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ADRENAL PHEOCHROMOCYTOMA: DIAGNOSTIC AND SURGICAL CHALLENGES – A CASE REPORT

General Surgery

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ABSTRACT

Pheochromocytomas are rare neuroendocrine tumors originating from chromaffin cells of the adrenal medulla, characterized by excessive catecholamine production. They constitute an uncommon cause of secondary hypertension, with only 50% of patients presenting with classical symptoms1. Here, we report the case of a 54-year-old male with a history of anxiety, hypertension, and palpitations, who was found to have an incidental left adrenal nodule. Confirming the diagnosis of pheochromocytoma, the patient underwent successful adrenalectomy following appropriate preoperative alpha- and beta-adrenergic blockade. This case underscores the importance of early recognition and management of pheochromocytoma to prevent associated morbidity and mortality.

KEYWORDS

Catecholamine secretion, phenoxybenzamine, pheochromocytoma, adrenal tumor, secondary hypertension

INTRODUCTION

Hypertension, defined as blood pressure exceeding 140/90 mmHg, is a leading cause of cardiovascular morbidity and mortality worldwide. Although the vast majority of cases are primary (essential) hypertension, approximately 3-10% are secondary to an identifiable underlying cause, some of which are potentially curable. Among these, pheochromocytomas, though rare, are critical to diagnose due to their potential for severe cardiovascular complications[2].

Pheochromocytomas have an estimated incidence of 0.4 to 9.5 cases per million people per year[3]. These tumors originate from catecholamine-secreting chromaffin cells, primarily located in the adrenal medulla. They can also arise extra-adrenally as paragangliomas. Clinical presentation is variable and may range from asymptomatic adrenal incidentalomas to hypertensive crises. The classic triad of headaches, palpitations, and excessive sweating suggests the diagnosis, but only half of affected individuals exhibit these symptoms. Biochemical confirmation of excessive catecholamine secretion, along with imaging studies, is necessary for diagnosis[4]. Definitive management involves surgical excision following adequate preoperative pharmacologic preparation to mitigate perioperative hemodynamic instability.

CASE PRESENTATION

A 54-year-old male presented to the outpatient department with recurrent abdominal pain. His medical history was significant for anxiety, hypertension, obesity, and palpitations. Additionally, he reported tremors and lower limb weakness. On physical examination, the patient appeared pale but had no palpable masses or other remarkable findings.

Laboratory investigations revealed urinary metanephrines of 204 µg/24h (normal <350 µg/24h). Contrast-enhanced computed tomography (CECT) of the abdomen identified a well-defined, hyperdense, solid, and avidly enhancing lesion measuring 59 x 51 x 50 mm in the left suprarenal region. The lesion exhibited internal areas of necrosis and derived its arterial supply from the middle suprarenal artery, draining into the left renal vein. It was closely related to the tail of the pancreas, upper pole of the spleen, and adjacent jejunal loops, though no overt infiltration was observed. Fat planes with the left kidney and stomach were preserved. No calcifications or fat attenuation were noted. The radiological findings were highly suggestive of pheochromocytoma.

A multidisciplinary team involving endocrinology and general surgery was consulted for treatment planning. The patient was started on phenoxybenzamine (10 mg/day) ten days prior to surgery to achieve adequate alpha-adrenergic blockade, followed by beta-adrenergic blockade to control reflex tachycardia. He subsequently underwent open adrenalectomy via a laparotomic approach. Histopathological examination revealed a well-encapsulated tumor with a classic Zellballen pattern and a rich vascular network, confirming the diagnosis of pheochromocytoma. There was no evidence of peri-adrenal adipose tissue invasion.

Postoperatively, the patient was monitored and no episodes of hypotension or hypoglycemia were documented. At six-month follow-up, ambulatory blood pressure monitoring was normal, and the patient remained asymptomatic.

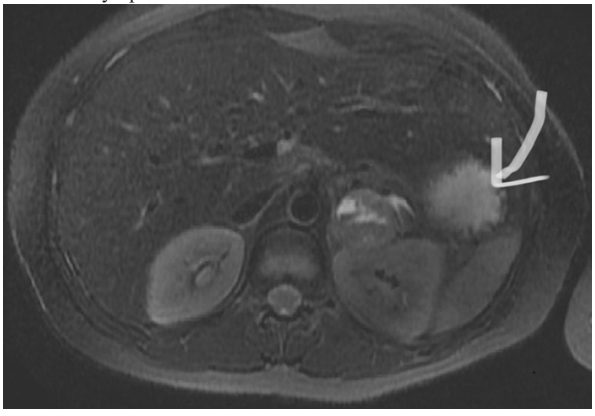

Fig-1.1

DISCUSSION

Pheochromocytomas are rare but clinically significant catecholamine-secreting tumors, accounting for 0.1-0.6% of cases of hypertension[5]. They typically present between the fourth and fifth decades of life and affect both genders equally[6]. Clinical manifestations range from asymptomatic adrenal incidentalomas to life-threatening hypertensive crises with potential cerebrovascular and cardiac complications.

Only 4% of adrenal incidentalomas are ultimately diagnosed as pheochromocytomas, with benign adrenal adenomas being far more common[7]. Approximately 50% of pheochromocytoma cases exhibit symptomatic paroxysmal hypertension due to excessive secretion of catecholamines such as epinephrine, norepinephrine, and dopamine[8].

Definitive diagnosis requires biochemical confirmation of excessive catecholamine release and imaging studies to localize the tumor. Additionally, genetic testing is recommended in all diagnosed cases, as 35-40% of pheochromocytomas are associated with hereditary syndromes, including von Hippel-Lindau (VHL) syndrome, multiple endocrine neoplasia type 2 (MEN2), and neurofibromatosis type 1 (NF1)[9]. Approximately 10% of pheochromocytomas are malignant, with local invasion or distant metastases being the only definitive indicators of malignancy[10].

Surgical resection remains the treatment of choice, achieving a cure in over 90% of patients[11]. Preoperative preparation is crucial to minimize perioperative morbidity and mortality. The Endocrine Society recommends alpha-adrenergic blockade, adequate fluid and salt intake, and subsequent beta-adrenergic blockade before surgery. Phenoxybenzamine is the preferred alpha-blocker, initiated at 10 mg every 6-12 hours and titrated up to 30-40 mg every 6 hours as needed. Beta-blockers should be introduced only after adequate alpha-blockade to prevent unopposed alpha-adrenergic vasoconstriction, which could precipitate a hypertensive crisis[12].

Postoperative complications include hypotension (occurring in 20-70% of cases) due to sudden catecholamine withdrawal[13]. Additionally, rebound hyperinsulinemia and depleted glycogen stores may lead to severe hypoglycemia, necessitating close monitoring of blood glucose and arterial pressure.

CONCLUSION

Pheochromocytoma, though rare, is a crucial differential diagnosis in patients with secondary hypertension. Timely recognition and appropriate perioperative management are essential for preventing life-threatening complications. Complete surgical resection, following optimal pharmacologic preparation, remains the cornerstone of treatment, offering excellent long-term outcomes. This case highlights the importance of considering pheochromocytoma in patients with suggestive clinical features and the critical role of multidisciplinary management in optimizing patient outcomes.

Conflicts Of Interest

The authors declare no conflicts of interest.

Ethics Statement

This study was conducted at Mathura Das Mathur Hospital, Jodhpur, Rajasthan, India. Informed consent was obtained from the patient for the publication of clinical data, and the study was conducted in accordance with Institutional Ethics Committee guidelines.

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