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Disease-Focused Panels

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Multimodality imaging of Bladder Paragangliomas

Presented By:
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Case Presentation:

- A 36 year old female presented with a history of intermittent lower abdominal pain for the past two months and post-micturition syncope.
- Review of systems- Episodic headaches, Palpitations and Sweating. BP 180/110
- Laboratory investigations revealed normal CBC, thyroid function tests and CBP.
- Urinalysis showed microscopic hematuria. 24-hour urine norepinephrine was elevated.
- Cystoscopy showed smooth elevated yellowish colored nodule measuring 3 x 2cm left posterolateral bladder wall.

Diagnostic Imaging :

Ultrasound Imaging features:

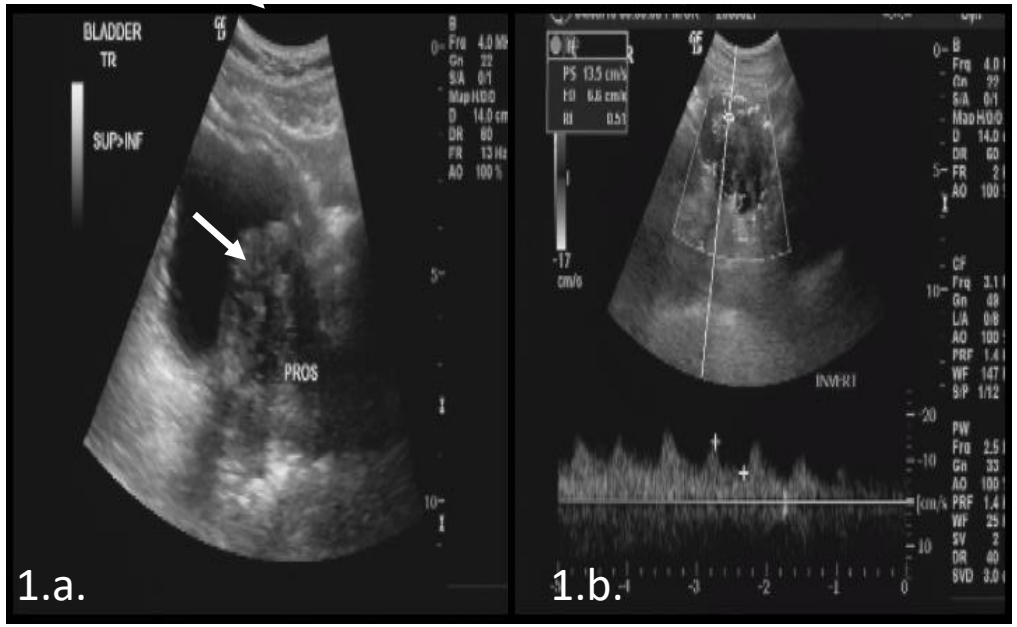

Fig 1a, 1b: Gray scale & color Doppler images showing a hetero echoic intravesical mass arising from the posterior bladder wall (white arrow). Arterial flow is demonstrated on the spectral waveform indicating a highly vascularized tumor.

CT Imaging Features:

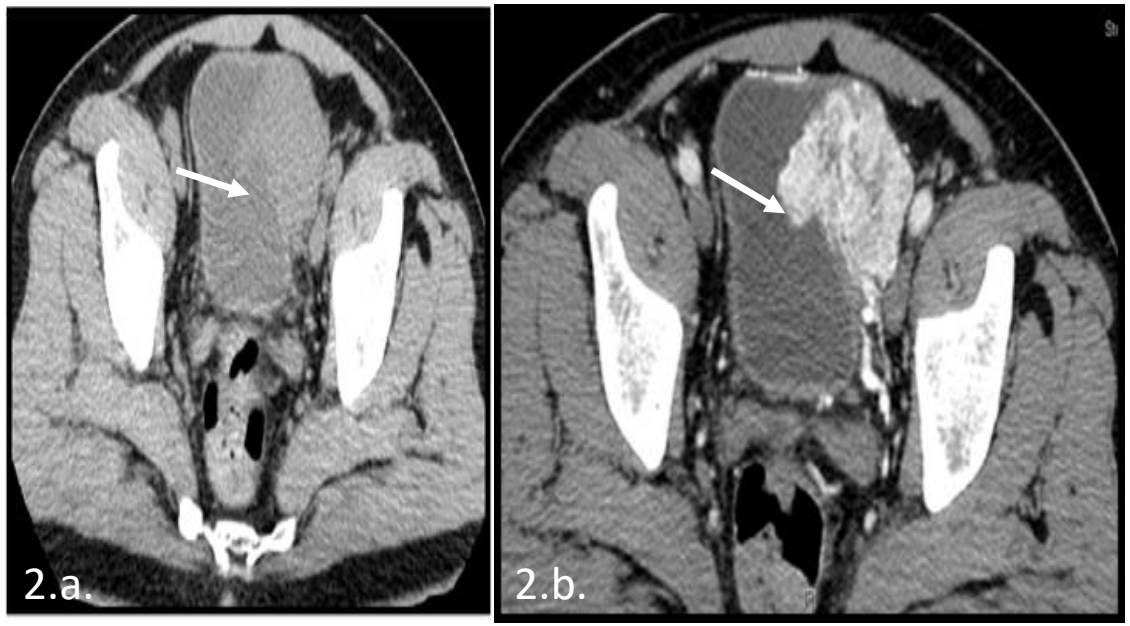

Fig 2a: Axial NECT shows a well defined homogenous soft tissue density mass (white arrow) in the left lateral wall of the bladder. Fig 2b: Corresponding CECT shows marked heterogenous contrast enhancement of the tumor (white arrow).

MR Imaging Features:

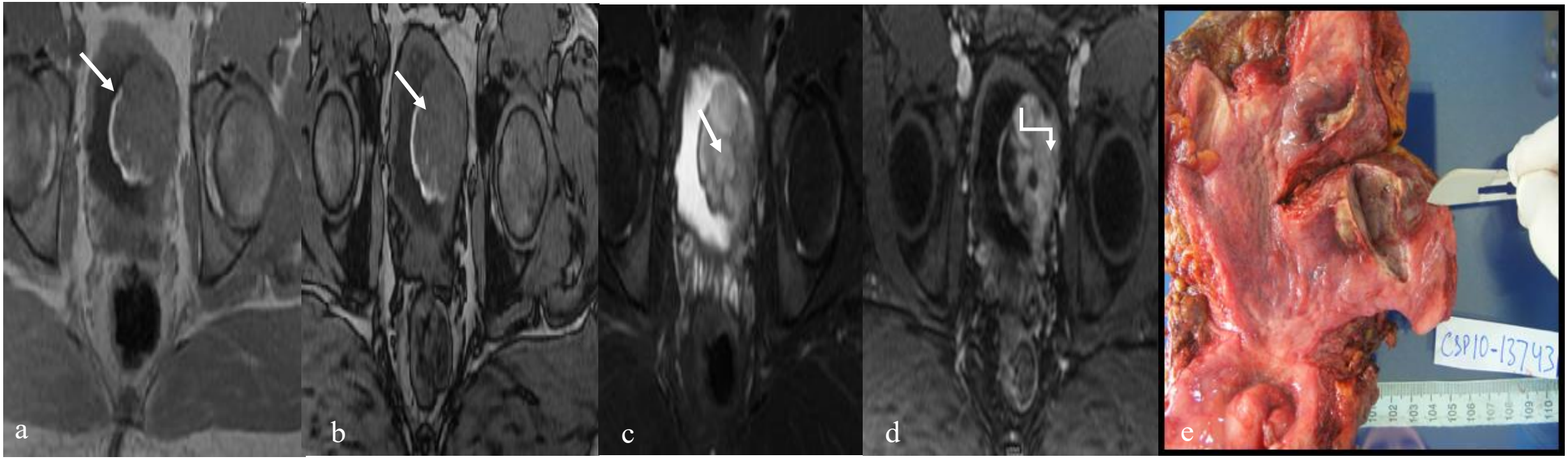

Fig 3a: Axial in phase T1W MRI shows an intermediate signal intravesical lesion with a rim of high signal due to hemorrhage (white arrow). Fig 3b: Axial opposed phase T1WI demonstrates no drop in signal, indicating low intravoxel fat content (white arrow). Fig 3c: Axial T2W fat saturated MRI shows high signal (white arrow). Fig 3d: Axial T1WI fat saturated MRI shows heterogenous lesion enhancement with characteristic 'salt and pepper' pattern due to increased tumor vascularity. 'Salt' represents enhancing parenchyma, 'pepper' represents a flow void (white arrow). Pathology proven paraganglioma.

Fig 3e: Gross specimen shows a surgical firm mass with cystic and hemorrhagic areas.

Histopathology of the specimen:

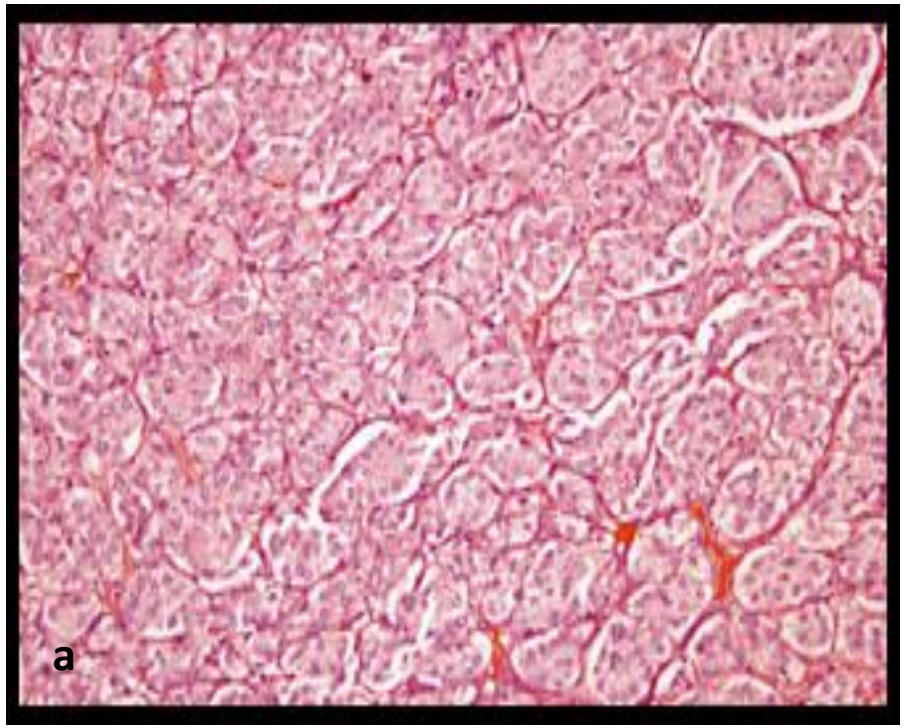

Fig: 4a. Cells arranged in classic clusters “zellballen” separated by fibrovascular cores (200X magnification). These clusters contain nest of chromaffin cells with pale eosinophilic stain surrounded by sustentacular cells (S-100 positive and cytokeratin negative). Also described as ”loose cell clusters with vascular coats”.

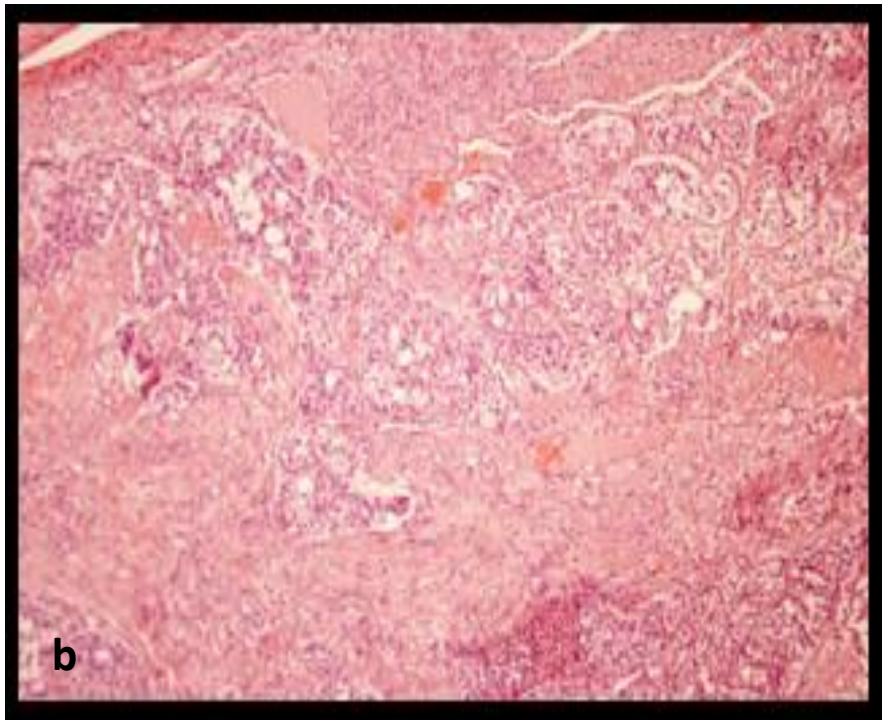

Fig:4b. Nests of pheochromocytoma cells invading in between bundles of smooth muscle (20X magnification)

Histologically extra-adrenal paragangliomas are identical to adrenal pheochromocytomas. The cells are arranged in Zellballen pattern surrounded by highly vascular fibrous network. Chief cells are positive for synaptophysin/chromogranin.

Discussion:

- Paragangliomas of the bladder are rare tumors originating from chromaffin cells of extra-adrenal sympathetic nervous system.
- They account for 0.1% of all the bladder tumors and less than 1% of all pheochromocytoma.
- Approximately 10% of all extra-adrenal paragangliomas occur in the bladder and 79.2% of genitourinary paragangliomas are seen in the bladder.
- Female to male ratio- 3:1; Common age group: 30-50 years.
- Associations: Familial paragangliomas, Carney triad (pulmonary chondromas, paragangliomas, GIST), NF1, MEN2, VHL.

Discussion:

- Usually discovered incidentally at imaging. Symptomatic patients may present with paroxysmal attacks of headache, palpitations, sweating, and tremors, secondary to catecholamine release.

- Associated with elevated levels of vanillyl mandelic acid (VMA) in plasma and in 24 hour urine collection (>6.7mg/24hrs).

- Elevated urinary normetanephrine and norepinephrines can establish the diagnosis. If these tests turn out normal in clinically suspected patients, I131 MIBG (Metaiodobenzylguanidine) scintigraphy is a good diagnostic test (sensitivity of 83-100%)

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Discussion:

- Differential diagnosis for bladder tumors:

  - Leiomyoma: MC in middle aged women; Mild to moderate even enhancement
  - Carcinoma bladder: Seen in elderly people; Uneven enhancement
  - Rhabdomyosarcoma: MC in children; Significant strength enhancement
  - Lymphoma: seen in mid to elderly people; Mild to moderate enhancement

- Extra-adrenal paragangliomas are more likely malignant than adrenal lesions. Malignant and metastatic tumors are associated with a poor prognosis.

- Gross & histopathological examination cannot differentiate benign and malignant tumors. severity of malignancy is based on depth of invasion, nodal involvement and extent of metastasis.
- Comprehensive radiological, hormonal and pathological correlation is therefore essential for accurate diagnosis.

Take home points:

- Bladder paragangliomas have non specific appearance. Despite characteristic imaging findings, definite diagnosis relies on pathology. Specific hormonal imbalance may support diagnosis.

- **CECT:** Increased vascularity gives characteristic intense heterogenous avid enhancement.

- **MRI:** T1WI contrast demonstrates ’salt and pepper’ pattern. T2WI shows markedly high signal secondary to increased water content from necrosis.

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