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Cardiac Paraganglioma : Diagnostic Work Up and Review of the Literature

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Abstract. Paraganglioma of the heart are potentially invasive, highly vascularized tumors for which complete resection may be curative. Derived from the cardiac wall in most instances, resectability can be assessed after integration of the data provided by MRI in T2 sequence, and coronarography. A fully documented case of a large cardiac pheochromocytoma of the left atrium and AV groove is reported and the pertinent literature on the subject is here presented.

Introduction

Pheochromocytoma and paraganglioma are rare, catecholamine secretory tumours that account for less than 1% of all cases of diastolic hypertension. Catecholamine secretory tumors originates from the adrenal medula (pheochromocytoma) or from extra-adrenal neural crest derivatives (paraganglioma) such as the sympathetic ganglia. Their neuroectodermic origin and the extensive distribution of neural crest derivatives explain the diverse localization of paraganglioma. They may be localized at the base of the skull, the pericardium, the atria, the periaortic areas (organ of Zuckerkandl) or to more remote locations such as the urinary bladder, the ovaries, testes, or prostate (1).

Approximately 1% of these tumours are seen in the chest. Half of them are reported to be in the posterior mediastinum. The diagnosis is obtained from the integration of preoperative diagnostic work-up, but the resectability depends on localization and invasiveness (1, 2).

In this case report we present a cardiac papraganglioma which is a rare entity consitituting less than 5% of all cardiac tumours. We emphasize the usefulness of the different preoperative diagnostic tools and the surgical management. An actualised review of the literature is provided.

Clinical observation

A 68 years old caucasian male was admitted to our hospital for exploration of a large (8 × 5 cm) tumour of the mediastinum discovered on CT-scan imaging. The patient had no previous medical history of hypertension, headache, hyper-hydrosis or dysmetabolic syndrome. He complained of arrhythmias which were described as isolated ectopics rather than runs or bursts of tachycardia. He had been investigated 12 years before and the para - cardiac lesion (at that time, a nodular mass of 1 cm in size) was diagnosed as a varicosity of the left lower pulmonary vein. In this normotensive adult, the enhanced contrast CT-scan disclosed a voluminous hyper-vascularized tumour abutting the left atrium and AV groove. It was not clearly visible whether the mass was within or extrinsic to the myocardium. Transoesophageal echocardiography revealed a bulging of the lateral wall of the left atrium, with deformation of the postero-lateral mitral annulus leading to a grade 1-2/4 mitral regurgitation. There was no left ventricular hypertrophy. Analysis of 24 h urinary collection revealed increased level of catecholamines (norepinephrine ; 690 mg/24 h (normal value : 10-110), methanephrines ; 2847 mg/24 h) and metabolites (vanillyl mandelic acid (9.3 mg/24 h ; NV 1-6)) with normal level of blood catecholamines. I-131 MIBG scan evidenced a single left para-cardiac focus of hyperfixation. 111 indium octreotid scan revealed only mild fixation. MRI showed a voluminous T2 hyperintense nodular structure in the posterior atrioventricular groove, located within the pericardial sac with compression of the sinistrolateral wall of the left atrium. A tiny slit of fat was noted at the interface with the left atrium suggesting resectability but no conclusion could be drawn at the level of the left AV groove and its structures (coronary sinus, circumflex artery) (Fig. 1). A coronary angiogram demonstrated a highly vascularized tumor fed by two pedicles derived from the proximal and distal circumflex artery. After preparation of the patient with phenoxybenzamine and b-blockers, surgery was conducted via median sternotomy. Cardio-pulmonary bypass was started with bi-caval canulation and aortic reinjection. A coronary sinus retrograde cardioplegic canula was placed into position. After gently lifting the heart to

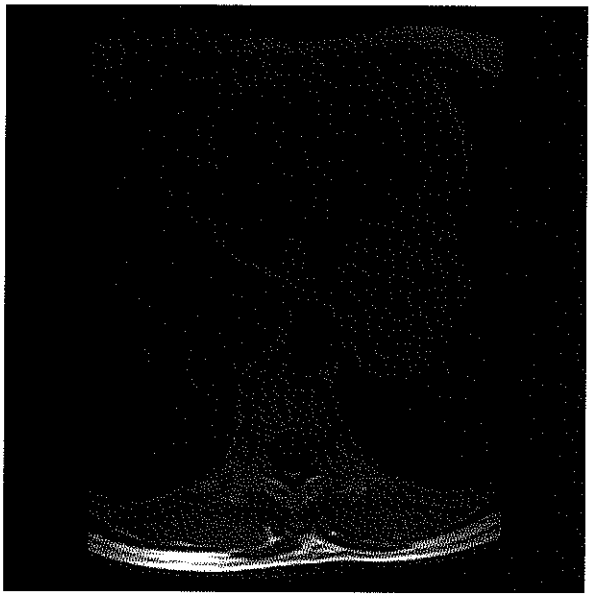

Fig. 1A
Transversal cut in RMN. Evidence of a voluminous tumor in contact with the left atrium and AV groove.

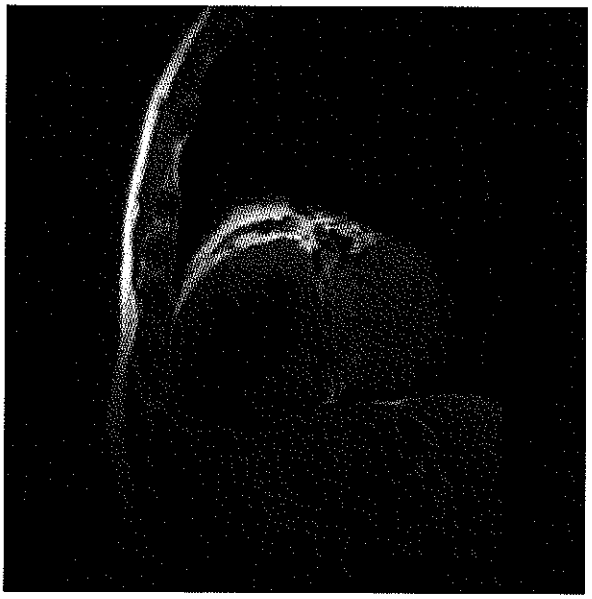

Fig. 1B
The same tumor in parasagittal plane

assess the tumour, it was noted that there was no evidence of hemodynamic instability and resection was initiated on full flow cardiopulmonary bypass without aortic cross-clamping. The mass was firmly adherent to the epicardium and its centre was situated on the lower part of the left atrium and the left AV groove (Fig. 2). To facilitate the delineation between the tumour and surrounding myocardium, we chose to perform the resection on the fully emptied beating heart. Resection with the use of low power electro-cautery was first initiated close to the AV groove to which the tumour was adherent but without invading the coronary sinus. Resection was performed "en bloc" for what appeared to be an encapsulated lesion fed by 2 pedicles, one derived from the distal circumflex and the other from large sinusoids from the left main and proximal circumflex. The patient was easily weaned from bypass and its post-operative course was uneventful, except for a re-hospitalisation for atrial tachyarrhythmia, three weeks later.

The lesion was a 137 g well delineated tumour of 7.5 × 6.5 × 4 cm. Different patterns of neuro-endocrine cells, positive for chromogranin, synaptophysin and CD 56 were recognized (Fig. 3). Neuro-endocrine granules were demonstrated on electronic microscopy. The diagnosis was extraadrenal functional paraganglioma.

Discussion

Cardiac paraganglioma is a rare pathology. Nevertheless, it is more frequently diagnosed nowadays. Recent literature comprises only 15 new cases since the excellent review of JEBARA *et al.* in 1992 (3), which reported over 32 cases. The diagnosis of this tumour is always challenging and should be considered in the presence of a highly vascularized nodular lesion of the atrial wall, interatrial septum or AV groove associated to increased catecholamine turn-over, either symptomatic or asymptomatic. Meta-iodo-benzyl-guanitidine (MIBG) analogue is taken up by chromaffin cells and incorporated into neuro-granules. MIGB scintigraphy can be useful in localizing extra-adrenal paraganglioma and metastases in difficult situations. The extra-medullary tissue is visible in 80-90% of cases on 123 -131 I MIBG scintigraphy. Ten percentof the tumours are unfortunately not seen by this technique (false negative), but false positive images are rare (1, 3). Specificity is nearly 100% in malignant lesions (1). Octreotide scanning can image the tumour in approximately 70% of the cases. A small proportion of patients with negative MIBG scan may be positive with octreotid scan (4, 5). In our patient, only mild fixation was evidenced on octroid 111 Indium scintigraphy.

Our observation confirms that MRI allows via its soft tissue contrast some degree of tissue characterization and appears particularly useful in tumours in close proximity to the heart or to the great vessels. Most paragangliomas give high T2 weighted signals (6) and this can differentiate the tumours from surrounding cardiovascular structures. Inside the tumour, signals are void from flowing blood. The coronary angiogram, which is advocated by JEBARA *et al.* (3), is in our opinion, of utmost interest as