG 2: CT chest

FIG 3: Intraoperative specimen

Discussion:

Neuroendocrine system is made up of peptide- and amine-producing cells which have migrated from the embryologic neural crest to various organs in the body. These cells can synthesize and secrete neuroendocrine peptides into circulation4. Well differentiated neuroendocrine tumors are referred to as carcinoid tumors, gastrointestinal tract being the most common site followed by lungs. Bronchial neuroendocrine tumors(NET) account for approximately 1 to 2 percent of all lung malignancies in adults and roughly 20 to 30 percent of all NETs. Globally, the incidence of bronchial carcinoid ranges from 0.2 to 2 per 100,000 population per year2.

Risk factors:

In bronchial carcinoids, the causality of smoking is not proven. But some studies have shown an association between atypical carcinoids and smoking5. Bronchial carcinoids are usually sporadic, although 10% have features suggestive of hereditary origin. Patients with the autosomal dominant syndrome of Multiple Endocrine Neoplasia type 1 (MEN 1) have a high frequency of foregut carcinoids2. Mean age of presentation of typical carcinoids is in the fifth decade with a slight female preponderance.

Fig 4: Histopathology Classification of bronchial neuroendocrine tumors (2004 WHO criteria):

The grading system proposed for all thoracic NETs by WHO and the International Association for the study of Lung Cancer uses either mitotic rate or the presence and extent of necrosis6. NETs include Typical carcinoid, atypical carcinoid, Large cell neuroendocrine carcinoma and Small cell neuroendocrine carcinoma. Typical carcinoids lack necrosis and show less than 2 mitoses/2 mm2 .Atypical carcinoids reveal punctuate necrosis and 2 -10 mitoses/2 mm2 .Typical and atypical carcinoids are determined as low grade and intermediate grade tumors respectively based on aggressiveness.

Clinical features:

Symptoms depend on the location of carcinoid tumor. Peripheral tumors are asymptomatic. Central carcinoids present with hemoptysis, cough, wheeze, chest pain. Recurrent pneumonia in the same pulmonary segment or lobe due to bronchial obstruction is common7. Less than 5% percent exhibit hormonally-related symptoms8. Carcinoid syndrome is rare with pulmonary carcinoid because foregut carcinoids often lack aromatic amino acid decarboxylase and cannot make serotonin and its metabolites. Bronchial carcinoid is the most common cause of ectopic adrenocorticotropic hormone (ACTH) production. Both typical and atypical bronchial carcinoids can cause Cushing's syndrome due to ectopic production of ACTH. Acromegaly from ectopic production of growth hormone releasing hormone (GHRH) is a rare manifestation of a bronchial carcinoid.

Imaging:

Majority of the carcinoids cause bronchial obstruction with distal atelectasis. It can also present as hilar mass. Segmental tumors can cause mucoid impaction (bronchocele) beyond the obstruction.

Central carcinoids present as hilar mass. Some cases present as solitary pulmonary nodule. In around 10% of the cases, chest X-ray may be normal. CT chest helps in accurate localisation of both intraluminal and extraluminal components of the tumor. Marked contrast enhancement is seen3. About 80 percent of typical bronchial carcinoids express somatostatin receptors by immunohistochemistry and may be imaged with radiolabeled octreotide. Octreotide scintigraphy can image the whole body and identify metastatic disease. Since NETs have a low proliferative activity, Positron Emission Tomography is recommended only when other imaging modalities are unequivocal.

Carcinoid syndrome:

Foregut carcinoids (including those arising in the lung) generally have a low serotonin content . This is because foregut carcinoids often lack aromatic amino acid decarboxylase and cannot make serotonin and its metabolites (including 5-hydroxy indole acetic acid). Bronchial carcinoids produce lesser quantities of serotonin than do midgut carcinoids, accounting for a lower rate of carcinoid syndrome. It is mostly encountered in patients with liver metastasis from bronchial carcinoid. Carcinoid syndrome is caused by systemic release of vasoactive substances. Acute symptoms include cutaneous flushing, diarrhea, and bronchospasm. Long-term sequelae of prolonged elevated hormone levels include venous telangiectasias, right-side predominant valvular heart disease, and fibrosis in the retroperitoneum and other sites.

Biochemical markers:

Carcinoids express neuroendocrine markers. The sensitivity and specificity of Chromogranin A, that is widely present in secretory granules for detection of NETs ranges between 70% and 100%. Neuron specific enolase is a useful marker for follow up of patients. Clinical usefulness of Urinary 5- hydroxyindoleacetic acid (5-HIAA) in bronchial carcinoids is variable because many a times 5-HIAA levels are normal.

Bronchoscopy :

Three fourth of the carcinoids are centrally located and reachable by bronchoscopy. They are seen as pink to red vascular mass with intact overlying epithelium. Endobronchial biopsy can be taken. Brushings and washings are usually non diagnostic as the epithelium overlying the tumor is normal.

Histopathology:

Tumor is composed of cytologically bland cells containing regular round to oval nuclei with finely dispersed ‘salt and pepper’ chromatin and inconspicuous small nucleoli. The cells are usually polygonal in shape and are arranged in distinct organoid, trabecular, or insular growth patterns with a delicate vascular stroma.

Treatment:

Surgery is the treatment of choice for carcinoid tumors. Lung parenchyma preserving bronchoplastic techniques like sleeve resection, flap resection are preferred in typical carcinoids2. However when the lung distal to obstruction is affected by infective process, lobectomy and pnemonectomy are carried out. Bronchoscopy directed endobronchial resection with laser is best reserved for debilitated patients. For patients with surgically unresectable and metastatic disease cisplatin based chemotherapy and radiotherapy are reasonale palliative options. There is insufficient data to recommend the use of adjuvant therapy after complete resection of loco regional disease. Control of symptoms from overproduction of hormones can be achieved using somatostatin analoges, interferon, serotonin receptor antagonists and anti diarrheal agents2.

Conclusion:

In young patients with no known comorbid illness presenting with hemoptysis, bronchial carcinoid should be considered as differential diagnosis. In patients presenting with hemoptysis, bronchoscopy will help in narrowing down the diagnosis.

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