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Research Paper Medical Science

Obstructive Paraganglioma of the Right Ventricle A Rare Primary Cardiac Neoplasm.

Miguel Maluf, MD PhD, Associate Professor, Cardiovascular Division - São Paulo Federal University - Brazil

Celia Silva, MD PhD From the Cardiovascular Division of São Paulo Federal University - Brazil

Cristina Mangia, MD PhD From the Cardiovascular Division of São Paulo Federal University - Brazil

ABSTRACT

A 31-year-old, white female, was referred with symptoms of tiredness, lower limbs edema and gain15 Kg. The clinic examination showed diastolic murmurinthe tricuspid area, liver palpable2-cm below thecostal marginand important edemain the lower limbs. Electrocardiography revealed no ST–T wave changes. Echocardiographic study showed a tumor mass, originating from the free wall of right ventricle (RV), with partially obstructingthe inlet ventricularcavity and tricuspid valve. Computed tomography of the chest showed a 5-cm mass, originating from theRV free wall, withprojectiontothe ventricular cavity. The tumor was successfully resected, this was an encapsulated tumor, totally resected without the need to open the ventricular cavity, decompressed ventricular cavity and releasing the mobility of the tricuspid valve. The histopathology features were consistent with paraganglioma. The patient had an uncomplicated recovery and after 20 months of follow-up, was asymptomatic and reintegrated into their routine activities.

KEYWORDS Tiredness, computed tomography, heart ventricle/pathology/surgery, heart neoplasms/diagnosis, paraganglioma/diagnosis/surgery

Case Reports

Material and Methods.A 31-year-old, white female was admitted, in January 2012, to a local hospital forin labor, being borna healthy child3,300 grs. In August 2012, 7 months postpartum period, the patient returnedwith symptoms of tiredness, lower limbs edema and gain15 Kg.

On clinicalexamination revealed: diastolic murmurinthe tricuspid area, liver palpable2.0cm below thecostal marginand edemain the lower limbs. Electrocardiography revealed no ST-T wave changes.Chest radiography unchanged.

Echocardiographic study showed a 5.0 x 4.8-cm tumor mass, originating in the free wall of right ventricle with partially obstructingthe inlet ventricularcavity anddeforming thetricuspid valve.(Figure 1)

Contrasted Computed tomography of the chest, showed a 5.0-cm mass in the free wall of the right ventricle, protruding into the inflow tract of the right ventricular cavity and affecting opening of the tricuspid valve. (Figure 2-3).

Surgical resection of the tumor was performed at August 28, 2012, recommended for definitive tissue diagnosis because the possibility of malignancy could not be excluded and to alive the inlet RV obstruction. (Figure 4) The surgical approach was performed, by external face (epicardial) of the tumor, closed to AV junction. Through a cross insição to the major axis of the RV, parallel al AV groove and 2cm below the bed of the right coronary artery was opened the fibrous capsule of the tumor was opened. The electrocautery was employing, due tointenseirrigation source, leavingthemusclesownfree wallRV. The tumor mass measuring5.0x4.8-cm,wasenucleated in its entirety, without thecoverageofthe right ventricular cavity. (Figure 5)

Care was takenin dissection, near theAVgroove,to prevent the injuryof the right coronaryartery.

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Results. The postoperative course was uneventful with discharge on the fourth postoperative day. Histology of the tumor confirmed the diagnosis of paraganglioma without capsular invasion. (Figure 6)

After 6th month of follow-up a Computadorized Tomography Angiography image, was performed and showed that theareaoftumor resectionbelow theAVgroove,without compromisingthe right coronary artery. (Figure 8 A)After contrast injection, We can observed the right ventricle and tricuspid valve free of obstruction. (Figure 8 B).

Comments

Cardiac paraganglioma is particularly rare, with ≈85 reported cases ranging from 8 to 79 years of age.[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. About 18% of paragangliomas are found outside of the adrenal glands: common locations for such tumors include the periaortic mediastinum and the retroperitoneum. [3], [4] Mediastinal paragangliomas can be either intracardiac or extracardiac. Intracardiacparagangliomas have been found mostly in the left atrium and less commonly in the interatrial septum, left ventricle, anterior surface of the heart, and right ventricular outflow tract. Only 7 cases of right atrial paraganglioma have been described in the world medical literature. [5], [6], [7], [8], [9], [10], [11]

Clinical presentation is variable, for it depends upon the location of the tumor in relation to cardiac structures. Intracardiac tumors may present with symptoms of valvular obstruction, dyspnea, or syncope.

In addition, presenting symptoms may be attributable to adrenaline secreted by the tumor. [5], [6], [7] Tumors that involve the coronary artery ostium can present as an acute coronary syndrome, whereas tumors that compress the great vessels can present with features of heart failure. [8], [9], [10]There have been few reports of asymptomatic presentation.

At present, paragangliomas are diagnosed mostly with the aid of noninvasive imaging, including computed tomography, magnetic resonance imaging,

Management is by open-heart surgical resection and can be complex, depending upon the size and location of the tumor. Of the cases reported heretofore in the world literature 4 out of 6 had successful surgical outcomes. [6], [7], [8], [9]

Our patient is in 20th surgical outcome with successful, and his tumor was probably nonsecreting.

In our patient, the histologic diagnosis was performed after tumor resection. This case affords an unusual symptomatic presentation of right ventricularparaganglioma as,obstructiveclinicalright heart chambers, followed by successful surgical resection.

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. After 20 months of follow-up, the patient is regained their usual activities.

Figure - Legend

Figure 1: Preoperative Echocardiographic study. Tumor of the right ventricle free wall, obstructing the right atrium (RA) and right ventricle (RV) cavity (blue arrows) and deforming the tricuspid valve (TV) (green arrow). LA: left atrium; LV: left ventricle.

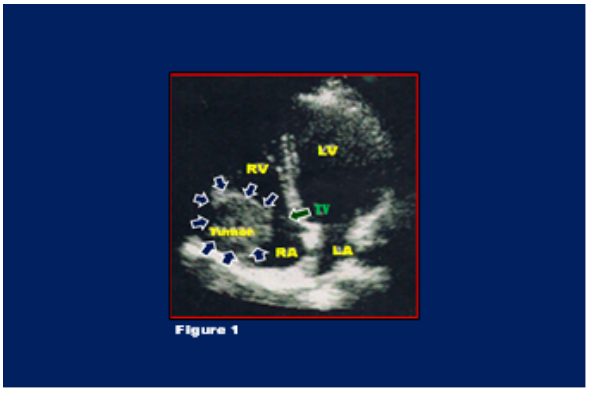


Figure 2: Preoperative Computed Tomography Angiography. Tumor of the right ventricle (RV), localizedbelow the Atrioventricular (A-V) junction (arrows)

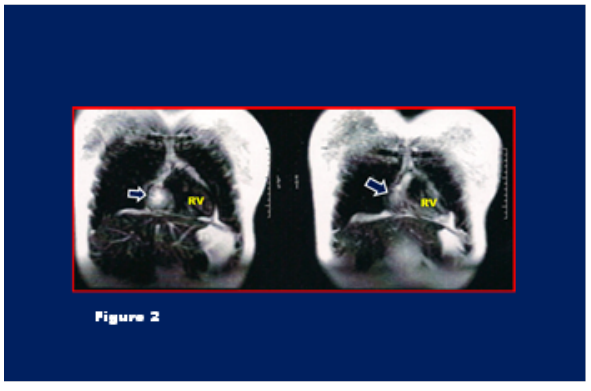


Figure3: Preoperative computed tomography angiography. Tumor of right ventricle (RV), located at Atrioventricular (A-V) junction, obstructing theright atrium (RA) and right ventricle (RV) cavity (blue arrows) anddeforming thetricuspid valve (green arrows).

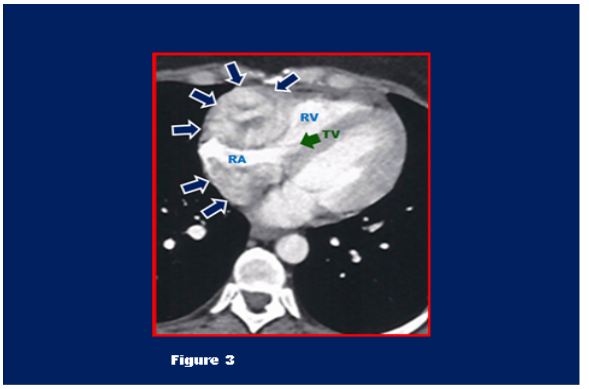


Figure 4: Intraoperative photography of Paraganglioma of theright ventricle (RV) (arrows). RA: right atrium.

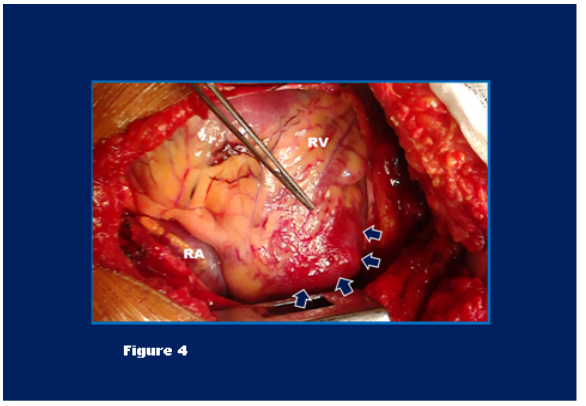


Figure 5: Intraoperative photographyof surgicalarea of tumor resection of the right ventricle (RV)(arrow). Procedure performed without opening the right cavities. RA: right atrium.

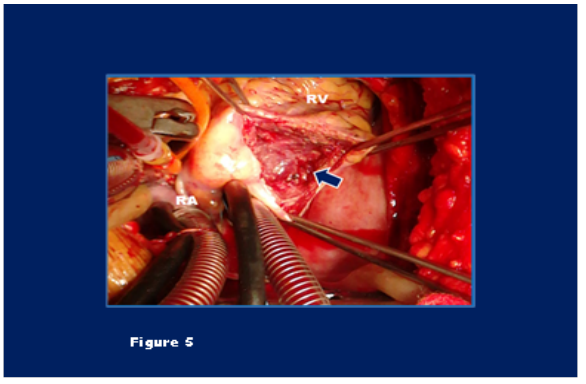


Figure 6: Photography of the Paraganglioma tumor. Measurements: 5.0 x 4.8cm.The bisected mass is well circumscribed and appears encapsulated and multiple dilated vascular channels in the cut surface.

Figure 7: Hematoxylin and Eosin stain. The tumor is composed of nests of cells bordered by slit-like vascular channels. The cells have a modest amount of eosinophilic cytoplasm and round to oval, bland nuclei. Occasional cells had larger nuclei.

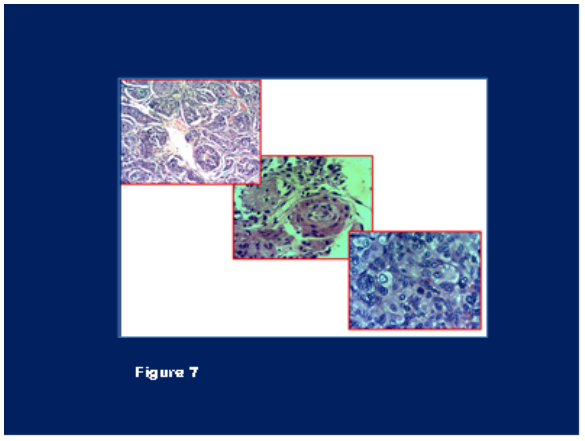
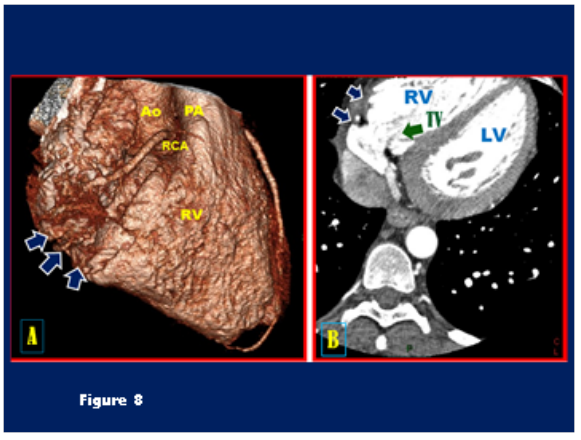


Figure 8: Postoperative Computed Tomography Angiography image, showed: A- Tumor resection areaof the right ventricle (RV), below theA-Vgroove,(blue arrows) preserving integrity of the right coronary artery (RCA). B - After contrast injection, observed the right ventricle (RV) and tricuspid valve (TV), free of obstruction. (green arrow). Ao: aorta; LV: left ventricle, PA: pulmonary artery.



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