A primary cardiac paraganglioma

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A 32-year-old woman presented with a 2-year history of intermittent hypertension. Clinical examination was unremarkable as well as routine biochemical blood examinations. Urinary as well as plasma levels of catecholamines and their metabolites were within normal limits. Magnetic resonance imaging angiography of renal arteries did not demonstrate any stenosis. The patient subsequently underwent transthoracic echocardiogram which demonstrated a 4.3 × 3.5 cm intracardiac mass protruding within the left atrium (LA) and was interpreted as myxoma. A transoesophageal echocardiogram confirmed the presence of a mass of moderate echogenicity enveloped by an outer and rather thick membrane, located on the lateral wall of LA (Figure 1A; LA, left atrium; Ao, Aorta; Tu, tumour). Further imaging with chest-computed tomography verified echocardiographic diagnosis (Figure 1B, arrow; Tu, tumour). A CT scan of the abdomen, pelvis, and adrenal glands did not provide any pathologic finding. Total body iodine-131 meta-iodobenzylguanidine scintillation scan showed no focal uptake.

Using full cardiopulmonary bypass, the left and right atrium were explored but surprisingly no tumour was found. The tumour was found to be located outside LA, on the junction of left superior pulmonary vein (LSPV) and the LA. The tumour was completely excised en bloc with part of the LSPV wall, and was then reconstructed with a pericardial patch (Figure 1C and D; LAA, left atrial appendage; Tu, tumour). Pathological examination of the mass with the use of immunohistochemistry demonstrated cells with granulated eosinophil cytoplasm, which were also positive for synaptophysin, NSE (NSE = neuron specific enolase) (Figure 1E), and chromogranin staining (Figure 1F), and ‘Sustentacular cells’ positive for S-100 protein staining. These findings were consistent with primary cardiac paraganglioma, which is a very rare cause of secondary hypertension. Since then, the patient has remained free from arterial hypertension, in 3 years follow-up.

Paragangliomas are neoplasms of neural crest origin, arising from extra-adrenal chromaffin tissue (thus known as extra-adrenal pheochromocytomas), and have been classified into brachiomeric, intravagal, aortico-sympathetic, and visceral-autonomic groups. According to autopsy findings, paragangliomas have an incidence of 0.1–0.3%, and only 1–2% of these cases have a thoracic location. Cardiac paragangliomas account for <5% of all cardiac tumours and they arise from cells of visceral paraganglia in the LA or the aorta. Paragangliomas are usually benign tumours, and their clinical presentation depends on symptoms caused by local invasion, and/or excessive production of catecholamines. They are found in 0.05–0.2% of hypertensive patients. Metastases can also occur in up to 10% of cases. The mainstay of treatment is complete surgical excision which is sometimes technically demanding. Local recurrence has been described.