A 16-year-old adolescent boy was admitted to a local hospital for surgical repair of an orbital fracture. He was seen by a pediatric cardiologist at 12 years of age for evaluation of palpitations and syncope, which was diagnosed as a vasovagal episode. He has had no subsequent symptoms or other health concerns and has excelled in sports. During recovery from the orbital fracture surgery, he was noted to have irregular heart rhythm and was subsequently referred for outpatient pediatric cardiology evaluation. His vital signs and physical examination were normal. Echocardiogram revealed a 2.5×2.6-cm mass between the anterior aspects of the proximal ascending aorta and main pulmonary artery with no obstruction to blood flow and no additional structural or functional abnormalities (Movie I in the online-only Data Supplement). Chest computed tomography identified small blood vessels from the proximal left and right coronary arteries entering the tumor. Cardiac magnetic resonance sequences were consistent with a high vascular supply to the tumor: hyperintense signal on T2-weighted turbo spin echo sequence, brisk perfusion of contrast on first-pass perfusion imaging (Movie II in the online-only Data Supplement), and hyperenhancement of the tumor periphery on late gadolinium enhancement imaging (Figure 1). Surgical resection of the tumor was recommended for definitive tissue diagnosis because the possibility of malignancy could not be excluded. Preoperative selective coronary angiography defined the feeding vessels from the left and right coronary arteries to the tumor (Figure 2 and Movies III and IV in the online-only Data Supplement).

Operative findings confirmed a 2×4-cm solid, encapsulated tumor attached to the base of the aorta and pulmonary root (Figure 3). The tumor was removed in its entirety, and nearby lymph nodes were biopsied. Histology of the tumor confirmed the diagnosis of paraganglioma with capsular invasion (Figure 4). The postoperative course was uneventful with discharge on the third postoperative day.

Figure 1. Cardiac magnetic resonance images of paraganglioma (arrows). A, Cine steady state free precession in the axial plane demonstrates a well-circumscribed tumor between the aorta and main pulmonary artery. B, Gadolinium first-pass perfusion imaging demonstrates brisk perfusion of the tumor (Movie II in the online-only Data Supplement). C, T1-weighted turbo spin echo imaging with fat suppression shows vascular channels in the tumor. D, T2-weighted turbo spin echo demonstrates hyperintense signal. E, Late gadolinium imaging sequence shows peripheral hyperenhancement of the tumor.
oncology evaluation, including chest computed tomography, \([^{123}\text{I}]\)m-iodobenzylguanidine, and \(^{18}\text{F}\)-labeled deoxyglucose positron emission tomography scans, found no evidence of metastases. Genetic testing revealed a succinate dehydrogenase subunit B gene mutation, which is associated with type 4 paraganglioma syndrome.\(^1\)

Paraganglioma is a rare neuroendocrine tumor that arises from the sympathetic paraganglia and can occur in various locations throughout the body.\(^2\) The tumor may or may not secrete catecholamines such as norepinephrine and can be metastatic. The clinical features of secreting tumors are similar to those of pheochromocytoma, including systemic hypertension, headaches, palpitations, chest pain, flushing, and sweating.\(^3\) Cardiac paraganglioma is particularly rare, with \(\approx 85\) reported cases ranging from 8 to 79 years of age.\(^4\) In our patient, this incidental finding led to the diagnosis of a succinate dehydrogenase subunit B mutation, which is associated with an increased frequency of malignant disease.\(^1\) From the standpoint of diagnostic imaging, cardiac paraganglioma should be considered in the differential diagnosis of tumors with rich vascular supply. This case highlights the importance of tissue diagnosis, given that its features on echocardiography, computed tomography, cardiac magnetic resonance, and angiography were indistinguishable from those of benign vascular tumors such as cardiac hemangioma.

**Disclosures**

None.

**References**


**Figure 2.** Selective coronary angiography of the right (A) and left (B) coronary arteries demonstrates multiple feeding vessels to the paraganglioma (arrows).

**Figure 3.** Intraoperative photograph of paraganglioma. Ao indicates aorta; RAA, right atrial appendage; and RVOT, right ventricular outflow tract.
Figure 4. Histology of paraganglioma. A, The bisected mass is well circumscribed and appears encapsulated. The cut surface is tan and has multiple dilated vascular channels (arrows). B, The tumor is composed of nests of cells (zellballen) bordered by delicate, slit-like vascular channels (arrows). The cells have a modest amount of eosinophilic cytoplasm and round to oval, bland nuclei. Occasional cells had larger nuclei (not depicted) (hematoxylin and eosin stain). C, The tumor cells show strong, diffuse cytoplasmic staining for chromogranin (immunoperoxidase stain). D, Sustentacular cells (arrows) surrounding the nests of tumor cells are strongly positive for S100, whereas the tumor cells have weak cytoplasmic and nuclear staining (immunoperoxidase stain). E, Succinate dehydrogenase B shows strong cytoplasmic staining in only a minority of cells (arrow), suggestive of succinate dehydrogenase gene mutation (immunoperoxidase stain). F, The proliferative index, as indicated by nuclear staining with Ki-67, ranged from <5% to focally up to 20%. In this field, the proliferative index is ~15% (immunoperoxidase stain).
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