Teaching Point
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Labile hypertension, increased metanephrines and imaging misadventures

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Introduction

Paragangliomas arise from extra-adrenal chromaffin cells and represent ~15% of adult pheochromocytomas [1]. Most catecholamine-secreting paragangliomas are located in the abdomen and pelvis [2]. [123I]Metaiodobenzylguanidine (MIBG) scintigraphy is commonly used to localize these tumours and, although highly specific (95–100%), it lacks sensitivity (77–90%) [3]. We report a case of pelvic paraganglioma that presented with elevated plasma and urinary metanephrine levels and [123I]MIBG scintigraphy suggestive of left adrenal pheochromocytoma. Magnetic resonance imaging (MRI) of the pelvis showed a pelvic mass that was surgically resected (histology-proven paraganglioma), with post-operative normalization of blood pressure and both urine and plasma metanephrines.

Case

This 58-year-old female with a 5-year history of hypertension was referred to our institution for further evaluation of a possible pheochromocytoma. One month prior to her visit, she experienced the onset of episodic blood pressure elevations associated with headaches, dizziness, sweating and palpitations. Her physical examination was normal and her blood pressure and metanephrine levels were elevated.

Fig. 1. MIBG scintigraphy: increased left adrenal and pelvic uptake.

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Fig. 2. Pelvic MRI: a 4 cm mass next to the urinary bladder.

Fig. 3. Pelvic tumour 7 cm × 6.5 cm × 5 cm, 70 g.
pressure was 130/80 mmHg on clonidine, 0.1 mg twice a day. Her 24 h urinary excretion of catecholamines was normal, but the total metanephrine excretion was 2.9 mg (normal, < 1.3). The plasma normetanephrine concentration was increased at 14.7 nmol/l (normal, < 0.9). Abdominal MRI scan demonstrated asymmetric enlargement of the left adrenal gland 1 cm in maximum diameter. \[^{123}I\]MIBG scintigraphy demonstrated asymmetric adrenal gland uptake (left > right). Increased \[^{123}I\]MIBG uptake in the pelvis was interpreted as normal urinary bladder activity (Figure 1). Initially, left adrenalectomy was considered, but subsequently we pursued an evaluation for extra-adrenal catecholamine-secreting tumour for two reasons. First, most patients with symptomatic sporadic adrenal pheochromocytomas have tumours much larger than 1 cm [4]. Secondly, there can be normal asymmetry in adrenal \[^{123}I\]MIBG uptake. Further testing included an MRI of the pelvis that revealed a 4 cm vascular mass localized to the right side of the urinary bladder (Figure 2), which, in retrospect, correlated with the area of increased pelvic uptake on \[^{123}I\]MIBG scintigraphy. After the patient had been adequately prepared with \(\alpha\) - and \(\beta\)-adrenergic blockade, the pelvic tumour was surgically resected (Figure 3). Histology confirmed the diagnosis of paraganglioma. Urinary 24 h excretion of total metanephrines and plasma normetanephrine levels normalized post-operatively. Two years later, she remains normotensive with normal fractionated plasma metanephrine concentrations and urinary catecholamine and metanephrine excretion.

**Teaching point**

In a patient with elevated catecholamines or metanephrines, the use of MIBG scintigraphy for tumour localization can be misleading due to a normal asymmetry in uptake by the adrenal glands. In addition, MRI scan of the adrenal glands may be helpful in further evaluation, as most adrenal pheochromocytomas are larger than 1 cm. Finally, increased pelvic uptake should be carefully evaluated, especially in a patient without radiological evidence of an adrenal tumour, because a pelvic paraganglioma easily can be misinterpreted as normal accumulation of contrast in the bladder.

**Conflict of interest statement.** None declared.

**References**