

Urinary Bladder Paraganglioma in a Post-Heart Transplant Patient

Dear Editor,

Both pheochromocytomas and paragangliomas are rare neuroendocrine tumours. Classical presentations of hypertension, paroxysmal palpitation and diaphoresis are related to catecholamine hypersecretion.¹

A 49-year-old Chinese man presented with exertional dyspnoea 8 years ago. With a presenting blood pressure (BP) of 160/90 mmHg, he was thoroughly investigated for secondary cause of hypertension. Echocardiogram demonstrated globally-impaired left ventricle (LV) with ejection fraction of 35%. Coronary angiography revealed no obstructive lesion with endomyocardial biopsy showing hypertrophy and interstitial fibrosis of myocardium. He was subsequently commenced on Carvedilol and Lisinopril.

He had been stable with New-York-Heart-Association (NYHA) class II until 2009 when he presented with out-of-hospital cardiac arrest due to ventricular fibrillation but successfully resuscitated. Biventricular pacemaker with defibrillator (CRT-D) was implanted in the presence of

widened QRS complex. In spite of this, he progressed to NYHA class IV and was referred for heart transplantation which was subsequently performed.

After the successful orthotopic heart transplantation, he was noted to have labile BP with systolic over 200 mmHg and persistent tachycardia of 110 beats-per-minute. During micturition, his systolic BP dropped profoundly to below 60 mmHg. With the strong suspicion of pheochromocytoma, a 24-hour urine sample was saved for catecholamine metabolites and urinary vanillyl mandelic acid (VMA) was found to be >20 times of upper limit of normal. Contrast computed tomography of abdomen and pelvis with intravenous contrast showed an avidly enhancing 2-cm mass within the urinary bladder (Figs. 1A and 1B) while ¹³¹I-metaiodobenzylguanidine (MIBG) scan demonstrated increased tracer uptake in corresponding region (Fig. 1C). Diagnosis of urinary bladder paraganglioma was confirmed with the patient started on phenoxybenzamine. Subsequently, radical cystectomy with ileal conduit was successfully performed for him who recovered uneventfully.

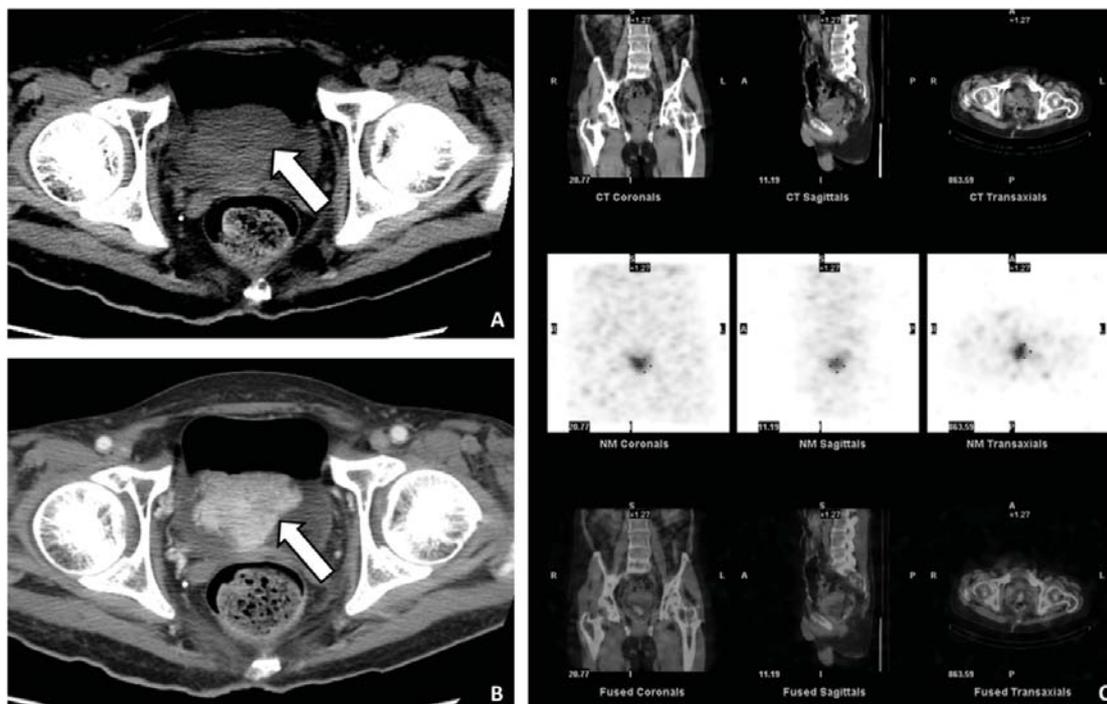


Fig. 1(A). Non-contrast axial computed tomography scan. An irregular bordered soft tissue lesion was noted within the urinary bladder [indicated by arrow]. (B). Axial computed tomography scan with intravenous contrast. An irregular bordered soft tissue lesion within the urinary bladder demonstrated avid heterogenous contrast enhancement [indicated by arrow]. (C). ¹³¹I-MIBG SPECT fusion scan showed increased uptake within the urinary bladder indicating the site of urinary bladder paraganglioma.

Chronic high blood level of catecholamine can facilitate various forms of myocardial damage, causing dilated or hypertrophied cardiomyopathy.² Diagnosis poses difficulties in the absence of hypertension.³ Indeed, the secretion of low level of catecholamine during early tumour development is insufficient to induce haemodynamic effects. In retrospect, it is likely the urinary bladder paraganglioma developed insidiously after the diagnosis of essential hypertension. With the further progression of his cardiomyopathy, his BP was even on the low side due to worsening left ventricular systolic function and cardiac output. This further lowered our index of suspicion of a catecholamine-induced cardiomyopathy, which classically presented with persistent or paroxysmal hypertension.⁴ Symptoms were revealed after transplantation with a normal functioning heart. Persistent tachycardia is common in denervated heart and may be overlooked. However, the occurrence of orthostatic hypotension during micturition in the background of uncontrolled hypertension should prompt further investigation. Urinary origin of paraganglioma should be suspected as tumour manipulation is notorious for causing catecholamine surge.⁵ Pathophysiological factors contributing to the phenomenon of hypotension in catecholamine-secreting tumour include intravascular volume depletion, abrupt cessation of catecholamine secretion due to tumour necrosis and desensitisation of adrenergic receptors.⁶

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