A 24-YEAR-OLD WOMAN presented to an outside hospital during her third trimester of pregnancy with hypertensive episodes (230/130 mm Hg), headaches, and palpitations. Pre-eclampsia was excluded because of the absence of proteinuria. Additional evaluation revealed markedly elevated 24-h urinary catecholamines (norepinephrine, 978 μg/24 h [0–100]; normetanephrine, 4067 μg/24 h [50–650]; epinephrine, < 3 μg/24 h [0–25]; metanephrine, 108 μg/24 h [30–350]; and vanillylmandelic acid, 13.6 mg/d [0–7]), which confirmed the diagnosis of pheochromocytoma. A magnetic resonance image (MRI) of the abdomen showed a 2.8 × 1.9-cm mixed-intensity mass medial to the left adrenal gland, which was presumed to be a pheochromocytoma. The patient was brought to term safely on phenoxybenzamine therapy (10 mg twice daily), and a healthy baby was delivered via elective cesarean section. Postpartum, the patient underwent an elective left laparoscopic adrenalectomy. However, pathology revealed only normal adrenal tissue, and the patient’s clinical syndrome did not resolve. Genetic testing for a RET proto-oncogene mutation was negative, and the patient was referred for additional evaluation.

During additional questioning, the patient reported having undergone prior surgery for a right neck mass, and her estranged father had been treated for a similar neck mass as well as an abdominal growth. Clinical suspicion for familial disease prompted functional imaging with 18F-DOPA positron emission tomography (PET)/computed tomography (CT), which revealed a posterior mediastinal mass with intense radiotracer uptake (Fig 1). Cardiac MRI and transesophageal echocardiography demonstrated a 4.5 × 2.7 × 3.9-cm mass abutting the postero-lateral aspect of the left atrium and the right inferior pulmonary vein (Fig 2). After preoperative medical conditioning with phenoxybenzamine and metoprolol, the patient underwent a right anterolateral thoracotomy. Cardiopulmonary bypass was instituted to expose the left atrium and to identify the tumor, which was found to originate directly from the left atrial endothelium near the right inferior pulmonary vein orifice. It was resected along with a portion of the atrial wall, and the defect was reconstructed using bovine pericardium. The patient recovered uneventfully; her urinary catecholamine levels rapidly normalized, and she became normotensive without the need for any medications. A postoperative echocardiogram demonstrated normal cardiac function. The pathology of the resected atrial tumor revealed a 3.5-cm paraganglioma with positive staining for S-100 and synaptophysin. Genetic testing revealed a mutation in succinate dehydrogenase subunit B (c.725 G → A) with a normal subunit D.

**DISCUSSION**

Pheochromocytomas are catecholamine-secreting tumors that develop from chromaffin cells in the adrenal medulla. Extra-adrenal pheochromocytomas, which are called paragangliomas, occur in the ganglia of the autonomic nervous system and represent approximately 18% of all pheochromocytomas. Cardiac paragangliomas are extremely rare. About 50 cases have been reported in the literature, and the left atrium is the most common site of origin (55% of cases). To our knowledge, the current case is the first description of cardiac paraganglioma that developed in the setting of a demonstrated genetic syndrome.

Paraganglioma syndrome (PGL) is characterized by multiple neoplasms of the paraganglia.
(chromaffin tissue). PGL is divided into 4 types based on the underlying genetic mutation. Three of the 4 types are caused by mutations in the subunits (A, B, C, and D) of succinate dehydrogenase (SDH), which is a key enzyme in the Krebs cycle and the mitochondrial electron transport chain. PGL1 is caused by mutations in SDHD, PGL2 is caused by mutations in an unidentified gene on chromosome 11, PGL3 is caused by mutations in SDHC, and PGL4 is caused by mutations in SDHB. 3 Although patients with PGL1-SDHD commonly have head and neck paragangliomas that are generally benign, patients with PGL4-SDHB can have abdominal and thoracic paragangliomas with greater malignancy potential. Half of patients with PGL4-SDHB develop disease prior to age 26. Up to 75% of the tumors are extra-adrenal in origin, and one third of affected patients develop metastases. 4 In the current case, localization of the patient’s tumor was achieved with the use of 18F-DOPA PET-CT, which is a novel functional scan with high sensitivity and specificity for detection of pheochromocytoma.

**REFERENCES**