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 I$^{131}$-metaiodobenzylguanidine (MIBG) scan was negative.
4. Finally, the diagnosis was suggested on C$^{11}$ 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 inclusion of pelvic imaging 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

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