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May 3rd, 8:00 AM

A Rare but Life Threatening Case of Labile Blood Pressures in Pregnancy

Kelsey Carrio MD
Rowan University SOM

Aarti Aggarwal MD
Rowan University SOM

Susan McGroarty PhD
Rowan University SOM

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Carrio, Kelsey MD; Aggarwal, Aarti MD; and McGroarty, Susan PhD, "A Rare but Life Threatening Case of Labile Blood Pressures in Pregnancy" (2018). *Stratford Campus Research Day*. 9.
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A Rare but Life Threatening Case of Labile Blood Pressure in Pregnancy

Carrio, K, MD, PGY3; Aggarwal, A, MD; McGroarty, S, PhD.

Inspira Health Network, Family Medicine Woodbury, NJ

INTRODUCTION

Pheochromocytoma is a rare but life threatening catecholamine secreting tumor and has a reported incidence of <0.2 per 10,000 pregnancies (1). Despite its rarity, untreated pheochromocytomas carry a risk of mortality for both mother and fetus, as high as 58% (2). This may be due to several factors, such as varying presentation, rarity, pregnancy precluding diagnosing studies (1,3,4). Often patients are thought to have superimposed preeclampsia and is important to note diagnostic differences in these severe cases. The infrequency with which it is encountered makes pheochromocytoma, especially in pregnancy, a formidable diagnostic challenge.

CASE

A 35 year old Japanese G1 diet controlled gestational diabetic with no other PMH presented to labor and delivery at 32 weeks and 2 days gestation for an evaluation of blood pressures (BP) in the 200s/100s. Patient's high BP persisted, and then she became symptomatic with headaches, orthostatic hypotension and blurry vision. Treatment was initiated with Mg drip for seizure prophylaxis and IV Labetalol. Pregnancy induced hypertension blood work revealed elevated liver enzymes, elevated uric acid and with her clinical presentation, she was deemed to be severe preeclamptic. During her 2nd dose of Labetalol her pressure suddenly dropped to 70s systolic. Maternal fetal medicine recommended urgent delivery via cesarean section. After a successful delivery, her labile BP continued. She was having significant orthostasis with a witnessed fall. With the help of Cardiology, Endocrinology & Critical Care specialists, patient's BP was stabilized with oral meds. Before discharge, endocrinology requested serum plasma metanephrines levels to be checked for completeness to rule out a more serious cause for labile blood pressures. Patient was deemed safe for discharge with self monitoring blood pressure and close follow up.

On postpartum day 12, her lab results came back with norepinephrine level of 2046 (normal range 7-65mcg/g), normetanephrine level of 4199 (normal <148pg/mL) and 24 h cortisol of 74.3 (normal 4-50mcg/24hr). Patient was called to return to hospital ASAP for further management. On arrival her BP was 200/138mmHg, we attempted to control her blood pressure with IV Labetalol and Cardura as was our only α blockade medication in stock. Patient was scanned with MRI the following morning and our suspicions were confirmed. Her MRI showed a large heterogeneous cystic & solid mass consistent with a pheochromocytoma/paraganglioma interposed between the aorta and IVC measuring 6.9 cm x 5.1 cm x 7 cm. Patient was transferred to Jefferson University Hospital, where she received 11 days of α blockade with Phenoxybenzamine followed by β blockade with Propranolol. She underwent exploratory laparotomy with resection of paraganglioma with no complications. Patient was doing well, discharged on day 13 in stable condition. Patient followed up 2 weeks later taking no medications, home blood pressures running in the 110s/70s. Her blood pressure in the office was 113/78, she had a successful surgical cure.

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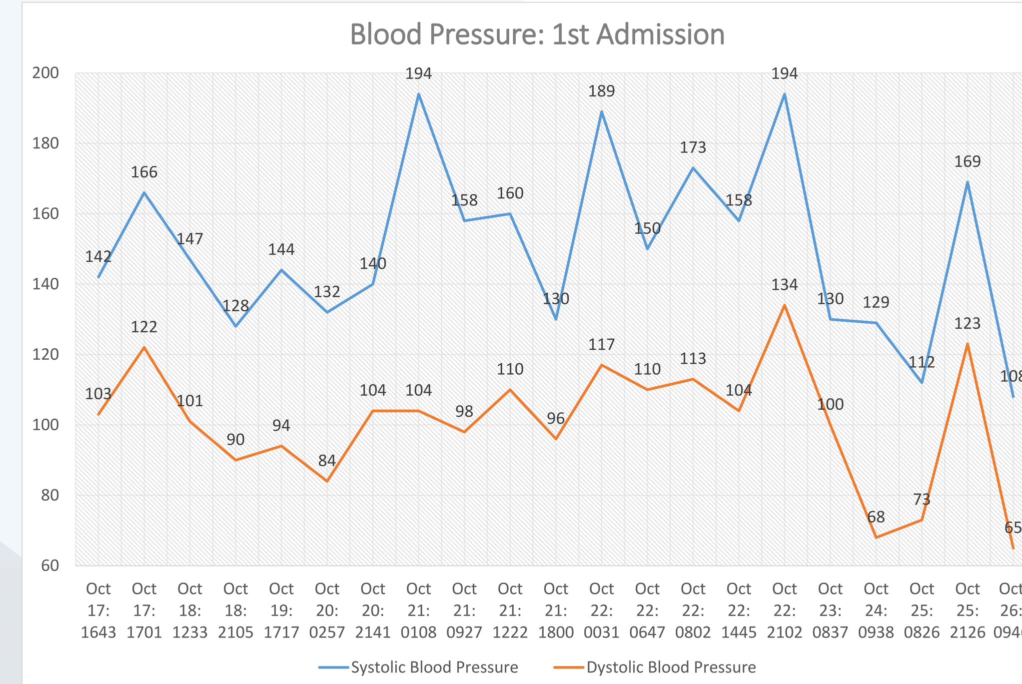
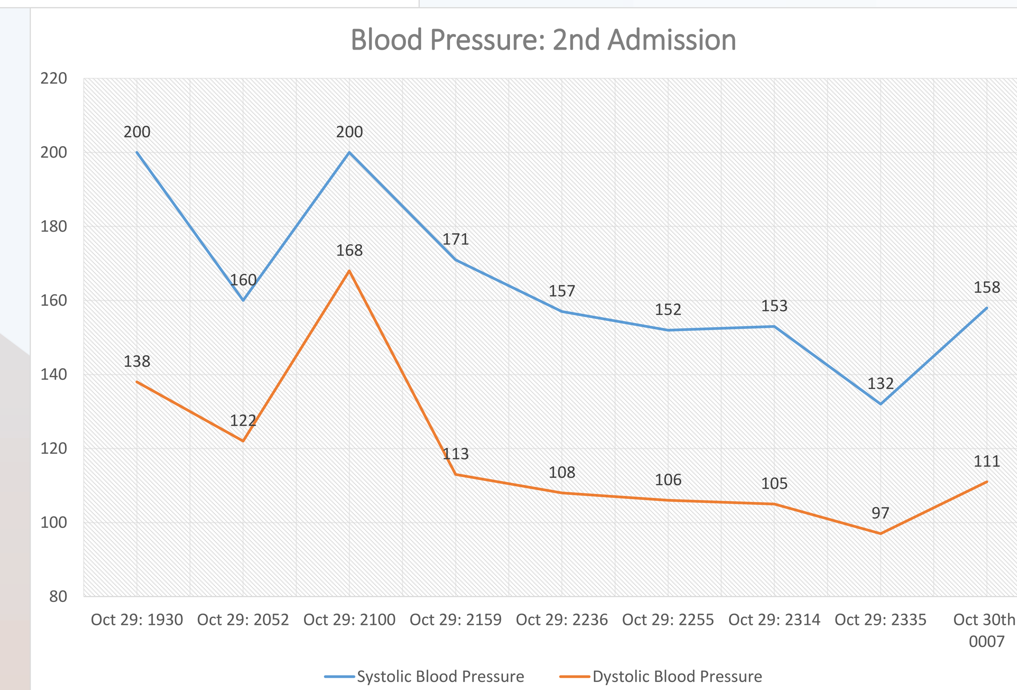
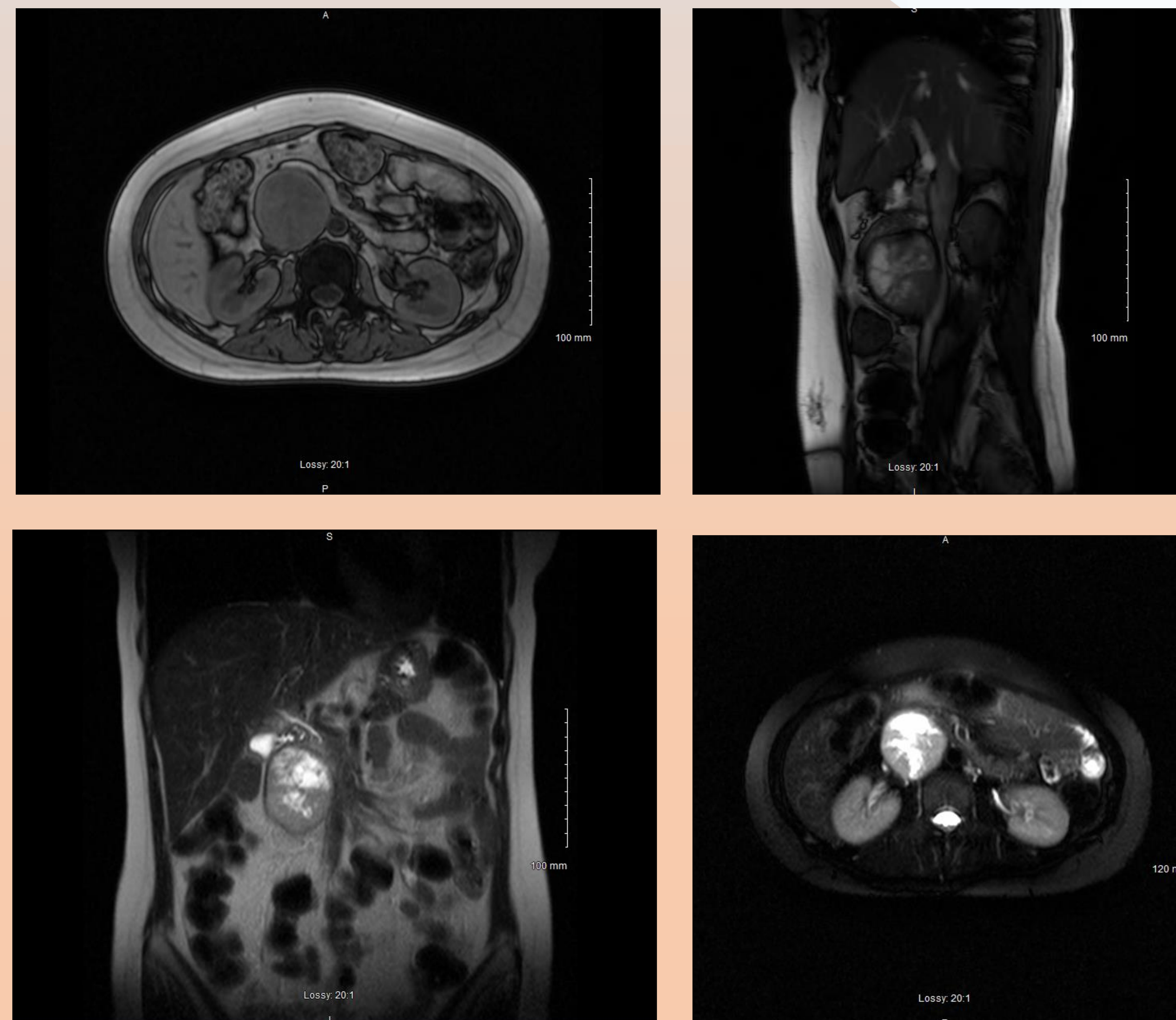
DATA





IMAGING



MRI Impression: Large 6 cm cystic solid mass right retroperitoneal space interposed between the aorta and IVC at the level of L2-L3 most consistent with a pheochromocytoma/paraganglioma felt to be extra adrenal in origin. There is significant mass effect upon the IVC, pancreas and duodenum.

LABS

- October 17, 2016
  - Uric Acid 10.4 (2.5-7 mg/dL)
  - Fibrinogen 658 (197-475mg/dL)
  - D-Dimer 1.43 (0-0.49ug/ml)
  - UDS negative
  - Urine protein 18 (0-11.9 mg/dL)
- October 24, 2016
  - Norepinephrine 2046 (7-65 mcg/g)
  - 24 hour cortisol 74.3 (4-50mcg/24hr)
  - Metanephrines plasma free <25 (<57pg/mL)
  - Normethanephrine plasma free 4199 (<148pg/mL)

DIFFERENTIATING PREECLAMPSIA VS PHEOCHROMOCYTOMA (5)

Features	Preeclampsia	Pheochromocytoma
Time of presentation	>20 wk. of gestation	Anytime during pregnancy
Hypertension	Usually sustained	Paroxysmal
Orthostatic hypotension	Absent	Present
Bipedal edema	May be present	Absent
Headaches	Usually in more severe preeclampsia	Present
Flushing	Absent	Present
Palpitations	Absent	Present
Weight gain	Present	Absent
Abdominal pain	Present	Absent
Proteinuria	Present	Often absent
Glucose	Normal	Elevated
Liver transaminases	Elevated	Normal
Catecholamine	Normal	Elevated
Thrombocytopenia	May be present	Normal

DISCUSSION

This case illustrated the potential for missing an important finding without proper consultants and medical management. Although a pheochromocytoma is rare, it is good to keep on your differential as a possible cause of labile blood pressures with headaches and significant orthostatics. Recognition of this condition is critical for appropriate therapy and surgical intervention to prevent mortality.

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