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Case Report

Incidentally Found Aorto-Pulmonary Middle Mediastinal Hypervascular Mass on Pulmonary CT Angiography in a Covid 19 patient

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Abstract

Paragangliomas (PGs) are rare neuroendocrine tumors arising from paraganglia, clusters of neuroendocrine cells scattered throughout the body. Mediastinal paragangliomas represent less than 2% of all paragangliomas and less than 0.3 % of the mediastinal tumors. These tumors may secrete catecholamines, however in up to 50% of cases they are nonfunctional and are diagnosed incidentaly or with symptoms of mass effcet to the adjacent structures. They should be considered in the differential diagnosis of hypervascular mediastinal masses. The typical radiological features are very guiding in the diagnosis and the management of the patient. We aimed to present a case of a hypervascular middle mediastinal mass incidentally found in a 69 year old woman and diagnosed with aortopulmonary nonfunctional PG radiologically.

Keywords: mediastinal paraganglioma; glomus tumor; hypervascular mediastinal mass

Introduction

Paragangliomas (PGs), also sometimes called as glomus tumors, are rare neuroendocrine tumors arising from paraganglia, which are clusters of neuroendocrine cells scattered throughout the body with the largest cluster found in the adrenal medulla. These cells are closely related to the autonomic nervous system, with either parasympathetic or sympathetic function. While the parasympathetic PGs are generally nonsecretory and present usually with mass-effect such as cranial nerve palsies, the symphathetic paragangliomas tend to be functional and secrete catecholamines resulting in presentations with palpitations, hypertension, headaches, hyperhidrosis and diabetes which resolves after the complete resection of the PG [1,2] Symphathetic PGs can be intraadrenal which are called as pheochromocytoma or extra-adrenal which can be located in the abdomen or thorax (mediastinal PG) [3]. Mediastinal PGs represent less than 2% of all PGs and less than 0.3% of the mediastinal tumors [4]. We present a case of a hypervascular middle mediastinal mass incidentally found in a 69 year old woman and diagnosed with aortopulmonary nonfunctional PG radiologically.

Case Resentation

A 69 year old female patient presented with cough, mild fever and chest pain. She had recent history of Covid-19 pneumonia about 8 months ago. Her laboratory results revealed increased level of D-dimer and Ferritin with slight increase in WBC count and CRP. There were no known any other diseases or primary malignancy. We performed IV contrast enhanced chest computed tomography [CT] for her presenting symptoms, which revealed a well defined middle mediastinal mass between left pulmonary artery and the descending aorta showing intermediate soft tissue density on precontrast images without any calcifications **(fig. 1a).** The size of the mass was abouut 3x3 cm and showed no enhancement on pulmonary arterial phase images **(fig.1b)** and heterogeneously intense enhancement on early arterial phase images **(fig. 1c)** with multiple serpentine tubular enhancing vasculature within the lesion. There were no accompanying pulmonary parenchymal nodules/masses, pathologic mediastinal lymph nodes or any parenchymal infiltration. Although we did not concerned malignancy and metastases, they were ruled out by positron emission CT (PET-CT). Our main differential diagnosis included mediastinal PG and mediastinal hemangioma. Due to the characteristic location of the mass in the middle mediastinum, the heterogenous serpentine contrast enhancement pattern immediately at the early arterial phase CT images and the lack of phlebolit on nonenhanced images, our initial diagnosis was mediastinal PG. Although there were no symptoms suggesting increased catecholamine secretion and her 24 hour urine vanillyl mandelic acid level was within normal limits, excision of the mass was recommended rather than biopsy because of hypervascularity and close proximity of the lesion to the great vessels. However, she refused operation due to her asymptomatic course and wanted to be followed up.

*informed consent has been taken from the patient.

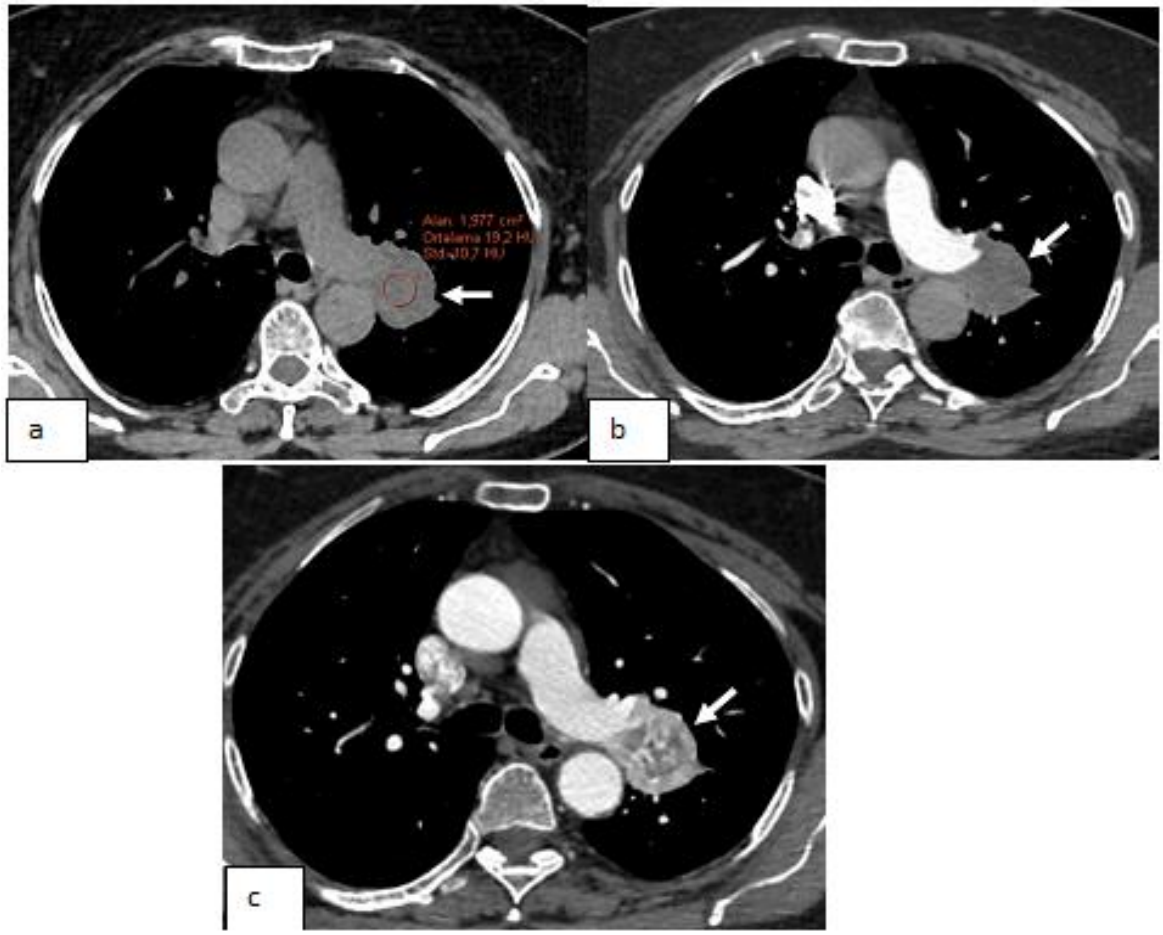

Figure 1a: Axial precontrast CT image and **b,c** Axial Pulmonary CTA images showing a well-defined middle mediastinal mass between left pulmonary artery and the descending aorta with intermediate soft tissue density on precontrast **(a, arrow)** without any calcifications and pulmonary arterial phase images **(b, arrow)** without enhancement. Heterogeneously intense enhancement on early arterial phase images with multiple serpentine tortious tubular enhancing vasculature within the lesion is demonstrated **(c, arrow).**

Discussion

Mediastinal PGs are rare tumors that can arise from two major clusters of sympathetic paraganglion cells including aortosympathetic paraganglia in the posterior mediastinum [paravertebral] and aortopulmonary paraganglia in the middle mediastinum [great vessels of the chest]. Additionally, although extremelly rare the third type of the mediastinal PG is cardiac PG [5]. These tumors may secrete catecholamines, however in up to 50% of cases they are nonfunctional. Most patients are asymptomatic and the diagnosis is incidental [1, 6]. Others may present with symptoms of local mass effect to the adjacent structures resulting in hoarseness, dysphagia, shortness of breath and chest pain [1,7]. In addition, PGs may develop distant metastases occuring most commonly to lymph nodes, liver, lung, and bone. Since there are no reliable histological marker for their malignant potential, the presence of metastases can be the only evidence for malignancy [8]. When the primary diagnostic concern is PG, biopsy is contindicated until biochemical screening is negative for catecholamine excess PGs are seen in a characteristic location in the paraaortic region of the middle mediastinum or paravertebral region of the posterior mediastinum corresponding to major cluster of sympathetic ganglion cells [3]. On nonenhanced CT they have typically intermediate density and show heterogenous intense enhancement following contrast administration. On MRI, they are usually markedly hyperintense on T2-w images representing light bulb appearance [9]. Larger ones display salt-and-pepper appearance referring to slow flow within dilated vascular structures or foci of hemorrhages (salt) and flow voids within high flow vasculature [pepper] [7]. Following contrast administration due to the presence of rich capillary network arising generally from internal mammmary artery, they demonstrate intense heterogenous, prolonged enhancement and delayed washout on dynamic imagings [9]. Differential diagnoses include mediastinal hemangioma and hypervascular metastases such as from RCC [10]. In hypervascular mediastinal metastasis, there is usually a known previous history of primary malignancy and the lesions are usually more than one in number with frequently accompanying pulmonary parenchymal involvement. In our patient there were no history of malignancy and thoracoabdominal imagings revealed no additional pathology. Therefore, our main differential diagnosis was mediastinal hemangioma. However, the middle mediastinum is the least common location for mediastinal hemangiomas with more than 50% of mediastinal hemangiomas being located in the anterior mediastinum [11, 12]. Phlebolithes can be characteristically demonstrated on nonenhanced images in cases of mediastinal hemangiomas [13]. In addition, although not always, hemangiomas, if larger than 1.5 cm in diameter are expected to demonstrate peripheral nodular discontinuous enhancement on early arterial phase images and progressive centripetal fill-in on delayed phase images over time. With the consideration of excessive ionising radiation we did not perform dynamic contrast enhanced CT. In our patient, due to the characteristic location of the mass in the middle mediastinum, the

heterogenous serpentine contrast enhancement pattern immediately at the early arterial phase CT images rather than only peripheral pathcy nodular enhancement and the lack of phlebolit on nonenhanced images, we diagnosed radiologically the lesion as mediastinal PG. Although the PG has been showed to be nonfunctional, we have recommended surgical excision rather than biopsy due to the hypervascularity of the lesion and its close proximity to the great vessels. She refused operation due to her asymptomatic course and wanted to be followed up.

Conclusion

Mediastinal PGs are rare lesions which constitute less than 0.3 % of the mediastinal tumors. However, they should be considered in the differential diagnosis of hypervascular mediastinal masses. The typical radiological features are very guiding in the diagnosis and the management of the patient

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