Case Report

A rare case of bilateral adrenal pheochromocytoma with unusual presentation: the case report

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ABSTRACT

Here we report a 25 years old male patient came to our emergency department with chief complaints of headache from 3 months, blurring of vision 7 days, abdominal pain 7 days, and vomiting from 2 days. Young male, with no known comorbidities, presented to our casualty with headache since, 3 months, episodic in nature, increased over the last 1 week, holocranial, gradually progressive in severity. last recorded Blood pressure outside was 220/120 mmHg and papilledema were noted on fundoscopy. USG abdomen and pelvis, CECT abdomen done and diagnosed to have bilateral adrenal pheochromocytoma. Patient underwent bilateral adrenalectomy was performed with good intra op BP control. Right adrenals were enlarged and shows multiple nodular lesions, left adrenal noted shows nodular lesions. Surgery remains the first line of management in malignant pheochromocytoma. Norepinephrine, epinephrine, and dopamine all act on their target receptors, which causes a physiological change in the body. Increase in catecholamines cause severe hypertension and multiple systemic complications like (cardiovascular, cerebrovascular events) and can lead to death if untreated.

Keywords: Adrenal glands, Adrenalectomy, Catecholamines, Chromogranin A, Metanephrine, Normetanephrine

INTRODUCTION

Pheochromocytomas and paragangliomas are catecholamine-producing tumors derived from the sympathetic or parasympathetic nervous system.1 These tumors may arise sporadically or be inherited as features of MEN type 2, von Hippel-Lindau disease, or several other pheochromocytoma-associated syndromes (SDHx).2

Here, we report a 25 years old male patient came to our ER with chief complaints of headache from 3 months, blurring of vision 7 days, abdominal pain 7 days and vomiting from 2 days. Young male, with no known comorbidities, presented to our casualty with headache since, 3 months, episodic in nature, increased over the last 1 week, holocranial, gradually progressive in severity. Initially dull and irritating type, gradually progressed to throbbing type of headache, aggravated on movement and relieved after a brief period of rest or analgesics. Headache was associated with blurring of vision since, 1 month aggravated over 1 week. No h/o radiation of pain to neck. No associated facial pain or eye pain, redness of eye, watering of eye. No precipitating factors (food, loud sounds and photophobia). No h/o diplopia, slurring of speech, deviation of angle of mouth, weakness of any of the limbs, h/o pain abdomen since 1 week, insidious onset, episodic in nature, non progressive, dull pain in the abdomen, non-radiating with no aggravating and relieving factors. It was associated with 2-3 episodes of vomiting over the last 2 days, Vomitus was non-projectile, non-blood or bile stained. Reports improvement in headache after vomiting. No h/o
fever, loose stools/constipation. No h/o chest pain, breathlessness, cough, syncopal episodes. No h/o giddiness, ear pain or ear discharge. No h/o loss of weight, tremors despite loss in appetite. In the 3 days prior to the EMD visit here, the patient had multiple hospital visits with the above-mentioned complaints and was found to have elevated blood pressure on multiple occasions, last recorded Blood pressure outside was 220/120 mmHg. No past history of hypertension in childhood. No h/o diabetes, dyslipidaemia, bronchial asthma, tuberculosis in the past.

CASE REPORT

Examination findings

24 years old young male, presented with the above complaints to the casualty. Patient is conscious, partially oriented. Vitals include pulse of 94 bpm, normal in rate, rhythm, volume, no radio-femoral, radio-radial delay. BP 90/60 mm of Hg in right brachial artery, supine position, (prior administration of antihypertensives outside where BP was noted to be 220/120 mm Hg). So2- 99% on RA. RR – 18 cpm. General physical examination - no pallor, icterus, cyanosis, generalized lymphadenopathy, clubbing or pedal edema. Papilledema was noted on fundoscopy - no neck stiffness, no neurocutaneous markers noted and no bruit noted over the carotids and abdomen.

Systemic examination

CVS- S1, S2 heard, no added sounds, no murmurs. JVP - normal, no signs of pulmonary arterial hypertension. RS -trachea central, apex beat - palpable (normal), bilateral chest movement symmetrical, bilateral air entry +, no added sounds. PA - soft, non-tender, no organomegaly, no bruit. CNS: HMF - normal. No sensorimotor deficits. No signs of meningeal irritation noted.

Radiological evaluation

USG abdomen with renal doppler: lobulated hypoechoic lesion with internal calcifications noted in right suprarenal region - likely to be adrenal mass. Renal artery doppler study - normal, i/v/o the above findings, CECT abdomen was done.

Figure 1(a and b): Bilateral papilledema.

Figure 2: Routine blood investigations.

Figure 3: (a) Plain CT abdomen and pelvis, (b) arterial phase, and (c) CECT abdomen and pelvis shows suprarenals.

Right side

A well-defined hypodense lesion measuring 38x36x51 mm (APXTSXCC) seen in the medial limb of right adrenal gland. The lesion shows central calcifications. Post contrast the lesion shows intense enhancement (pre-contrast HU 20-40, post contrast HU 120-140 HU in arterial phase) with poor washout (delayed phase 60- 80 HU). The lesion shows multiple non enhancing necrotic areas within.

Left side

Multiple lesions (4 in number) with similar enhancement characteristics are seen in the left adrenal region with
indistinct medial and lateral limbs and no evidence of calcifications. The lesion on left side shows loss of fat plane with left renal vein. Features are suggestive of solitary right adrenal and multiple enhancing left adrenal lesions as described above - possibly pheochromocytoma. In view of indistinct fat planes with left renal vein malignant changes cannot be ruled out.

**Biochemical analysis**

Plasma free metanephrines (liquid spectrometry with tandem mass spectrometry). Free metanephrines: 3.36 ng/l (7.90-88.70), free normetanephrines: 1540 ng/l (20.10-135.40), 3-methoxytyramine: 17.20 ng/l (<18.40). Syndromic evaluation of pheochromocytoma was done: (MEN 2). USG neck - normal thyroid gland. Serum calcitonin - 2.68 pg/ml (0-18.2) (VHL) CT brain (plain) - no evidence of any intracranial pathology. An I-131 MIBG scan was done, to look for extra adrenal tracer uptake.

![Figure 4: MIBG scan.](image)

**Procedure**: I-131 MIBG was administered by slow intravenous injection. Relevant drugs were stopped 2 days before. Patient was put on 10% dextrose fluids before and one hour after injection. Arterial and posterior views of the head, chest, abdomen, pelvic and umbilical obtained at 24 and 48 h post injection.

**Findings**: There are areas of intense concentration and retention in both left and right suprarenal area weighting 25.4mm on right side and 23.1mm on left side area symmetrically. (Left sided lesions scan separately in CT scan and are seen together due to limited resolution of gamma camera.)

**Impression**: There is no abnormal concentration elsewhere in the body. Normal concentration is seen in inferior glands, liver and spleen and normal excretion in urinary bladder.

**Conclusion**: Suggestive of MIBG avid supra renal tumours bilaterally: No evidence of abnormal concentration in the body.

![Figure 7: Gross specimen.](image)

Endocrinology and oncurosurgical opinion were sought and patient was planned for B/L adrenalectomy. After 1-week medication with alpha blockers, he was started on beta blocker tab Labetalol 100mg TID which was continued until surgery. Intermittently patient had orthostatic hypotension which was managed with adequate fluid intake and high sodium in diet. Bilateral adrenalectomy was performed with good intra op BP control. Right adrenals were enlarged and shows multiple nodular lesions, left adrenal noted shows nodular lesions.

![Figure 8: (a) Residual adrenal tissue and (b) tumor in Zellballen pattern.](image)

**Post-operative management**

Blood pressure continued to fluctuate despite adequate analgesia and complete tumor removal. Blood pressure was controlled with: tab Labetalol 100MG 1-1-1 and tab Nicardia Retard 20MG 1-0-1. Biopsy reports confirmed...
bilateral pheochromocytoma with adequate tumor free margins in the resected specimen.

With adequate BP control, he was discharged with: tab labetalol 100 mg (1-0-1) and tab nicardipine (R) 20 mg (1-0-1). After bilateral complete adrenalectomy, steroid replacement was started: tab wysolone 5 mg (1-0-0), with an advice to double steroid dose in case of any intercurrent illness.

DISCUSSION

Pheochromocytomas and paragangliomas are catecholamine-producing tumors derived from the sympathetic or parasympathetic nervous system. These tumors may arise sporadically or be inherited as features of MEN type 2, von Hippel-Lindau disease, or several other pheochromocytoma-associated syndromes (SDHx).

The prevalence of PPGL in patients with hypertension in general outpatient clinics is between 0.2-0.6%. In children with hypertension, prevalence is approximately 1.7%. About 5% of patients with incidentaloma on imaging are proven to have pheochromocytoma. Clinical importance is Tumors hyper-secrete catecholamines which can lead to cardiovascular, neurological morbidity and mortality. PPGLs enlarge and cause mass effect symptoms by encroaching upon or extending into adjacent area. Help in earlier diagnosis and treatment in other family members.

Preoperatively

Alpha blockade followed by beta blockers (phenoxbenzamine/prazocin/phenotolamine/metyrosine) (1-2) weeks before surgery. During surgery localized pheoadrenalectomy (laparoscopic/open) was done. Bilateral adrenalectomy (cortex preserving). Intra operatively I.V. nitroprusside (0.5-5 mcg/kg b.w), phenotolamine and nicardipine, severe hypotension. Post-operatively, rapid decrease in blood pressures. Hypotension: adequate fluid replacement with salt rich diet, hypoglycemia- hypoglycemia (due to removal of catecholamine suppression on insulin secretion).

CONCLUSION

Surgery remains the first line of management in malignant pheochromocytoma. Norepinephrine, epinephrine, and dopamine all act on their target receptors, which causes a physiological change in the body. Increase in catecholamines cause severe hypertension and multiple systemic complications like (cardiovascular, cerebrovascular events) and can lead to death if untreated.

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