Occult Extra-adrenal Pheochromocytoma In The Urinary Bladder

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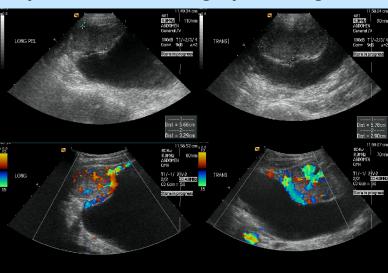
Background & Case Presentation

- Pheochromocytoma is an uncommon neuroendocrine tumour arising from the chromaffin cells associated with increased catecholamine production
- Extra-adrenal pheochromocytoma (EAP) accounts for 10-15% of pheochromocytoma in adults and 30-40% in children
- EAP are sporadic, and tend to be larger than the adrenal counterpart at the time of detection
- A 6-year-old girl presented with a 1 year history of hypertension, diaphoresis and episodic post-micturition headache
- Urine analysis had revealed elevated noradrenaline, adrenaline and vanillylmandelic acid levels, highly suspicious for pheochromocytoma

Diagnostic Loopholes and Eventual Management

- **1.** The initial CT abdomen and renal artery stenosis USG scan in another hospital **did not include the pelvis** in the scanned region, and were otherwise unremarkable.
- **2.** Fluorodeoxyglucose-positron emission tomography (FDG-PET) showed no pelvic uptake.
- **3.** The I¹³¹-metaiodobenzylguanidine (MIBG) scan was negative.
- **4.** Finally, the diagnosis was suggested on C¹¹ L-3,4-dihydroxyphenylalanine (L-DOPA) PET scan of which a focus of increased uptake was noted at the anterior bladder.
- **5.** A dedicated **pelvic USG** confirmed the presence of a soft tissue mass at the urinary bladder dome.
- **6.** Uneventful surgical resection was subsequently performed.
- **7.** Follow-up imaging revealed no recurrence of UBP.

Representative Sonographic Images



Ultrasound images demonstrate a 5.8 x 2.9 x 5.7 cm hypoechoic intramural soft tissue mass at the dome of the urinary bladder. The epithelial lining of the bladder was preserved. Colour doppler imaging had revealed prominent peripheral vessels which extended to the rest of the bladder wall.

Discussion

- ✓ Although rare, UBP should be considered as one of the possible extra-adrenal sites
- ✓ The authors recommend the <u>inclusion of pelvic imaging</u> as part of the routine protocol for the screening of pheochromocytoma
- ✓ This case also illustrates the high spatial resolution and multi-planar capability of US evaluation, which mirrored the intra-operative findings of histopathologically proven EAP

References

- 1. Ansari MS, et al. Malignant paraganglioma of the urinary bladder: A case report. Int Urol Nephrol. 2001;33:343–5.
- 2. Purandare NC, et al. Pheochromocytoma of the urinary bladder. Ultrasound Med. 2005;24:881-3

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