Hypertension in a Pregnant Woman

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25 year old G3P0020 female at 36 weeks 2 days, noted to have BP 174/126 at prenatal visit

Asymptomatic at the time of exam

Admitted to ICU for further care

Past Medical History: Obesity, HTN, 2 first trimester miscarriages
  - Started on labetalol for HTN during second trimester
  - Noted episodes of diaphoresis, palpitations, lightheadedness
• **Social History:** Quit smoking 18 months prior, no EtOH, sedentary lifestyle

• **Family History:** Mother with paraganglioma resected 30 years ago, initially in chest (“grapefruit sized”), then along thoracic spine

• **Medications:** Ferrous sulfate, docusate PRN, labetalol

• **Physical Exam:** BP 174/126, repeat 162/120; 170/124 in other arm. HR 95. Well-appearing pregnant female in no acute distress. No peripheral edema, no bruits.
Further Studies

- Urinalysis negative for protein, RBCs
- Elevated free plasma normetanephrine at 794 (0-145). 24 hour urinary normetanephrines 2646 mcg/24h (normal 103-390), normal urinary VMA, elevated total urinary metanephrines 2770 mcg/24h (normal 145-510).
- BMP: Na 135, K 4.2, Cl 103, Cr 0.74
- Noncontrast MRI abdomen/pelvis: 2.5x2.4x1.5cm retrocaval mass concerning for extra-adrenal paraganglioma

Lee et al. JMIS 2014;17(1):15-19
Workup and Treatment

- **Treatment**: Phenoxybenzamine followed by propranolol, nicardipine infusion titrated to goal BP <130/80

- Elective Caesarean section at 37w2d with MFM, Anesthesia, Endocrine

- **Postpartum**: Endocrine and General Surgery consulted
  - CT w/contrast to further characterize mass
  - +Succinate dehydrogenase B (SDHB) mutation
  - I-123 MIBG testing negative for metastases

- Current plan: **Surgical resection** next week
Differential of Hypertension in Pregnancy

- Up to 10% of pregnancies
- 2 primary categories:
  - Chronic (essential/secondary) hypertension: Endocrine (thyroid, adrenal, pheochromocytoma/paraganglioma), cardiomyopathy, renal (renal artery stenosis, fibromuscular dysplasia), etc.
  - Pregnancy related: Preeclampsia-eclampsia and gestational HTN
- First-line treatment: labetalol, hydralazine, nifedipine
  - Pregnancy class C

Wilson et al. BMJ 2003;326:845
Pheochromocytoma/paraganglioma

- 0.2% patients with HTN, 0.002% of pregnancies
  - Maternal/fetal mortality ~50% if undiagnosed
  - Symptomatic during pregnancy due to palpation, fetal movement, uterus

- **Pheochromocytoma**: Adrenal, secretory

- **Paraganglioma**: Extra-adrenal, sympathetic ganglia
  - 30-50% part of a hereditary syndrome
  - Differential: MEN 2A and 2B, NF1, VHL
  - Hereditary: Mutations in SDHA/B/C/D, VHL and NF1

Pheochromocytoma/paraganglioma

- Diagnosis based on exam, FH, imaging, labs and genetic testing
  - Consider with episodic symptoms, orthostatic hypotension, cardiomyopathy, multiple/recurrent tumors, onset <45y
  - Autosomal dominant, variable penetrance
  - CT > MRI (90-100% sens / 70-80% spec)
  - Metastases: I-123 MIBG scintigraphy or FDG-PET
- Labs: **Blood/urine metanephrines** more sensitive (95-100% sens) than plasma catecholamines

*Young et al. J Clin Endocrinol Metab 2002;87:4101-5*
Pheochromocytoma/paraganglioma

- Management: **alpha blockade** (titrate to low-normal SBP) followed by **beta blockade** (titrate to HR 80) followed by **surgical removal**.
  - Treatment reduces surgical risk due to catecholamine surges
  - In pregnancy, remove before 24 weeks or after delivery
  - Remove SDHB+ tumors as quickly as possible due to metastatic potential
  - Genetic counseling/screening of family members
  - Lifelong screening via labs and imaging

*GeneReview: Hereditary Paraganglioma-Pheochromocytoma Syndromes, Kirmani and Young. 2014, NCBI.*
Thank you!

References: