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Case Report

A case of carcinoid syndrome after CT guided lung biopsy of a neuroendocrine tumor ☆

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ABSTRACT

Carcinoid tumors of the lung are an uncommon malignancy that can rarely lead to carcinoid syndrome. Carcinoid tumors represent less than 1% of all lung cancers. This case describes a rare presentation in which a previously asymptomatic woman began to experience tachycardia, shortness of breath, and anxiety after biopsy of a pulmonary carcinoid tumor. Chest CT, histology, and clinical course supported the diagnosis and the patient's symptoms quickly resolved with medication. Ultimately, surgical removal of the tumor led to complete resolution of symptoms.

Introduction

Carcinoid Syndrome is a rare complication of neuroendocrine tumor caused by the release of biologically active molecules from the tumor site into the bloodstream. Patients typically present with shortness of breath, heart disease, hypotension, flushing and GI upset [1]. Carcinoid tumors represent less than 1% of all lung cancers and about 25% of all carcinoid tumors are found in the lungs [2]. The diagnosis of carcinoid syndrome is typically made through biopsy and serum measurement of 5-hydroxyindoleacetic acid (5-HIAA) levels; a specific marker for carcinoids producing serotonin [3]. While carcinoid tumors can cause systemic symptoms, the majority of cases will be asymptomatic [4]. While biopsy of carcinoid tumors has been shown to exacerbate carcinoid syndrome in other cases [5,6], to our knowledge there is no documented case of this phenomenon in a lung biopsy. We present a unique case of an asymptomatic pulmonary carcinoid tumor in which carcinoid syndrome was triggered by computed tomography (CT) guided biopsy.

Case

The patient is a 56-year-old female with a past medical history of a left lower lobe pulmonary nodule discovered on chest X-ray in 2016. The nodule was followed and showed an interval increase in size on a repeat CT scan in 2021. A positron emission tomography (PET) scan revealed hypermetabolism within the left lower lobe compatible with malignancy (Fig. 1). A

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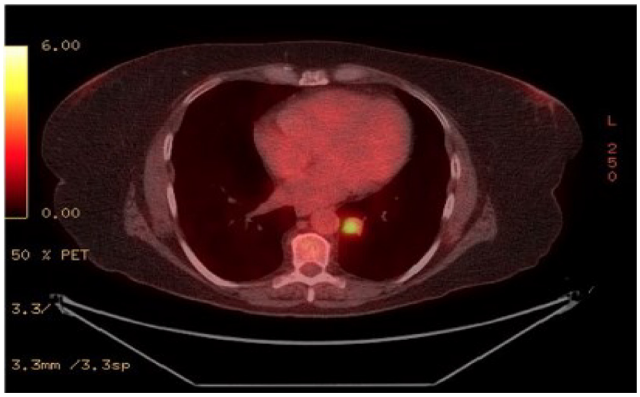

Fig. 1 – PET scan reveals area of high standardized uptake value (SUV) lateral to the descending aorta. Area of uptake coincides with known left lower lobe nodule from prior CT.

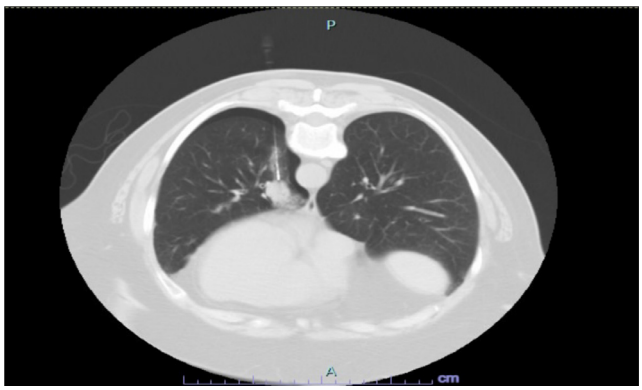

Fig. 2 – CT scan showing biopsy sample of the left lower lobe pulmonary nodule.

biopsy was ordered to confirm the findings. Subsequently, the biopsy of her left lung nodule was performed (Fig. 2) and the patient was noted to have a small pneumothorax which resolved within hours. She was discharged that same day without symptoms. Pathology report released 2 days later revealed the nodule stained diffusely positive for synaptophysin, chromogranin CD56, TTF-1, and AE1/3 indicating the nodule was consistent with a carcinoid tumor (Fig. 3). On post biopsy day 3, the patient returned to the hospital for shortness of breath, chest pain and palpitations. Initial vitals were BP: 89/65 T: 98.8°F (37.1°C) HR: 177 RR: 18 SpO2 98 % on room air. Cardiovascular exam was significant for tachycardia and an irregularly irregular rhythm. EKG showed atrial fibrillation. Laboratory results including complete blood count, comprehensive metabolic panel, troponins, d-dimer, and urinalysis were within normal limits. A diagnosis of atrial fibrillation with rapid ventricular rate was made. Hematology/Oncology was consulted and it was found that due to the patient's rapid onset of tachycardia and hypotension following the biopsy and histologic evidence of carcinoid tumor, the patient's symptoms were likely due to carcinoid syndrome exacerbated by biopsy.

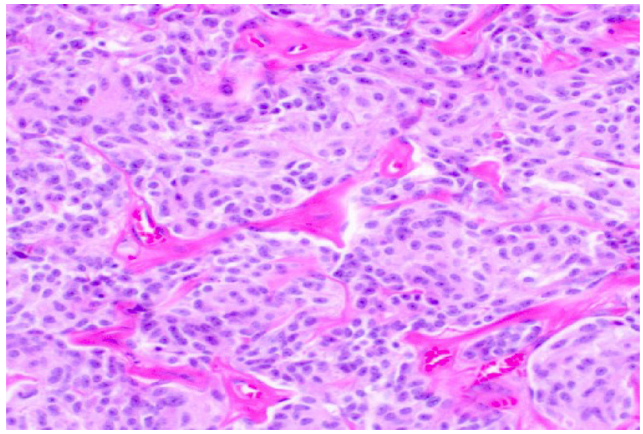

Fig. 3 – Pathological image of tumor obtained after surgical resection consistent with carcinoid tumor.

Treatment

Patient was started on diltiazem and low intensity heparin for atrial fibrillation. She was also given repeated boluses of normal saline for hypotension. By the first night of her hospital stay the patient had converted to normal sinus rhythm and her vital signs had normalized. On hospital day 2 she was discharged on low dose Metoprolol and Eliquis, and given instructions for outpatient testing of urine 5-HIAA and cortisol. She was also scheduled for left lower lobectomy for treatment of the tumor. The nodule was surgically removed a few months later without complications.

Outcome and follow-up

On post op day 1 the patient had several episodes of palpitations which were successfully treated with metoprolol. The rest of her stay was uneventful. A 6 month follow up test of urine 5-HIAA was found to be within normal limits. At the 3-year follow up, the patient had no evidence of tumor recurrence on CT and was asymptomatic.

Discussion

Carcinoid tumors are slow-growing neuroendocrine tumors that arise from amine precursor uptake and decarboxylation cells (APUD cells). Despite ongoing research, the etiology of these tumors is not completely understood. Carcinoid tumors are predominantly found in the gastrointestinal tract and are asymptomatic because most of the hormones produced are metabolized by the first-pass effect of the liver [7]. The lungs are the second most common site for neuroendocrine tumors after the gastrointestinal system accounting for 25% of all neuroendocrine tumors [8]. Neuroendocrine tumors of the lung are broadly categorized into typical carcinoid, atypical carcinoid, large cell neuroendocrine carcinoma, and small cell lung carcinoma [2].

Carcinoid syndrome occurs in 3.1%-6.7 % of affected patients and is characterized by diarrhea, flushing, and shortness of breath [5]. Palpitations and hemodynamic instability may occur if the tumor metastasizes to the liver and bypasses first-pass metabolism. Various factors including percutaneous fine needle liver biopsy, bronchoscopy, external manipulation during ultrasound studies, surgery, anesthesia, transesophageal echo and mammography have been implicated as triggers for carcinoid syndrome [5,6,9]. Notably, pulmonary carcinoid tumors are rarely associated with carcinoid syndrome. To our knowledge, there are no documented cases of acute carcinoid syndrome arising from CT guided biopsy in the literature to date.

In our patient, acute carcinoid syndrome was suspected due to clinical symptoms and histopathological confirmation after biopsy. Our patient's clinical symptoms likely stemmed from the release of vasoactive substances by the tumor upon manipulation of the nodule. We were unable to confirm the diagnosis as the patient was noncompliant with postdischarge testing instructions for urinary 5-HIAA. The severity of carcinoid syndrome is linked to the level of urinary 5-HIAA excretion [10]. Currently, there is limited literature addressing the management of acute carcinoid syndrome induced by lung biopsy. Upon onset of carcinoid syndrome, symptomatic relief, and hemodynamic stabilization can be achieved through somatostatin analogues and adequate fluid resuscitation. Surgical resection remains the primary therapeutic approach for pulmonary carcinoid tumors with 5-year survival of patients ranging from 67% to 96% [3]. Additionally, patients may benefit from a multidisciplinary approach as other management options include chemotherapy and radioligand therapy [8]. Post-treatment, regular follow-up evaluations are imperative to monitor patient progress.

In summary, the presence of solitary nodules should prompt consideration of carcinoid tumors, given the potential for acute carcinoid syndrome. Adequate caution must be exercised during biopsy, and patients should be educated on symptom recognition, as timely detection of acute carcinoid syndrome is crucial for effective management. If carcinoid syndrome is suspected, endocrinological workup including 5-HIAA, serotonin, chromogranin A should be performed immediately [8]. Furthermore, a workup should be performed to rule out systemic metastases or local spread. Treatment strategies encompass a multidisciplinary approach, incorporating surgical resection, somatostatin analogues and chemotherapy to control symptoms and optimize patient outcomes.

Patient consent

Written consent was obtained from the patient by the attending physician for this case and can be provided upon request.

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