A CARDIAC PARAGANGLIOMA

Image in cardiology

M. Makotoko*, R.A. Plaatjies#, M. Swart+ and J.H.A. Venter+

* Medi-Clinic Heart Hospital, Pretoria, South Africa
# Pelonomi Private Hospital, Bloemfontein, South Africa
+ Bloemfontein Medi-Clinic, Bloemfontein, South Africa

Address for correspondence:
M. Makotoko
Medi-Clinic Heart Hospital
551 Park Street
Pretoria
0083
South Africa

Email:
makoali@vodamail.co.za

A 31-year-old man presented with a month's history of severe headache, palpitations and excessive sweating. He had no prior illnesses and was not taking any medication. He was a non-smoker and a teetotaller. His parents had both died but he did not know the cause of their deaths.

Clinical examination revealed an ill-looking young man, who was febrile with a temperature of 38.7°C, tachycardia of 109/min, BP of 180/116mmHg. His apex beat was thrusting but not displaced. His heart sounds were normal and he had a grade 3/6 ejection systolic murmur localised to the aortic area. He had signs of lobar consolidation in his right upper lobe. Abdominal examination was normal.

His echocardiogram showed a normal sized left ventricle with no hypertrophy. The left ventricle ejection fraction was 71%. There was a round mass situated in the right atrio-ventricular groove within the pericardial space. Its diameter was 3.7cm.

Chest x-ray showed a normal cardiothoracic ratio and cardiac silhouette. There was a lobar consolidation of his right upper lobe.

A 64-slice CT of the chest confirmed the presence of the tumour in the right atrio-ventricular groove below the right coronary artery. Random glucose was elevated at 16.4mmol/l.

The clinical combination of severe hypertension, diabetes mellitus and a tumour led to the differential diagnosis of a phaeochromocytoma. A 24-hour urine collection for metanephrine and nor-metanephrine excretion was done. The nor-metanephrine excretion was markedly raised at 17 677nmol/24hhrs (n: 24 to 3 418), chromogranin A was 89u/l (n: 0 to 23). The metanephrine excretion was within normal limits: 482nmol/l (n: 160 to 2 478nmol/l). Phaeochromocytoma or, since the tumour is outside the adrenal medulla, a paraganglioma), was therefore confirmed.

The patient was put on NovoMix insulin for his diabetes mellitus and Coversyl plus, one tablet daily, Adalat XL 30mg daily, and Doxazocin 4mg daily for his hypertension. His pneumonia was also treated. He was discharged home but booked to undergo surgical resection of the tumour after two weeks. The anatomical correlations that were seen on the pre-operative images of the echocardiogram and multi-slice CT were confirmed at operation. A branch of the conus artery was supplying the tumour and had to be sacrificed. The histology of the tumour was of a paraganglioma.

Post-operatively the patient's blood pressure and glucose went back to normal and he never needed any further treatment. His recovery was uneventful. He attends annual examinations.

Our patient presents with a very rare cause of hypertension. Cotesta et al.,[1] state that phaeochromocytoma is a cause of hypertension in only 0.1% of hypertensive people. Cardiac paragangliomas are even rarer, only 37 have been reported in the medical literature in adult patients.[2]

Paragangliomas are tumours that arise from neuroectodermal chromaffin cells found outside the adrenal medulla. Similar tumours within the adrenal medulla are called phaeochromocytomas. 10% of phaeochromocytomas are malignant, 10% are extra-adrenal and 10% are familial.[1] Phaeochromocytomas arise as isolated tumours or as part of the Multiple Endocrine Neoplasia syndrome (MEN 2), where, in addition there is medullary thyroid carcinoma (MTC) and hyperparathyroidism. In MEN 2B there is phaeochromocytoma, medullary thyroid carcinoma and mucosal neuromas on the lips and tongue. Phaeochromocytoma can also occur in conjunction with the Von Lippel-Lindau disease, cerebellar or retinal angiomas.[3]

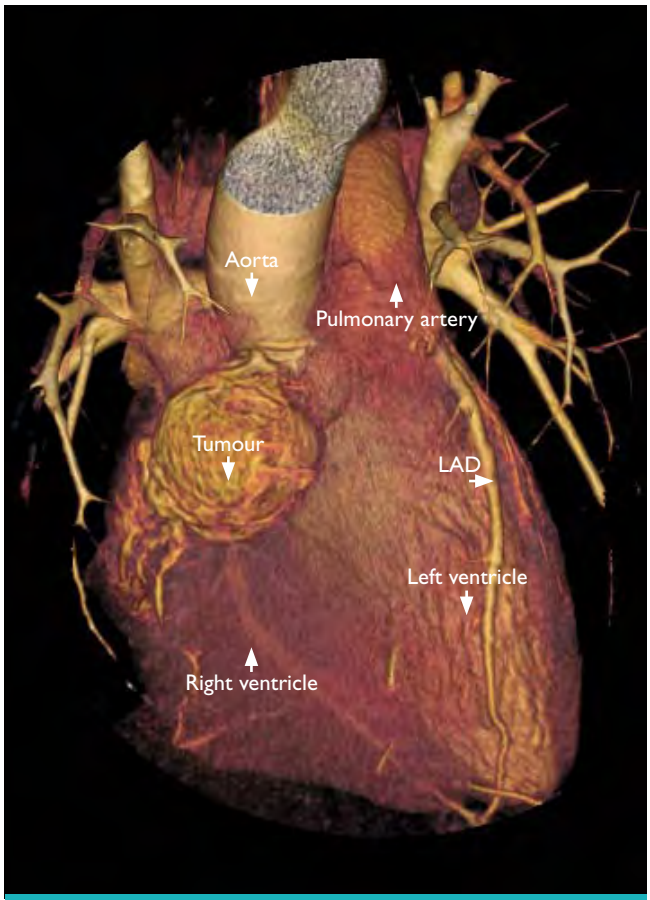

FIGURE 1: A 64 slice cardiac CT was obtained. In this 3-dimensional reconstruction a mass is demonstrated in the AV groove, in close association with the proximal right coronary artery.

The majority of patients with phaeochromocytoma present with sustained, as opposed to paroxysmal hypertension. However, 50% of these patients will still get paroxysms of severe hypertension associated with severe headache, palpitations and angina.

Extra-adrenal phaeochromocytomas are said to excrete noradrenaline almost exclusively, as did our patient. In a series by Steinstrom et al., 23% were found to have diabetes.[4] After demonstrating excessive pro-duction of catecholamines, localisation of the tumour can be done through the various imaging modalities: CT scan, magnetic resonance imaging or by using the metaiodo benzyl guanidine (MIBG) test whereby the chromaffin tumour will take up the MIBG.

Definitive treatment is by removal of the tumour. Our patient did very well and did not require any pharmacological treatment after the tumour was removed. As there is a risk of recurrence, long-term follow up is necessary.

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