Peripheral Bronchial Carcinoid Tumor Presenting as a Right Cardiophrenic Angle Mass

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A 50-year-old woman presented for evaluation of an enlarging right cardiophrenic angle mass. Two years prior she complained of intermittent nausea, diarrhea, and flushing. Initial chest radiography and computed tomography (CT) suggested a pericardial cyst. Due to the onset of increasing dyspnea on exertion, lower extremity edema, and weight gain repeat CT was performed revealing a solid tumor. An Indium-111 octreotide scan showed somatostatin activity limited to the pericardiac mass. Histology after resection confirmed the diagnosis of peripheral bronchial carcinoid. The traditional differential diagnosis for a right cardiophrenic angle mass was misleading in this patient.

Introduction

The differential diagnosis for a right cardiophrenic angle mass includes both benign and malignant entities. The most common lesions are pericardial cyst, pericardial fat pad, and foramen of Morgagni hernia. Bronchial carcinoid tumors do not typically present with symptoms of the carcinoid syndrome and most patients are asymptomatic for several years before presenting with symptoms of endobronchial obstruction.
Case Report

A 50-year-old woman with obesity, hypertension, and severe sleep apnea presented for a second opinion regarding an enlarging right cardiophrenic angle mass. She suffered from intermittent nausea, vomiting, and diarrhea two years prior which improved after a partial colectomy for adherent endometriosis. She had also complained of light headedness and flushing accompanying her nausea with an associated syncopal episode 1 year prior. She had recently developed a four week history of increasing dyspnea on exertion, lower extremity edema and weight gain. She denied any fevers, chills or night sweats.

During her evaluation a chest mass was present on radiography (Fig. 1A), but its encapsulated appearance and low attenuation on initial computed tomography (CT) of the thorax (not shown) was thought to represent a pericardial cyst. A second CT scan (Fig. 1B) revealed that the right middle lobe mass had an attenuation of 40 Hounsfield units (HU) in the arterial phase, 61 HU in the venous phase, and 51 HU in the delayed equilibrium phase. Enhancement was only apparent when measured directly on the imaging workstation. This low level of enhancement and tumor wash-out was interpreted as consistent with a solid neoplasm. CT of the abdomen and pelvis was negative for other possible primary neoplasms.

Since the mass was abutting the anterior pleural surface, ultrasound-guided needle biopsy was performed. The morphologic features and strong positivity for neuroendocrine markers on immunohistochemical staining were consistent with carcinoid tumor. An Indium-111 octreotide scan (Fig. 1C), performed after initial biopsy to further characterize the tumor, was positive for somatostatin receptor activity limited to the pericardiac mass. Urine 5-hydroxyindole acetic acid (5-HIAA) levels and plasma serotonin levels were not elevated at the time of evaluation. Thoracoscopy and wedge resection revealed that the tumor originated from a right middle lobe bronchiole and was partially adherent to, but did not invade the pericardium. Pathologic evaluation of the mass confirmed peripheral bronchial carcinoid tumor (grade I neuroendocrine carcinoma).

Discussion

The classic differential diagnosis for a right cardiophrenic angle mass includes pericardial fat pad, pericardial cyst,
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Bronchial carcinoid tumors represent approximately 2% of all primary lung tumors [2]. They usually present in the fourth or fifth decade and have an equal incidence in men and women [3]. They are classified as central or peripheral based on location in the lung. Central bronchial carcinoid tumors typically present as endobronchial masses with symptoms of obstruction such as wheezing, coughing, dyspnea and lobar infection [4]. Peripheral carcinoid tumors originate in peripheral bronchioles and can present as pericardiac masses. Bronchial carcinoids originate in the bronchial mucosa from neuroendocrine cells termed Kulchitsky cells [5]. Bronchial carcinoids are further classified as typical or atypical depending on their level of differentiation and potential for aggressive behavior.

Bronchial carcinoids usually present with respiratory symptoms from endobronchial obstruction without the classic symptoms of carcinoid syndrome (diarrhea, flushing, and wheezing). The carcinoid syndrome occurs in less than 5% of pulmonary carcinoid cases [6]. Carcinoid heart disease seen in association with carcinoid tumors of abdominal origin is characterized by fibrous lesions on the right-sided cardiac chambers and valves and less-commonly left-sided cardiac lesions as vasoactive substances released by the tumor are inactivated in the pulmonary capillaries. However, patients with pulmonary carcinoids more often present with left-sided cardiac plaques as serotonin released by the tumor accesses the left heart directly via the pulmonary venous drainage before it can be metabolized in the pulmonary capillaries [7]. Bronchial carcinoids are usually slow-growing, encapsulated tumors amenable to surgical resection. Ectopic corticotrophin secretion from bronchial carcinoids accounts for 1% of all Cushing’s syndrome cases [8].

On CT, bronchial carcinoids are usually smooth, round, and can be lobulated or contain calcifications. Carcinoid tumors are usually vascular and enhance markedly on CT. A study of 12 surgically proven pulmonary carcinoid tumors had an average CT attenuation after contrast enhancement of 80.5 to 179 HU [9]. In the case we present, initial CT of the chest showed minimal enhancement due to delayed scanning after the contrast bolus. On repeat CT, tumor enhancement was still less than usually expected with carcinoid tumors probably due to properties of the tumor itself. Although no areas of ischemia or frank necrosis were identified, regions of cellular devitalization and non-necrotizing granulomas were present in the pathology specimen.

Exacerbation of the patient’s symptoms and growth of the tumor resulted in aggressive management leading to the correct diagnosis. The traditional differential diagnosis for a right cardiophrenic angle mass was misleading in this patient. Initially the patient was thought to have a pericardial cyst and while pericardial cysts can have high attenuation on noncontrast CT, it is important to examine post-contrast images carefully to appreciate subtle enhancement to diagnose an unsuspected solid mass. Proper timing of CT scanning after the contrast bolus is also critical to detect enhancement of a neoplasm. Differentiating between a benign and malignant pericardiac process was essential in the proper management of this patient.

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