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A rare case of unexpected cardiac incidentaloma causing syncope

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A 68-year-old woman presented with recurrent paroxistic tachycardia followed by syncope occurring in right lateral decubitus.

A transthoracic echocardiogram showed a cardiac mass close to the right atrium (*Figure 1, yellow arrows*) and to the inferior vena cava, apparently not interfering with diastolic filling.

A transesophageal study showed a right para-atrial mass close to the atrioventricular groove but not infiltrating the cardiac wall (*Figure 2, Panel A, yellow arrows*) with regular surface and solid texture.

A contrast enhancement computed tomography scan confirmed the anatomical details (*Figure 2, Panel B, yellow arrows*) and the relationship with the right coronary artery (*Figure 2, Panel B, red arrows*).

In the hypothesis that the compression of the inferior vena cava was responsible for the syncope in the right lateral decubitus, the patient underwent a surgical intervention without cardiopulmonary bypass.

Intraoperatively, high doses of alpha and beta blockers were used to manage hypertensive crisis up to 220/110 mmHg.

The tumour was successfully removed and appeared as a nodular reddish, encapsulated, 7,5 x 5 cm mass (*Figure 3, right*); the histological examination documented a prominent vascular network separating nests of neuroendocrine cells (Synaptophysin +) (*Figure 4, left*) and a peripheral sustentacular cell population (S100 +) (*Figure 4, right*), compatible with paraganglioma. Postoperative urine and serum cathecolamines and

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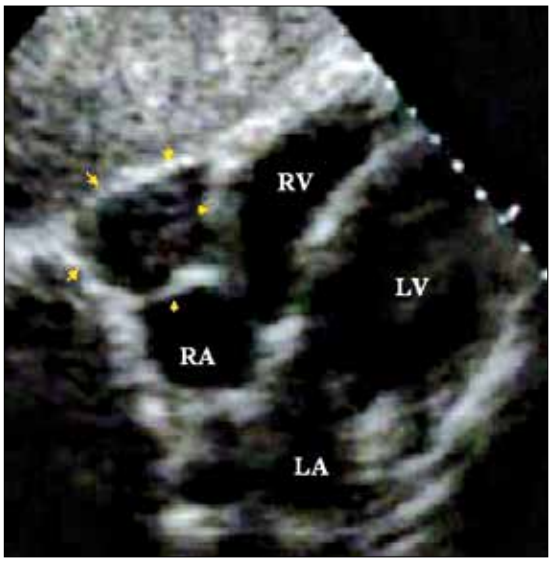

Figure 1 - *A standard transthoracic echocardiogram from the subcostal approach shows a cardiac mass close to the right atrium (yellow arrows).*

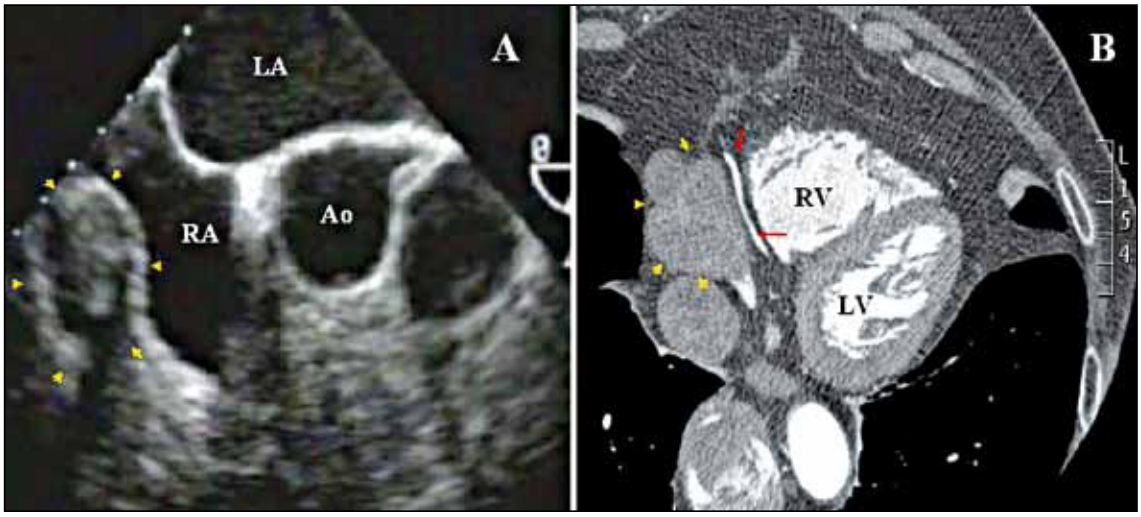

Figure 2 - *A transesophageal echocardiogram reveals a right para-cardiac mass apparently not infiltrating the cardiac wall (Panel A, yellow arrows); the computed tomography scan confirms the anatomical position of the mass (Panel B, yellow arrows) and its spatial relationship with the right coronary artery (Panel B, red arrows).*

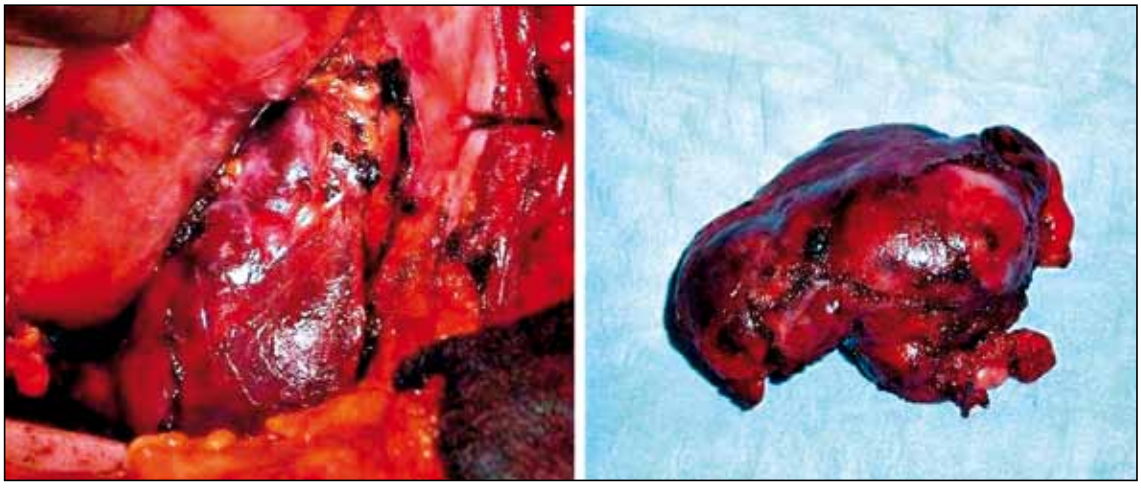

Figure 3 - *Surgical inspection of the para-cardiac mass "in situ" (left) and after resection (right).*

metanephrines were in normal range. The patient was discharged in 6th postoperative day in a good clinical condition.

Paragangliomas represent 18% of total adrenal tumours (1-3); they arise from paraganglia (small adrenal accessory organs embryologically deriving from neural crest migration) and usually are located in the paraaortic and para-caval sympathetic chains within the organs of Zuckerkandl, the urinary bladder and the sympathetic chain of the neck or mediastinum.

They can cause clinical symptoms as a result of cathecolamine release in the bloodstream as well as for compressive syndromes.

Cardiac paragangliomas are commonly located in the left atrium and less frequently in the right atrium, in the atrial septum

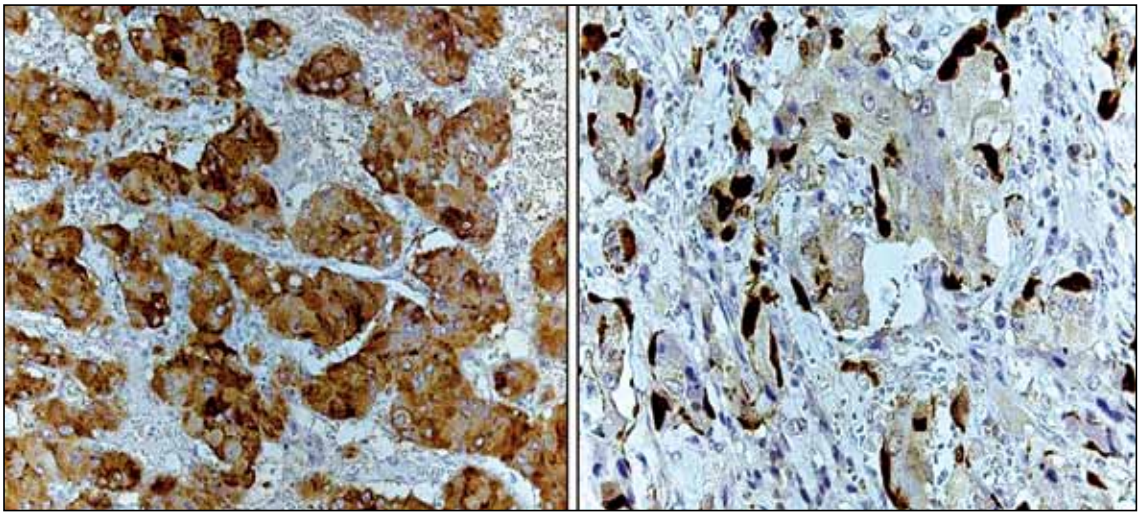

Figure 4 - *The histological examination confirms the diagnosis of paraganglioma showing a prominent vascular network with nests of neuroendocrine cells (Synaptophysin +) (left) and a peripheral sustentacular cell population (S100 +) (right).*

and in the left ventricle. Diagnostic tools include serum and urinary catecholamines and metanephrines, computed tomography, magnetic resonance imaging, iodine-131-meta iodobenzylguanidine scanning and PET.

We described a patient with atypical symptoms and detected the mass as an incidental finding (incidentaloma) at echocardiography.

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