Case report

A cardiac paraganglioma presenting with atypical chest pain

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Abstract

Primary cardiac tumours are rare. The majority are benign and 75% are atrial myxomas. One of the more unusual benign tumours affecting the heart is a cardiac paraganglioma. A 56-year male was presented with a 6-month history of vague, left-sided chest pain, intermittent paraesthesia of the left arm and dyspnoea on bending. Echocardiography documented a large, highly vascular mass, attached to the intra-atrial septum. All investigations, haematological, biochemical, neuroendocrine tumour markers and urinary catecholamine levels, were within normal limits. Macroscopically the tumour involved the whole of the atrial septum, roof of the left atrium and extended to surround the superior vena cava, excluding total resection. The feeding vessels were ligated in the hope of infarcting the remainder of the tumour. In over 2-years of follow-up, the patient remains clinically stable. To our knowledge this is the first reported case where vascular ligation has been used to control a cardiac paraganglioma.

Keywords: Mediastinal paraganglioma; Echocardiography; Angiography; Vascular ligation; Benign

1. Introduction

Primary cardiac tumours are extremely rare. The majority are benign and of these 75% are atrial myxomas. [1] One of the more unusual benign tumours affecting the heart is a cardiac paraganglioma, commonly referred to as a cardiac phaeochromocytoma, irrespective of functional status. We present a case of a cardiac paraganglioma presenting with atypical chest pain.

2. Case report

A 56-year male was presented to the local cardiology department with a 6-month history of vague, left-sided chest pain, intermittent paraesthesia of the left arm and dyspnoea on bending. There was no past medical history of note or identifiable cardiac risk factors, he was on no regular medication and physical examination was unremarkable. Resting 12-lead ECG showed first-degree AV block. Trans-thoracic echocardiography was performed which identified a large intra-cardiac mass adherent to the intra-atrial septum. An echocardiogram 3-years earlier had been reported as 'normal'. He was transferred for further assessment.

Repeat echocardiography with a transoesophageal (TOE) study documented a large 6 × 6 × 4 cm mass attached to the intra-atrial septum, partially filling the right atrium and pushing the atrial septum into the left atrium (Fig. 1a). There was no demonstrable haemodynamic effect on intra-cardiac flow. The mass appeared highly vascular and a CT scan of the thorax was performed (Fig. 1b). This confirmed the vascular nature of the tumour, raising the possibility of a sarcoma. No other intra-thoracic masses were identified.

Cardiac catheterisation demonstrated normal epicardial coronary arteries. Two large feeding vessels were identified, one originating from the right coronary artery, which was the principle vascular supply of the tumour, the other from the proximal circumflex artery (Fig. 1c). All haematological and biochemical investigations were within normal limits including neuroendocrine tumour markers and urinary catecholamine levels.

A tissue diagnosis was important and the extensive vascularity of the tumour would have made percutaneous or transoesophageal biopsy very hazardous. Surgery with the aim of total resection, plus cardiac reconstructive surgery, was therefore performed via a median sternotomy. The patient was placed on cardiopulmonary bypass, the aorta cross-clamped and the heart arrested with cold blood cardioplegia. Macroscopically the tumour involved the whole of the atrial septum, the roof of the left atrium and extended to surround the superior vena cava, which excluded total resection. A large biopsy (30 × 18 × 20 mm) was taken and after closure of the right atrium the cross-clamp was removed. The feeding vessels were ligated using colour doppler on TOE to confirm successful interruption of blood flow to the tumour. This was performed in the hope of infarcting the remainder of the tumour.
Histopathological examination of the resected mass showed a highly vascular tumour with a dense sclerosed core. There were extensive thin walled channels around which there were nests of pale cells with round nuclei and prominent pale pink cytoplasm. The nuclei were pleomorphic and there was no mitotic activity (Fig. 2).

The tumour cells were immunohistochemically positive for the neuroendocrine marker chromogranin, plus PGP9.5, synaptophysin and gamma-enolase. S100 showed strong nuclear and weaker cytoplasmic staining with sustentacular cells staining strongly. The tumour cells were also strongly positive for CD56 and negative for cytokeratin. The diagnosis was a cardiac paraganglioma.

In over 2-years of follow-up, the patient's clinical condition remains stable with no change in tumour size, but tumour vascularity has markedly reduced. Evidence of metastatic spread was excluded with MIBG and whole body CT scanning. No further treatment has been given.

3. Discussion

Cardiac paragangliomas are rare extra-adrenal tumours derived from the neural crest, associated with the autonomic nervous system, and account for 0.3% of all mediastinal neoplasms [2]. They can occur at other extra-adrenal sites including the carotid body and glomus jugulare of the middle ear or as part of Carney’s triad [2]. Presentation is in adulthood with a female predominance and less than 50% are functional, as in this patient [2–4]. The tumour can arise from several sites including the intra-atrial septum, left and right atrium, intra-pericardial aorta, pulmonary artery and the left ventricle [3,5–7].

The patient may present with constitutional symptoms including malaise, weight loss and fever. A functional, catecholamine-secreting tumour can give rise to paroxysmal hypertension, sweating, palpitations and headaches [2]. Clinical presentation also relates to tumour site [1,8]. Intracardiac blood flow can be compromised either directly by the tumour mass or through disruption of valvular function. In our patient, there is an evidence of superior vena cava involvement with the associated risks of the development of superior vena cava obstruction. These tumours are locally invasive and can produce symptoms secondary to pericardial involvement or through invasion of the conduction system. Embolic phenomenon may also occur. In some the tumour is an incidental finding [1,4].
There are no proven histological criteria to differentiate benign from malignant cardiac paragangliomas, and there is no direct correlation between tumour size and malignant potential [2,3,9]. As such the incidence of malignancy varies depending on definition used [7]. Paragangliomas are classically benign, slow growing and locally invasive. They are highly vascular tumours that ‘parasitise’ the coronary circulation [1]. The principle feeding vessels typically originate from the left coronary system although in this patient a large collateral from the right coronary artery principally supplied the tumour [7].

Complete surgical resection, where possible, is the treatment of choice [4]. Prior to surgery, tumour extent needs to be visualised. Echocardiography will define tumour size and site whilst coronary angiography will demonstrate coronary artery involvement [5]. Because paragangliomas are highly vascular, complete resection can be problematic and should be performed on cardiopulmonary bypass. In cases where the tumour is functional the anaesthetic management is similar to an adrenal phaeochromocytoma with combined alpha and beta-blockade [2,5,6,8]. Total cardiopulmonary bypass with cardioplegic arrest to isolate the heart from the systemic circulation before manipulation is advisable. Unlike adrenal phaeochromocytomas, cardiac paragangliomas have a less well-defined capsule, making resection more difficult. As the tumours are of locally invasive nature, disease-free excision margins must be achieved. Extensive cardiac reconstructive surgery may therefore be required, with its associated risks [6]. When total surgical resection is not possible orthotopic cardiac transplantation is advocated [10].

As cardiac paragangliomas are rare there is no consensus on long-term management. The use of chemotherapy and radiotherapy is of limited value [1]. If complete tumour excision is possible, long-term prognosis is good; what is unclear is long-term outcome in patients such as ours where the tumour is unresectable.

To our knowledge this is the first reported case where vascular ligation has been used to control a cardiac paraganglioma.

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References