Pheochromocytoma Presenting as Hemorrhagic Stroke.

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Objective:
- Recognize pheochromocytoma as a differential diagnosis in a patient presenting with hemorrhagic stroke and uncontrolled hypertension.
- Discuss challenges in management of pheochromocytoma encountered in a patient with cerebrovascular accident (CVA).

Case Report:
- 59-year-old gentleman presented to emergency department with sudden onset headache, word finding difficulties and blurry vision.
- His past medical history was significant for hypertension and recurrent left axillary artery thrombus.
- His medication list included amldipine, lisinopril, labetalol and Rivaroxaban.
- On presentation, Blood pressure was 186/110 mm Hg, pulse 97, temperature 97.9 °F and respiratory rate of 11/minute.
- On physical examination, the patient was alert but with intermittent episodes of aphasis and right homonymous hemianopsia. He did not have dysarthria, motor or sensory deficits.
- His initial laboratory evaluation was unremarkable.
- Imaging: Computed tomography (CT) of the head identified a large left occipitoparietal intraparenchymal hemorrhage with 4 mm of midline shift to the right. There was no vascular malformation on CT angiography of the head.
- Patient was given prothrombin complex concentrate.
- During the course of hospitalization, his blood pressure remained exceedingly difficult to control requiring multiple antihypertensive agents.
- Hematology was involved given the history of recurrent left axillary artery thrombosis.
- Hypercoagulable work up including antithrombin, protein C, protein S, Factor V Leiden, prothrombin, lupus anticoagulant was unremarkable.
- CT of the chest, abdomen and pelvis was obtained which revealed 5.7 x 4.4 cm left adrenal mass.
- Hormonal workup for the adrenal mass revealed markedly elevated serum norepinephrine 11536 pg/ml (80 – 520), normetanephrine 4922 pg/ml (<148) and urine total metanephrines 11883 (149-603 mcg/g cr), consistent with pheochromocytoma.

Imaging

On further questioning, patient did endorse episodic headache, palpitations, flushing and lightheadedness prior to admission, occurring without any precipitating event.
- Episodic severe catecholamine-induced hemodynamic disturbance contributed to his presentation as stroke.
- In our patient, difficult to control blood pressure in hospital, high levels of catecholamines is suggestive of pheochromocytoma as a cause of stroke.

Treatment: Patient was started on alpha blockade during the hospital stay with a good blood pressure control, followed by volume expansion and beta blockade to prepare for surgery
- The decision was made by endocrine and neurosurgery to proceed with adrenalectomy 6 weeks after the acute stroke.

Discussion:
- Pheochromocytoma is a rare catecholamine-secreting tumor occurring in 2-8/1000,000 individuals. The most common clinical presentation is sustained or paroxysmal hypertension.
- Cerebrovascular accident is an atypical initial presentation of pheochromocytoma. In literature, there are total of seven cases reported which included ischemic stroke and transient ischemic attack in patients with pheochromocytoma.
- As an internist, it is crucial to be aware of pheochromocytoma and keep it in the differential diagnosis for uncontrolled hypertension in both outpatient and inpatient setting. (Does not mean screening in all patients)
- Any patient with adrenal mass should undergo complete hormonal workup irrespective of the size of mass and presence of hypertension.
- It is important to recognize pheochromocytoma as the cause given the management is different. Beta blockers can cause worsening hypertensive crisis leading to more end organ damage.
- Beta blockade before alpha adrenergic blockade should be avoided given the concern for worsening hypertensive crisis.
- Elective surgeries after acute stroke are usually postponed however surgical resection is the only definitive treatment of pheochromocytoma. In different case report, the surgery was done 2-10 weeks after the stroke. It is important to ensure adequate preoperative management with complete alpha blockade before surgery.
- Multidisciplinary approach including neurology, neurosurgery, endocrine surgery is warranted and further studies are needed.

References: