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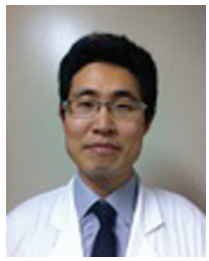
Anterior Mediastinal Paraganglioma Mimicking Thymoma



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ABSTRACT

A 54-year-old man was referred to our institution with hemoptysis and hoarseness of 1 year's duration. A computed tomography (CT) scan showed an anterior mediastinal mass (2.5 cm × 1.0 cm), which was diagnosed as thymoma. The tumor was resected under a sternotomy. The tumor had invaded the anterior wall of the ascending aorta. With the patient under cardiopulmonary bypass, the aortic wall invaded by the mass was resected, and arterial reconstruction was performed with patch material. The tumor was revealed to be a tumor of neuronal origin. The patient's postoperative course was uneventful. The patient was discharged on postoperative day 9. One year after the operation, a follow-up chest CT evaluation showed no specific complications or recurrence.

INTRODUCTION

Paraganglioma is a rare tumor that arises from extra-adrenal chromaffin cells in the autonomic nervous system [Herrera 1993]. Paragangliomas represent 10% to 18% of all chromaffin tissue–related tumors and affect a wide variety of sites, such as the abdomen, pelvis, neck, and mediastinum. Mediastinal paragangliomas originate from para-aortic (middle mediastinum) and para-vertebral (posterior mediastinum) sympathetic chain ganglia [Young 2006; Balcombe 2007].

A number of para-aortic paragangliomas have been reported; however, a paraganglioma mimicking a thymoma has never been reported. We present a case of an aortic paraganglioma that was initially misdiagnosed as a thymoma.

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CASE REPORT

A 54-year-old man was referred to our institution in May 2005. The chief complaints were hemoptysis of approximately 70 mL and hoarseness for 1 year. His vital signs were stable, and he had no symptoms. No other medical history was relevant except for hypertension of 15 years.

A computed tomography (CT) scan revealed a well-enhanced anterior mediastinal mass 2.5 cm × 1.0 cm in size (Figure 1). Increased fluorodeoxyglucose uptake of the anterior mediastinal mass was observed in a positron emission tomography examination (Figure 2). The mass was diagnosed as thymoma, and surgery was planned.

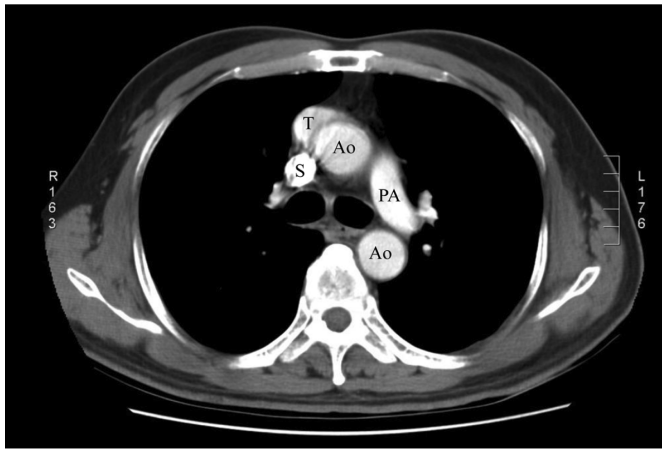

Figure 1. Axial computed tomography scan showing the tumor (T) adjacent to the aorta (Ao), the superior vena cava (S), and the pulmonary artery (PA).

The operation was performed via a median sternotomy. The thymus was dissected from the pericardium, and the innominate vein was partially resected and sutured, because the tumor was located close to the innominate vein. The tumor had invaded the anterior wall of the ascending aorta. With the patient under cardiopulmonary bypass, the aortic wall was resected, and arterial reconstruction was performed with patch material.

The tumor was 2.5 cm × 2.3 cm × 1.1 cm and was a hard mass. Immunohistochemical studies revealed positivity for

chromogranin, synaptophysin, and S-100 protein. These results confirmed the tumor to be neuronal in origin. The patient's postoperative course was uneventful. The patient was discharged on postoperative day 9. A follow-up chest CT evaluation 1 year after the operation showed no specific complications or recurrence.

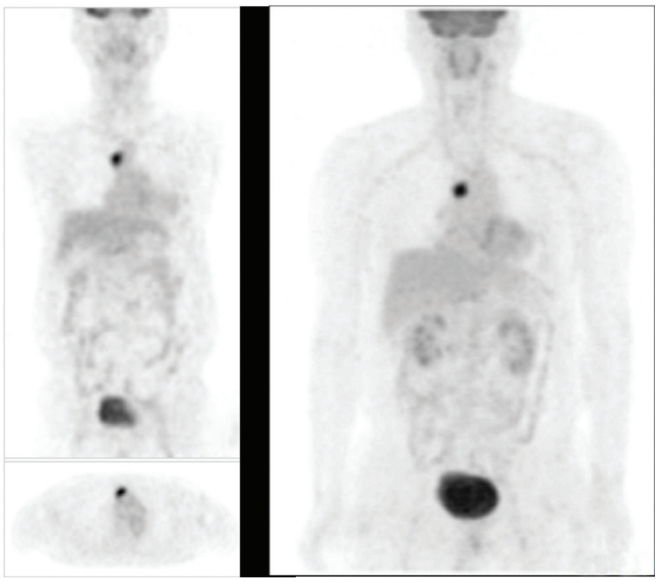

Figure 2. Image of positron emission computed tomography scan showing the anterior mediastinal mass with its increased fluorodeoxyglucose (FDG) uptake.

DISCUSSION

Paragangliomas are endocrine tumors that arise from paraganglionic tissue. Mediastinal paragangliomas are rare tumors that tend to remain asymptomatic until discovered incidentally or until they grow to a size large enough to cause symptoms. The majority (75%) of paragangliomas arise in the anterior or middle mediastinum.

Paragangliomas arise from the adrenal medulla, the carotid and aortic bodies, and the organs of Zuckerkandl [Sclafani 1990]. Paragangliomas are diagnosed incidentally in most patients, or the mass causes symptoms that are recognized. Fewer than 2% of paragangliomas are functional tumors arising in the chest [Herrera 1993], so the clinical presentation, such as hypertension, and symptoms such as headache, palpitations, or sweating caused by catecholamine hypersecretion are generally absent.

A 24-hour urinary metanephrine and catecholamine measurement is useful for diagnosing functional tumors. In our case, however, these levels were not checked because the patient was asymptomatic and the tumor was thought to be a thymoma.

Paragangliomas are highly enhanced on CT scans except for poor enhancement of central necrotic areas, and they are usually located in the bifurcation of great vessels. Uniform and homogeneously enhanced masses have also been reported [Balcombe 2007]. Thymomas are the most common primary tumor of the anterior mediastinum. Thymomas are round or lobulated, homogeneous-enhanced masses on CT scans, but they can be heterogeneous, or even cystic, because of areas of hemorrhage and necrosis [Marom 2010]. In our case, the mass was initially diagnosed as a thymoma because of its higher prevalence and similar appearance in a contrast CT scan of the chest.

Pathologic confirmation is crucial to diagnose paragangliomas, because of their ambiguous characteristics on imaging studies. The pathologic characteristics of a paraganglioma are its highly vascular nature and clusters of cells with fine granular eosinophilic cytoplasm occurring between the many capillaries [Bird 1991]. Immunohistochemical positivity is essential to diagnose a paraganglioma.

Complete surgical resection with vascular reconstruction is recommended, because the tumors are relatively resistant to chemotherapy and radiation. Because these tumors are high vascular and usually located near great vessels, resection with the patient under cardiopulmonary bypass is frequently mandatory. Low-dose α-adrenergic blocking agents should be used, because hemodynamic instability may occur with clinically and biochemically silent paragangliomas [Erickson 2001]. Fortunately, our patient showed no hemodynamic instability during the operation.

A paraganglioma in the anterior mediastinal region is rare but can mimic other tumors, such as a thymoma. Paragangliomas may cause hormonal-related crises associated with significant morbidity and mortality [Erickson 2001; Young 2006]. Surgical resection is challenging because of the high degree of vascularity. A paraganglioma should be considered in the diagnosis when an anterior mediastinal mass is observed.

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