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Multimodality imaging of aortopulmonary paraganglioma

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A 72-years old woman was admitted to our department for a long lasting (3-hours) thoracic pain episode and increasing palpitations during the last month. At admission clinical examination was unremarkable and laboratory test showed slightly increased urinary as well as plasma levels of catecholamines. Transthoracic echo (Fig. 1, panel A-B) showed a vascularized, hypo-echoic mass with a central hyperechoic region that did not appear to interfere with flow (Online Video 1) in the great vessels. Subsequent CT scan confirmed the presence of a 6 × 7 cm mass with a central necrotic core, located under the aortic arch, over the pulmonary trunk (Fig. 1, panel C-D) and vascularized by thyroid arteries. Total body iodine-131 meta-iodobenzylguanidine scintillation scan showed tracer accumulation in the anterior mediastinum (Fig. 1, panel E), which suggested the presence of chromaffin hyperfunctionating cells, thus confirming the diagnosis of aortopulmonary paraganglioma.

Paraganglioma are extremely rare neoplasm of the neural crest, originating from either parasympathetic or sympathetic paraganglia. Only 10% of sympathetic paragangliomas are located in the thorax and are usually associated with catecholamine excess, presenting with hypertensive crisis, headache, sweating and palpitations [1]. However, some may be pauci-symptomatic and the diagnosis may be delayed until compressive symptoms occur [2]. High index of suspicion for malign forms should be maintained in patient with RET, VHL, SDHD and SDHB mutations [3]. Complete surgical excision with adequate pre-

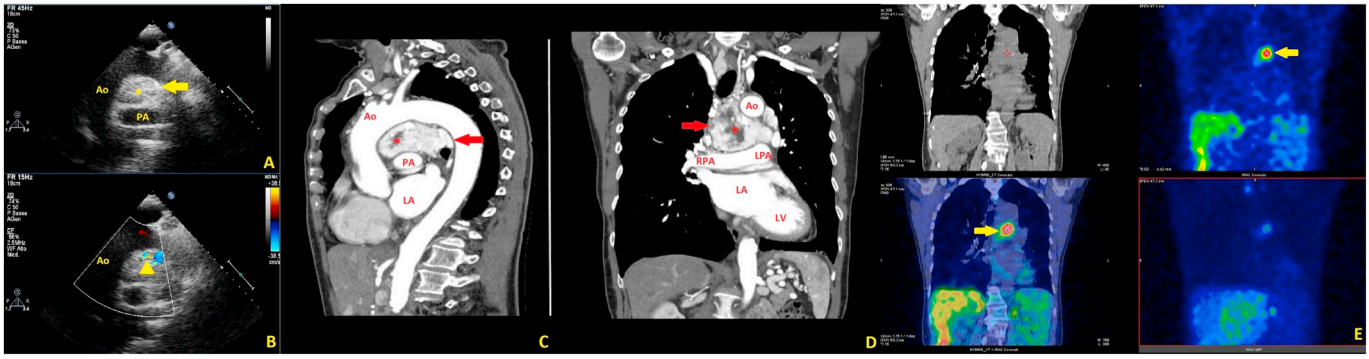

Fig. 1. Transthoracic echo (suprasternal window) showing a hypoechoic mass (yellow arrow) with a central hyperehoic region (yellow asterisk) (A), with abundant vascularization evident with color-doppler (yellow triangle) (B). CT scan showing a contrast-enhanced mass (red arrow) with a central necrotic core (red asterisk), sagittal plane (C), frantal plane (D). Total body iodine-131 meta-iodobenzylguanidine scintillation scan with tracer accumulation in the anterior mediastinum (yellow arrow) (E). Ao = aorta; PA = pulmonary artery; LA = left atrium; LV = left ventricle; RPA = right pulmonary artery; LPA = left pulmonary artery. (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)

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operative alfa and beta-blockade is the main stone of the therapy [1].

Although rare, catecholamine secreting tumours should always be researched in the diagnostic work-up of patients with hypertension, palpitation or chest pain, as in this case, remembering that some may be located in the mediastinum which can be initially explored by echocardiography from suprasternal view.

Supplementary data to this article can be found online at https://doi.org/10.1016/j.ejim.2019.05.005.

References

[1] Ramlawi B, David EA, Kim MP, Garcia-Morales LJ, Blackmon SH, Rice D, et al. Contemporary surgical management of cardiac paragangliomas. Ann. Thorac. Surg. 2012;93:1972.

[2] Erickson D, Kudva YC, Ebersold MJ, Thompson GB, Grant CS, van Heerden JA, et al. Benign paragangliomas: clinical presentation and treatment outcomes in 236 patients. J. Clin. Endocrinol. Metab. 2001;86:5210.

[3] Boedeker CC, Neumann HP, Maier W, Bausch B, Schipper J, Ridder GJ, et al. Malignant head and neck paragangliomas in SDHB mutation carriers. Otolaryngol. Head Neck Surg. 2007;137:126.