Occult extra-adrenal pheochromocytoma in the urinary bladder

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DESCRIPTION

Pheochromocytoma is an uncommon neuroendocrine tumour arising from the chromaffin cells associated with increased catecholamine production. More than 85% of the tumours are infradiaphragmatic in location, with para-aortic region being the most common site.¹

Extra-adrenal pheochromocytoma (EAP) accounts for 10%–15% of pheochromocytoma in adults and 30%–40% in children.² EAP is sporadic and tends to be larger than the adrenal counterpart at the time of detection.³

Herein, the authors report a rare case of occult bladder EAP that evaded initial imaging modalities due to diagnostic pathway loopholes. A 6-year-old girl had presented with a 1-year history of hypertension, diaphoresis and episodic postmicturition headache. Urinalysis revealed elevated norepinephrine, epinephrine and vanillylmandelic acid levels, highly suspicious for pheochromocytoma. The contrast CT scan of the abdomen and ultrasound (US) for renal artery stenosis performed at another hospital did not include the pelvis in the scanned region and were otherwise unremarkable. Fluorodeoxyglucose-positron emission tomography (PET) showed absent pelvic uptake. The I¹³¹-metaiodobenzylguanidine scan was negative. The diagnosis was finally suggested on C¹¹ L-3,4-dihydroxyphenylalanine PET scan of which a focus of increased uptake was noted at the anterior bladder.

A pelvic US demonstrated a 6 cm hypoechoic intramural mass at the dome of the urinary bladder with preserved epithelial lining (figure 1). Colour Doppler imaging (figure 2) revealed prominent peripheral vessels which extended to the rest of

Figure 2 Colour Doppler imaging of the bladder mass (A,B), demonstrating intravesical vascularity. Prominent peripheral vessels are seen extending to the rest of the bladder wall.

the bladder wall. No vascular communication with the adjacent bowel loops was seen. The histopathological diagnosis of EAP was confirmed after an uneventful surgical resection of the bladder tumour.

Learning points

- ▶ Although rare, urinary bladder pheochromocytoma should be considered as one of the possible extra-adrenal sites.
- ▶ The authors recommend the inclusion of pelvic imaging as part of the routine protocol for the screening of pheochromocytoma.
- ▶ This case illustrates the high-spatial resolution and multiplanar capability of ultrasound evaluation, which mirrored the intraoperative findings in this case.

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Figure 1 An ultrasound of the pelvis of a 6-year-old girl, demonstrating a hypoechoic intramural soft tissue mass at the urinary bladder dome, measuring 5.8×2.9×5.7 cm. Sonographic images are represented in (A) the sagittal view and (B) the transverse view.

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