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CASE REPORT

Pheochromocytoma Presenting with Upper GI Symptoms: A Case Report

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ABSTRACT

Pheochromocytomas are rare tumours originating in chromaffin cells, representing 0.1–1% of all secondary hypertension cases.[1] A case of pheochromocytoma in a 34 year old male presenting with unusual symptoms of abdominal discomfort and vomiting is presented. He has been generally well in the past but started feeling excessively tired for last 4 years and had dyspepsia off and on. On presentation his blood pressure was 240/120 mmHg, and heart rate was 130bpm. There was no neurological deficit and rest of the physical examination was normal. Fundoscopic examination revealed copper wiring and AV nipping but no hemorrhages or papilledema.The aim of this case report is to emphasize the importance of secondary causes of hypertension and to keep in mind the unusual presenting symptoms of pheochromocytoma.

Keywords: Gastrointestinal Disorders, Pheochromocytoma, Tumors

INTRODUCTION

Pheochromocytomas and paragangliomas are very rare tumors of the sympathetic nervous system that secrete catecholamines, noradrenaline and adrenaline, and their metabolites. These tumors may arise sporadically or may be associated with multiple endocrine neoplasia type 2, von Hippel–Lindau disease, or several other pheochromocytoma-associated syndromes.[2] More than 85% of cases of pheochromocytomas arise in adrenal medulla and 15% are extra-adrenal that arise elsewhere in the sympathetic chain anywhere from the base of the brain to the urinary bladder.[5] The classic "rule of tens" for pheochromocytomas states that ~10% are bilateral, 10% are extra adrenal, and 10% are malignant.[2]

Its clinical presentation is so variable that pheochromocytoma has been termed "the great masquerader". Among the presenting manifestations, episodes of palpitation, headache, and profuse sweating are typical, and these manifestations constitute a classic triad. The presence of all three manifestations in association with hypertension makes pheochromocytoma a likely diagnosis.[2] other unusual symptoms include abdominal pain, nausea, vomiting, dyspnea, anxiety and the fear of impending death, fatigue, pallor, weight loss, and tremors.

CASE REPORT

A 34 year old male with complains of abdominal discomfort and vomiting for last two weeks and 2 high blood pressure readings at his physician's clinic presented to Cardiology Department. He has been generally well in the past but started feeling excessively tired for last 4 years and had dyspepsia off and on. On presentation his blood pressure was 240/120 mmHg, and heart rate was 130bpm. There was no neurological deficit and rest of the physical examination was normal. Fundoscopic examination revealed copper wiring and AV nipping but no hemorrhages or papilledema. He was admitted to cardiology ward for blood pressure control and to assess the end organ damage and started on intravenous Isosorbide Dinitrate along with Amlodipine and Lisinopril.

Complete blood count and serum biochemistry were normal except for deranged urea and creatinine which settled within a week. ECG showed sinus tachycardia and hyperacute T waves but negative cardiac troponins. CT Brain plain revealed no haemorrhage or ischemia. Echocardiography was normal.

It was noticed that here was a marked variation in his blood pressure and pulse hour to hour (systolic B.P ranging from 70 to 260 mmHg, diastolic B.P ranging from 40 to 160 mmHg and pulse rate ranging from 80 to 150bpm) During the episode of hypertension, he seemed anxious, complained of palpitations, and was all sweaty, each episode lasting for less than an hour.

Ultrasound abdomen gave a suspicion of a suprarenal mass. CT Abdomen without contrast showed a well circumscribed left supra renal mass of 7.6x8 cm size, homogenous texture. Plasma Metanephrine and Normetanephrine were more than 10-fold high. 24 hour urinary Vanillylmandelic Acid was more than 3 fold high. Serum Aldosterone and Parathyroid hormone (PTH) were within the normal range.

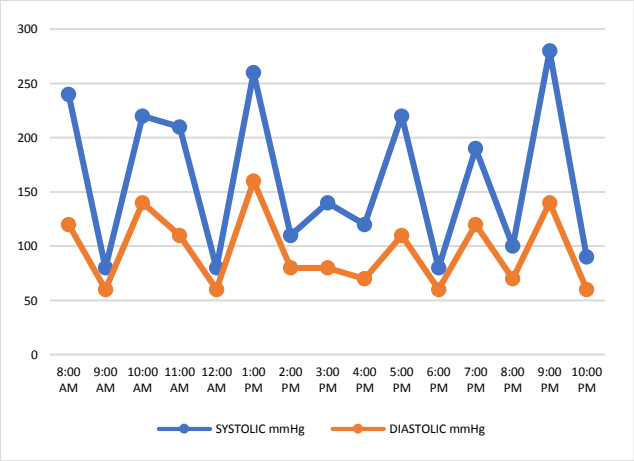

Figure 1: Patient's hourly blood pressure.

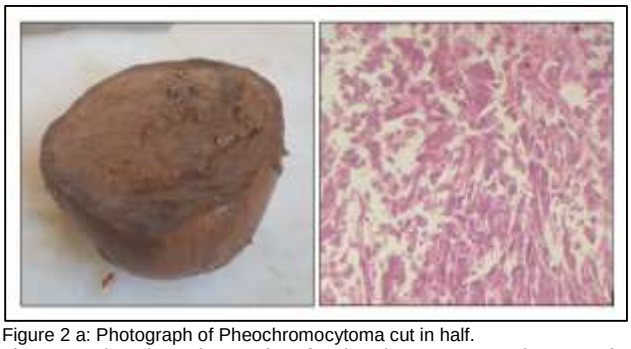

Figure 2 a: Photograph of Pheochromocytoma cut in half.
Figure 2 b Photomicrograph of Pheochromocytoma demonstrating characteristic 'zellballen' nests of cells

Alpha Blockade was achieved during the first week with Tab. Doxazocin 2mg x 8 hourly which along with calcium channel blocker controlled blood pressure to a point that he had a postural drop but tachycardia was still a problem. During the second week Tab Labetolol which has the properties of both alpha and beta

blockers was added to the treatment and it controlled the heart rate and other symptoms like palpitations and sweating as well.

Left Adrenalectomy performed and a single soft tissue mass measuring 7x7x4 cm and weighing 50 grams was removed with intact capsule. There was marked intra intraoperative fluctuation in blood pressure and 7 litres of crystalloids and 3 units of packed cell volume had to be transfused. On cut section the tumor was hemorrhagic, yellow and tan. Histopathology proved the diagnosis of pheochromocytoma.

The patient is doing reasonably well postoperatively with blood pressure readings within normal range.

DISCUSSION

Although Pheochromocytoma is a 'great masquerader', Hypertension is the key. Hypertension may be in paroxysms or may be sustained. The paroxysms or spells can occur spontaneously or be provoked by something. Among the precipitants are postural change, exercise, anxiety and drugs like beta blockers, metoclopramide and some anaesthetic agents. Anything that increases intraabdominal pressure can also precipitate a spell like change in position, lifting, defecation, exercise, colonoscopy, pregnancy, trauma. The types of spells experienced by patients are highly variable, however the spells tend to remain same for each patient. The frequency of pheochromocytoma spells vary from many times a day to once a month. The duration of a pheochromocytoma spell is also variable, ranging from few minutes to several hours, most commonly 15 to 20 minutes.[3] In this case the spells lasted for less than an hour.

A case report by Elden Baumgarten M.D says that many of the cases of so called 'Essential Hypertension' may actually be due to Pheochromocytomas which in the long run results in end organ damage and death so they prefer to consider this type of tumor as benign anatomically but malignant physiologically because it is the physiologic activity of this tumor that is malignant and not its anatomic consideration.[6] This emphasizes the importance of timely diagnosis and potential cure.

The cornerstone of diagnosis of pheochromocytoma and paraganglioma is biochemical confirmation of excessive catecholamines production and detecting the tumor on imaging. So if the physician is aware of the classical as well as non classical presentations of this neuroendocrine tumor and suspects it clinically, only then he can plan the workup investigations.[5] Tragically, nearly 50% of these tumors are discovered on autopsies so the clinicians must keep pheochromocytoma in mind whenever they encounter paroxysmal or even sustained hypertension with hyperadrenergic features.[8]

Surgical resection is the therapeutic goal but pre operative management is crucial. Pre operative management includes effective alpha blockade first and then beta blockade only in cases who have tachycardia or arrhythmias. The rationale behind this is that unopposed alpha blockade can precipitate hypertensive crises. Intra operative and post operative management need a team of anaesthesiologist and surgeons. There can be hypertensive crisis during intubation and tumor handling which may need intravenous phentolamine and sodium nitroprusside infusion. Intravenous Esmolol is used for intraoperative arrhythmias.[7] Intra operative and postoperative hypotension is managed with intravenous crystalloids as in this case.

Phaeochromocytomas have also been linked to the release of other hormones such neuropeptide A and endothelin-1 [9]. Although the functions of these hormones are unknown, they might contribute to the pathophysiology of gastrointestinal problems. Ileus develops as a result of a heavy tumour burden and excessive catecholamine release that lasts for a long time. Intermittent symptoms can be brought on by paroxysmal inhibition of the intestinal smooth muscles caused by intermittent catecholamine release [10].

CONCLUSION

Pheochromocytoma is one of the secondary causes of hypertension. It can be fatal by causing hypertensive emergency but if diagnosed, surgical removal is the definitive treatment in more than 90% of the cases.

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