Right atrial paraganglioma: an unusual primary cardiac tumour

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Received 16 January 2008; received in revised form 21 February 2008; accepted 24 February 2008; Available online 10 April 2008

Abstract

We present a case of an unusual presentation of a rare primary cardiac tumour. There are no more than 50 previously reported cases of primary cardiac paraganglioma in the world literature and only a small proportion of these involve the right side of the heart. Diagnosis is difficult and surgical management is varied and complex.

Keywords: Cardiac; Cardiac tumour; Paraganglioma

1. Case report

A 51-year-old male presented to a regional hospital complaining of chest pain and palpitations. This was of acute onset; there were no similar previous episodes and no cardiovascular history of note. On examination he was normotensive and was discovered to have an irregular heart rate, confirmed as atrial fibrillation on electrocardiogram. Otherwise his cardiovascular exam was normal and cardiac enzymes revealed no abnormality. Of note, his brother had been diagnosed with thoracic paraganglioma 12 years previously. Urine was analysed for metanephrines and vanillyl mandelic acid (VMA) and returned within normal limits. A transthoracic echocardiogram and subsequent transoesophageal echocardiogram showed a 3.5 cm broad based mass in the right atrium involving the inter-atrial septum. CT thorax and abdomen were performed as staging scans and showed no other abnormality. He was transferred to a cardiothoracic unit for further management.

A coronary angiogram identified a well vascularised mass in the right atrium receiving branches from the left circumflex and right coronary arteries (Fig. 1). The coronary arteries were otherwise normal.

An exploratory sternotomy was performed and the patient was placed on cardiopulmonary bypass. A 6 cm tumour was identified originating in the right atrium, invading the superior vena cava proximally, the right main pulmonary artery and right superior pulmonary vein. A frozen section was sent from the mass and returned a provisional result of paraganglioma.

Despite attempts to mobilise the tumour, the extensive invasion of vascular structures rendered the mass unresectable and the chest was closed.

Over the course of the next 36 h, the patient developed accelerated affects of tumour progression as manifested by SVC obstruction, acute respiratory distress syndrome (ARDS) and subsequent multiorgan failure leading to his eventual demise.

Permanent sections of the tumour showed the characteristic packeted appearance of paraganglioma originally described in German as ‘zellballen’. There was also significant cytologic atypia including large bizarre cells that are a recognised feature of endocrine tumours. Immunohistochemical staining was positive for the endocrine markers chromogranin and synaptophysin. S100 outlined the characteristic sustentacular cell network (Fig. 2).

2. Discussion

Paraganglioma is a tumour of the autonomic nervous system. It is of neural crest cell origin most frequently occurring within the adrenal gland where it is known as phaeochromocytoma. Approximately 18% of paraganglioma are found outside the adrenal gland [1]. Primary cardiac paragangliomas are extremely rare with less than 50 cases reported in the literature [2]. Most of these tumours are found in the left atrium while other less frequent sites include the inter-atrial septum, anterior surface of the heart and left ventricle.
Malignancy is determined by tumour behaviour rather than histological examination with the distinguishing features being distant spread or local tissue invasion.

The most common symptoms of cardiac paraganglioma are those of catecholamine excess, namely, headache, sweating, palpitations and dyspnoea. Clinical suspicion is raised in cases of resistant hypertension particularly in patients with a history of phaeochromocytoma. However, more unusual presentations have been reported such as acute myocardial infarction and stroke [3], congestive heart failure [4], hypertensive crisis in labour [5] and upper limb paraesthesia [6]. A family history of phaeochromocytoma may indicate MEN2, Von Hippel—Lindau syndrome, neurofibromatosis type 1 and familial phaeochromocytoma—paraganglioma syndrome [7], the latter being the most likely in this case as no features of the other syndromes were present. Familial phaeochromocytoma—paraganglioma syndrome is a group of syndromes caused by germline mutations of the genes SDHB, SDHC and SDHD, although patients with apparently sporadic phaeochromocytomas may also carry some mutations [8]. Location in the heart is not particularly associated with familial syndromes. The family of this man have been referred for genetic counselling.

There are only four previous cases of primary paraganglioma arising from the right atrium in the world literature [2,3,9]. All of these cases involved functional tumours with three presenting with persistent hypertension preoperatively and one case demonstrating intraoperative hypertensive crisis on palpation of the tumour [10]. Our case is unique in that there were no biochemical abnormalities and none of the classic signs of catecholamine excess. The only specific sign (atrial fibrillation) was most likely due to direct invasion of the conduction system of the heart.

3. Imaging

Echocardiography and coronary angiography are useful in identifying cardiac paraganglioma. Consistent findings are of a highly vascular, broad based tumour frequently ‘parasitising’ one or both of the coronary arteries (Fig. 1). Iodine 131-labelled meta-iodobenzylguanidine (I-MIBG) scintigraphy has been used for diagnosis especially in cases of resistant hypertension where a phaeochromocytoma is suspected. As a diagnostic tool, CT appears to be of more benefit in retroperitoneal [2] but either CT or MRI is necessary for preoperative staging.

4. Management

Surgical resection and reconstruction with pericardial patch or synthetic graft is the gold standard treatment for primary cardiac tumours. This approach, however, can be rendered impossible due to extensive involvement of the great vessels. Cardiac autotransplantation has been described with most success reported with left sided sarcomas [10]. There is no experience in the worldwide literature regarding this technique with locally invasive right atrial tumours.

5. Conclusion

Cardiac paraganglioma is an extremely rare primary tumour of the heart. The patient typically presents with symptoms of catecholamine excess. The usual investigations for phaeochromocytoma are necessary, however the most useful, specific to primary cardiac tumours, are coronary angiography and echocardiography. While surgical excision is the standard management, this can be technically difficult and is complicated by the position of the tumour and involvement of large vascular structures.

References

