Giant primary paraganglioma of the left ventricle

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Primary paraganglioma (PP) of the cardiac structures is extremely rare.[1] This tumor is highly vascular, with a high tendency toward fatal hemorrhagic complications and invasion of the surrounding tissues.[2] Its most frequent location has been shown to involve the interatrial septum, cardiac atria, intrapericardial aorta, and pulmonary artery.[1,3,4] We present a case of a patient with a giant PP with an unusual localization in the left ventricle.

CLINICAL SUMMARY

A 50-year-old woman was admitted to our department with left-sided chest pain and dyspnea at rest. There was no past medical history of cardiac disease, and the physical examination was unremarkable. Transthoracic echocardiography was performed, which revealed a large cardiac mass adherent to the left ventricle. The patient underwent computed tomographic ultrafast scanning, which showed the vascular nature of the tumor. No other intrathoracic masses were identified. Cardiac catheterization showed no concomitant coronary artery disease and identified 2 feeding vessels: a branch originating from the proximal portion of the right coronary artery, which was the main vascular supply, and a second perfusion branch from the circumflex artery (Figure 1). All additional hematologic and biochemical investigations were negative in terms of markers for neuroendocrine tumor and urinary catecholamine levels.

Surgical intervention was performed through a standard median sternotomy on extracorporeal circulation and with cardiac arrest. At the operation, the paraganglioma was shown to be deeply embedded in the left ventricular wall, mainly involving the basal and middle segments of the anterolateral wall (Figure 2, *A* and *B*). A cleavage plane was found at the subepicardial level, just underneath the last portion of the coronary main trunk and anterolateral vessels. Meticulous dissection and excision was carried out, taking care to preventively ligate and cauterize the numerous feeding vessels encountered along the wide area involved (Figure 2, *C*). The excised paraganglioma was an 8 × 7–cm mass. Histology confirmed the preoperative diagnosis of PP with pleomorphic nuclei and rare mitosis. The tumor cells were arranged in balls and clusters separated by endothelial-lined spaces with a classic pattern characteristic termed ''Zellballen.'' Immunohistologic staining was positive for chromogranin, synaptophisin, and neuron-specific enolase. Positive staining to neurofilaments and protein S-100 was also shown. The margin of resection was negative for tumor.

After mass excision, the epicardial layer was approximated to the underlying muscle surface to fill in the dead space and to enhance hemostasis. Biologic glue (Flowseal, Baxter AG) was also applied afterward. The postoperative course was uneventful. The patient was extubated after 8 hours, readmitted in the ward on the first postoperative day, and discharged on the fifth day after the operation. Transthoracic echocardiographic analysis before discharge showed preserved biventricular contractility. The patient has been recently assessed at 1 year from surgical intervention. Computed tomographic scanning has shown neither recurrent neoformation at the site of surgical excision nor evidence of metastasis. Finally, global left ventricular performance was maintained at transthoracic echocardiography.

DISCUSSION

PP of the chest is extremely rare, with less than 0.1% of the hypertensive population being affected.[1] Its frequent extra-adrenal location is in the posterior mediastinum.

Primary PPs originating from the heart are uncommon, and their treatment is similar to that of all other cardiac tumors: they should be completely excised when possible.[3] Furthermore, many cardiac paragangliomas are functionally active and can also cause symptoms related to catecholamine excess, including palpitations, headache, and sweating,[4] and in these cases preoperative β- and α-blockade should be considered.

Our patient showed an unusual ventricular localization of a PP with a high vascularity of the tumor deriving from the coronary system. This abnormal perfusion by coronary arteries, together with a tendency of the tumor to be strictly adherent to the surrounding structures without a well-defined capsule, made the extracorporeal circulation with cardiac arrest necessary for the total resection of the PP. Mass removal required meticulous dissection, vessel visualization and ligation, and final coverage of the dissected area to avoid life-threatening hemorrhage. The tumor was manipulated with great care to avoid sudden hypertension caused by norepinephrine release. If a complete excision of the tumor is possible, long-term prognosis is good. Concern exists about long-term prognosis in patients in whom the tumor was

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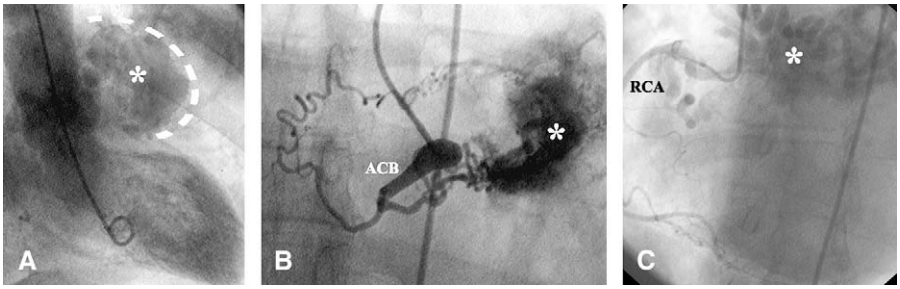

FIGURE 1. Preoperative coronary and aortic angiogram. A, Left ventriculography and related aortic angiographic analysis showing the location of the primary paraganglioma and the anatomic relationship with the cardiovascular structures. B, Perfusion of the primary paraganglioma from a branch of the circumflex coronary artery. C, A second perfusion branch of the primary paraganglioma arises from the proximal portion of the right coronary artery, showing a tortuous course and initial signs of vessel aneurysm.

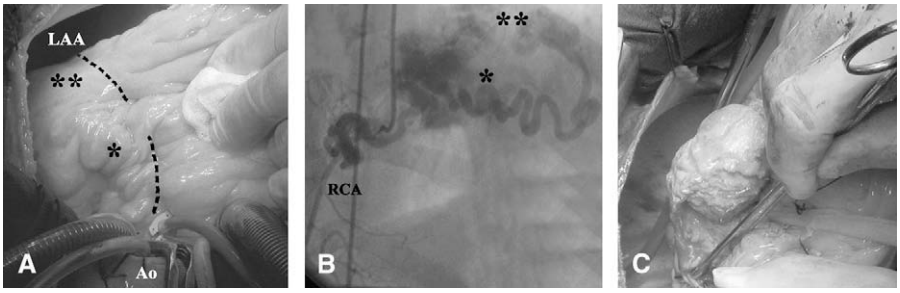

FIGURE 2. Intraoperative view of the tumor. Panel A shows the ventricular vision after sternotomy and pericardiotomy: the ventricular mass is evident *(double asterisk)*, showing a coronary branch *(asterisk)* coming from the right coronary artery. Panel B shows the entire course of the main feeding branch of the primary paraganglioma, coming from the right coronary artery and ending in a sort of vascular fistula. C, The tumor excision is documented, showing the intimate relationship and depth at the left ventricular level.

unresectable. The use of chemotherapy and radiotherapy, in such cases, is of limited value.[5]

References

1. Lee HH, Brenner WI, Vardhan I, Hyatt J, Terlecki M. Cardiac pheochromocytoma originating in the interatrial septum. *Chest*. 1990;97:760-2.
2. Orringer MB, Sisson JC, Glazer G, et al. Surgical treatment of cardiac pheochromocytomas. *J Thorac Cardiovasc Surg*. 1985;89:753-7.
3. Chan JKM, Pontefract D, Andrews R, Naik SK. Paraganglioma of the left atrium. *J Thorac Cardiovasc Surg*. 2001;122:1032-3.
4. Jebara VA, Uva MS, Farge A, et al. Cardiac pheochromocytomas. *Ann Thorac Surg*. 1992;53:356-61.
5. Vander Salm TJ. Unusual primary tumors of the heart. *Semin Thorac Cardiovasc Surg*. 2000;12:89-100.

A case of antiphospholipid syndrome presenting with a floating thrombus in the ascending aorta

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We report the case of a patient with primary antiphospholipid syndrome (APS) with a presenting symptom of dyspnea. Transesophageal echocardiography and aortic dissection computed tomography showed a floating thrombus in the distal ascending aorta.

CLINICAL SUMMARY

A 57-year-old man was admitted with the symptom of gradual dyspnea for 1 month. He had no medical history of