Gastroenteropancreatic & Genitourinary
Neuroendocrine tumors
Bladder NET (Paraganglioma)

Presented By:

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Case 1: 65 year-old hypertensive male with flank pain, with occasional palpitations and night sweats

Axial (A) and Coronal (B) CT scans in the excretory phase, showing a 1.9 cm nodule involving the right posterolateral wall of urinary bladder

(C) MIBG scan shows corresponding uptake within the right-sided bladder wall nodule, consistent with paraganglioma
Diagnosis: Paraganglioma

- Although initial consideration was urothelial cancer. Cystoscopy revealed an extrinsic mass indenting the urinary bladder with overlying normal appearing mucosa/urothelium.

- Given the history of long standing hypertension, along with occasional palpitations and night sweats (as well as had prior history of syncope following prostate biopsy on further questioning), adrenergic or sympathomimetic tumor was suspected.

- MIBG scan was diagnostic. Partial cystectomy was performed, confirming the diagnosis.
Imaging features of paragangliomas:

• **US:**
  A well-defined heterogeneously echogenic mass with significant internal vascularity

• **CT:**
  A hypodense hypervascular mass. CT localizes the mass, shows extension, evaluate nodal and regional involvement.

• **MRI:**
  Minimal hyperintensity on T1-weighted images, (hyperintense foci and hypointense foci) representing classical salt and pepper appearance. Hyperintense on fluid sensitive images. Problem solving, evaluate nodal and regional involvement.

• **NM:**
  High uptake on MIBG scans and High FDG-avidity on PET CT scans.
Discussion:

• Paragangliomas are extra-adrenal pheochromocytomas, accounting for about 15% of all pheochromocytomas
• They are extremely rare in the urinary bladder (0.06% of all bladder tumors)
• More common in females than males (3:1) and, occur primarily between 30 and 50 years of age
• Common symptoms are hematuria, hypertension, headache, palpitations and post-micturition syncope
• Back/flank pain may be the only presenting symptom of some paragangliomas.
• These can even be silent (about 17%), which may be quite dangerous as may be easily misdiagnosed
Discussion:

- About 88% of cases of paraganglioma show elevated levels of urine metanephrine and serum catecholamine.
- Both computed tomography (CT) scan and magnetic resonance imaging (MRI) are useful for localization of both the primary tumor and any metastases, and management planning.
- Functional imaging that targets the catecholamines pathways, is very helpful not only for original tumor localization/diagnosis, but especially also after surgical removal for detection of recurrence or metastases.
- $^{131}$Iodine metaiodinebenzylguinidine (MIBG) scans have high sensitivity (77-90%) and specificity (95-100%) for pheochromocytoma/paraganglioma detection.
World Health Organization (WHO) histological classification of neuroendocrine neoplasms of the genitourinary tract in adults.

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<th>Kidney</th>
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<th>Prostate</th>
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<td>Well-differentiated neuroendocrine tumor</td>
<td>Adenocarcinoma with neuroendocrine differentiation</td>
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<td>Small Cell neuroendocrine carcinoma</td>
<td>Small Cell neuroendocrine carcinoma</td>
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<td>Paraganglioma</td>
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Courtesy of Dr. Dheeraj Reddy Gopireddy
References:


