A case of micturition syncope

Kimberly Bundick, PA-S

Yale SCHOOL OF MEDICINE
Physician Associate Program

Agenda

Purpose

- Utilize case to illustrate classic finding of an interesting pathology

Agenda

- Case study
- Epidemiology, etiology of disease
- Diagnosis and treatment
- Key Takeaways

Limit

- 5 minutes

Chief Complaint

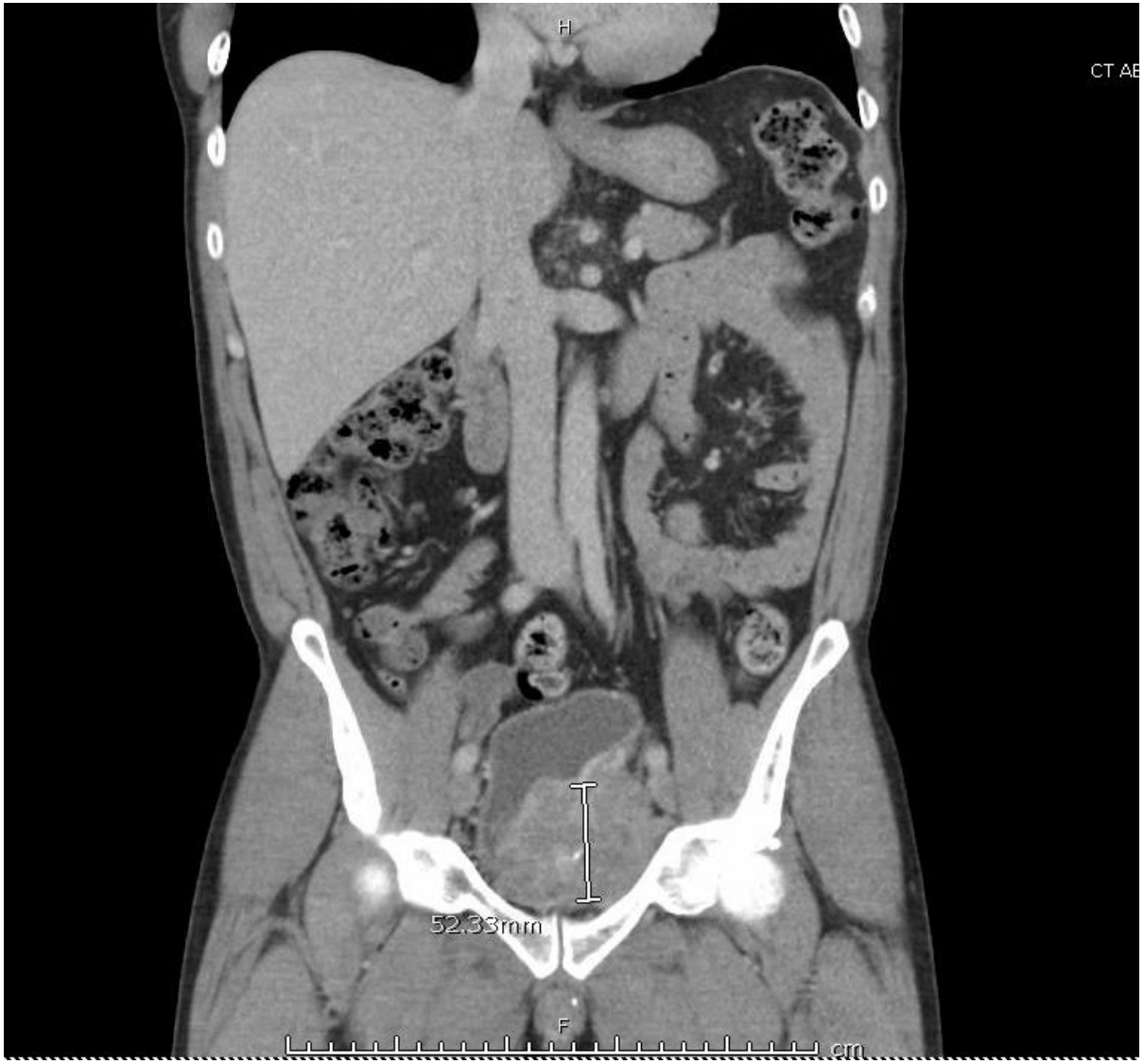
Indication for study:

- 40-year-old male, fainting and tachycardia with urination

Additional information:

- Recent hospitalization for uncontrollable hypertension at OSH
- 24-hour urine catecholamines were markedly elevated

CT Abdomen Pelvis without Contrast
Coronal View



CT Abdomen Pelvis without Contrast
Findings and Impression

FINDINGS:

There is a **heterogeneously enhancing mass containing a few calcifications centered within the left anterolateral bladder wall, compatible with a neoplasm.** Given the lab abnormalities and history, this likely reflects a **bladder pheochromocytoma.** This is associated with bilateral pelvic adenopathy, some of which is calcified. Along the left pelvic sidewall, this measures up to 1.7 cm, while along the right pelvic sidewall, this measures up to 1.3 cm.

IMPRESSION:

Findings highly concerning for a **bladder pheochromocytoma** with pelvic adenopathy as described

Paragangliomas are rare catecholamine-secreting tumors

DESCRIPTION

- Paragangliomas, much like pheochromocytomas, are rare neuroendocrine, catecholamine-secreting tumors
- Paragangliomas can be sympathetic (typically symptomatic, located in the sympathetic paravertebral ganglia of thorax, abdomen, and pelvis), or parasympathetic (typically asymptomatic / nonfunctional and located along the glossopharyngeal and vagal nerves in the neck and at the base of the skull)
- Distinction from pheos is important given key differences in location, clinical behavior, aggressiveness and metastatic potential, biochemical findings, and association with inherited genetic syndromes

EPIDEMIOLOGY

- Given overlapping characteristics with pheos, specific incidence is unknown but estimated at 0.8 per 100,000

ETIOLOGY

- ~30 – 50% are a component of an inherited syndrome; genetic testing is recommended for all patients with a paraganglioma and in selected patients with pheochromocytoma to appropriately characterize risk

MALIGNANT POTENTIAL

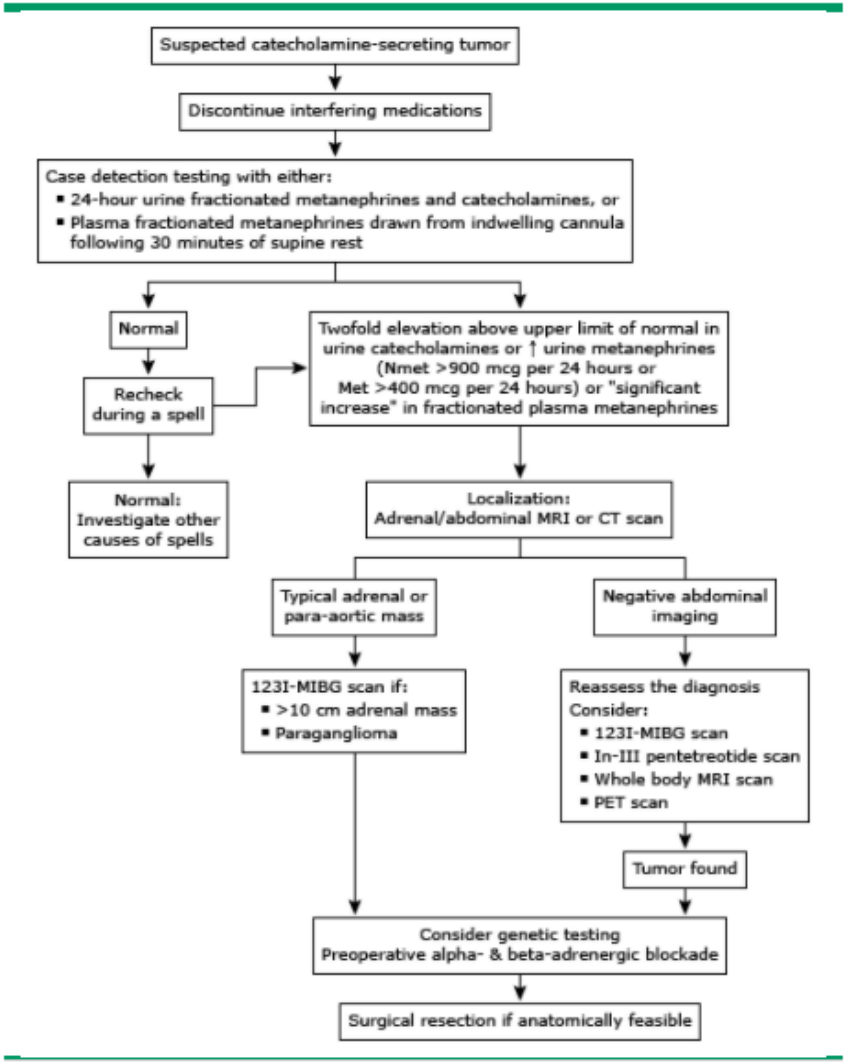
- Up to 25% of paragangliomas are malignant; malignant behavior indicated by metastatic spread

Micturition syncope is the classic feature of bladder paragangliomas

- Production of vasoactive catecholamines leads to symptoms of catecholamine excess including hypertension, episodic headache, sweating, and tachycardia
- Hypertension is the most common feature
- Micturition syncope is the clinical hallmark of a catecholamine-secreting bladder paraganglioma
  - –The triad of hypertension, hematuria, and symptoms on micturition or sexual activity is considered almost diagnostic of the condition, and is reported variably in 50 to 100 % of patients
  - –Some patients have only painless hematuria

Diagnosis is confirmed with lab testing and imaging

Evaluation and treatment of catecholamine-producing tumors



LAB TESTING

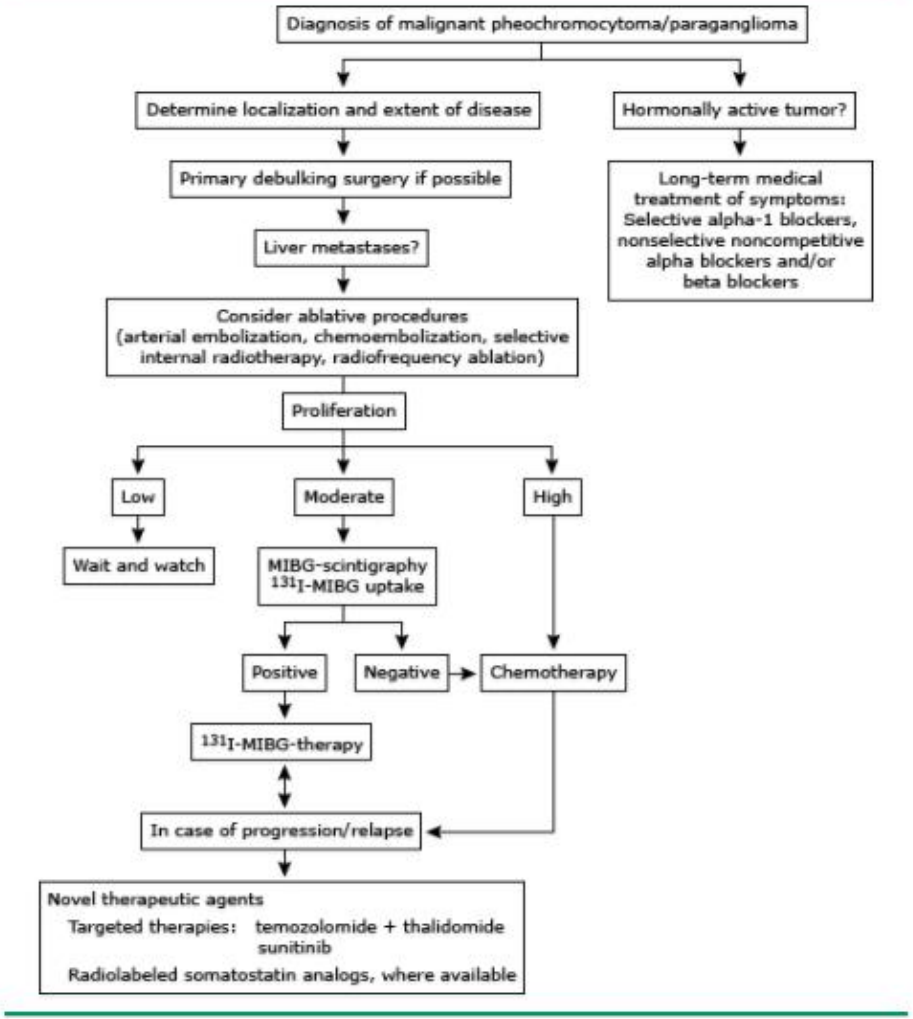
- Urinary and/or plasma fractionated metanephrines and catecholamines

IMAGING

- Radiologic imaging is an important component of assessment for information
- Location and a high degree of vascularity may to permit a presumptive preoperative diagnosis of a paraganglioma in setting of catecholamine secreting tumors
- Appropriate modalities:
  - Ultrasound
  - computed tomography
  - Magnetic resonance imaging (MRI)
  - Angiography
  - Radioisotope imaging using metaiodobenzylguanidine (MIBG)
  - 18F-fluoro-2-deoxyglucose positron emission tomography (FDG-PET)

Treatment of paragangliomas is focused on symptom control and surgical resection if possible

Treatment algorithm for malignant pheochromocytoma and paraganglioma



- There are no curative options for paragangliomas
- Symptoms of catecholamine excess should be controlled using combined alpha- and beta-adrenergic blockade
- Primary and metastatic lesions should be resected if possible
- The clinical course of metastatic paraganglioma is highly variable, with reported five-year survival rates that range widely from 12 to 84%

Key Takeaways

- Paragangliomas are rare, catecholamine secreting neuroendocrine tumors
- Present with symptoms of catecholamine excess, with the classic presentation of bladder paragangliomas being micturition syncope
- Diagnosis is made with a combination of clinical presentation, 24-hour urine catecholamines and imaging
- Treatment is focused on symptom management and resection, if possible

References

- Carty, S. et al. *Paragangliomas: Epidemiology, clinical presentation, diagnosis, and histology*. Uptodate.com, Oct. 2017
- Carty, S. et al. *Paragangliomas: Treatment of locoregional disease*. Uptodate.com, Oct. 2017.
- Carty, S., et al. *Paraganglioma and pheochromocytoma: Management of malignant disease*. Uptodate.com, Oct. 2017