CASE REPORT

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Pheochromocytoma is an Eventful Tumor: A Case Description

[1]Bhargav PRK, [2]Amar V

[1]Assistant Professor, Department of Endocrine Surgery, Mamata Medical College and Hospital, Khammam, Andhra Pradesh, India

[2]Consultant General and Laparoscopic Surgeon, Mamata Medical College and Hospital, Khammam, Andhra Pradesh, India

Correspondence: Bhargav PRK, Assistant Professor, Department of Endocrine Surgery, Mamata Medical College and Hospital Khammam-507002, Andhra Pradesh, India, Phone: 08742-252010, Mobile: 09490130798, e-mail: kingbhargav@gmail.com

ABSTRACT

Pheochromocytoma is a tumor arising from adrenal medulla in > 90% of cases. It is a great mimic, featuring in differential diagnosis of disorders of multiple organ systems. Here we present a prototypical case of right adrenal pheochromocytoma, highlighting this unique feature of the tumor. It presented with repeated hypertensive crises with predominant involvement of one major organ system in each episode. All of them were reversible with timely intervention and successful removal of the tumor, highlighting the functional basis rather than organic pathology.

Keywords: Pheochromocytoma, Hypertensive crises, Adrenalectomy.

INTRODUCTION

Pheochromocytoma is a neuroendocrine tumor arising from adrenal medulla in more than > 90% of cases.[1] It is a great mimic with protean clinical manifestations.[2] To highlight this aspect, we present a case of right adrenal pheochromocytoma with a plethora of events during its clinical course.

CASE REPORT

A 37-year-old lady presented to us with history of palpitations, excess sweating and weight loss for 6 months. Blood pressure (BP) was 200/130 mmHg with postural drop to 180/110 mmHg. Pulse was bounding and 115/minute. Rest of the systemic examination was normal.

Patient was admitted with a diagnosis of secondary hypertension. Renal, drug intake, vasogenic, psychogenic causes were excluded. 24 hours urinary VMA value was 28.8 mg (7-11.1). Computerized tomogram showed a 6 × 4 × 4 cm heterodense right adrenal mass (Fig. 1). According to our departmental protocol, which is to give a trial of graduated alpha blockade for 10 to 14 days with maximum dose of 8 to 20 mg prazosin for adequate control in terms of Roizen's criteria before surgery. We add calcium channel blockers and ACE inhibitors if BP is refractory for most of the day.

Preoperative preparation was done with graduated antihypertensive medications with prazosin 15 mg/day (final dose), nifedipine 20 mg/day, enalapril 2.5 mg/day, high salt and fluid intake. There was symptomatic improvement and BP has reduced to 140/100 mmHg with mild fluctuations. We had reasonable control of BP for most of the day except during paroxysmal attacks of hypertensive crises as described subsequently.

On day 5 of admission, she complained of exertional dyspnea and cardiac evaluation revealed dilated cardiomyopathy with ejection fraction of 43%. On day 8, she developed sudden abdominal pain, distension and vomitings with maximum BP of 210/110 mmHg lasting for 1 day. Relieved with antiemetics, antispasmodics and potassium syrup. On day 10, she complained of vomitings, drenching sweats and had cold, cyanosed extremities with oliguria (300 ml urine output in 24 hours). Recovered with IV fluids and symptomatic treatment. On day 14, she developed tachypnea and was turning blue. She was intubated and put on ventilator. Evaluation revealed pulmonary edema, which improved with frusemide,

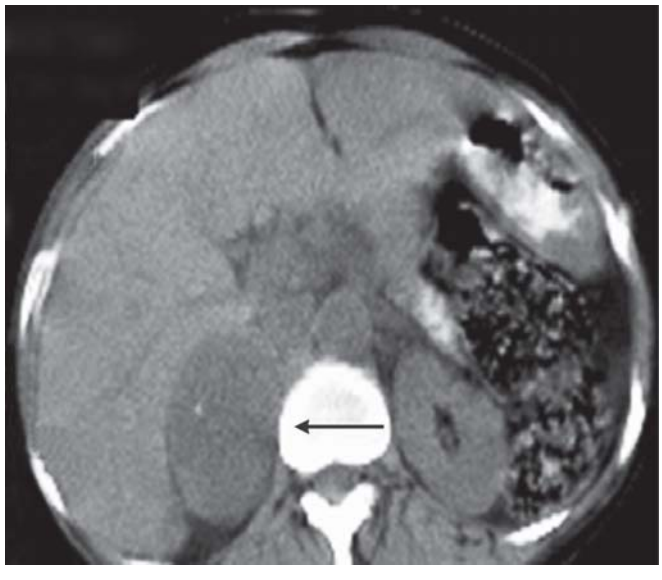

Fig. 1: CT abdomen showing a heterodense right adrenal tumor (arrow)

hydrocortisone and ventilation. On day 15, she developed convulsions and her consciousness deteriorated with bilateral dilated pupils. Diagnosed as hypertensive encephalopathy with maximum BP of 210/120 mmHg and put on mannitol, frusemide and hydrocortisone.

In view of repeated pheochromocytoma crises, she was taken up for surgery on day 16. We performed right open adrenalectomy and retrieved a 85 gm highly vascular tumor with characteristic intraoperative BP swings from maximum of 240/120 mmHg (tumor handling) to minimum of 70/45 mmHg (5 minutes after adrenal vein ligation). She recovered from hypotension after 15 hours with inotropic support. By 24 hours, she regained consciousness with pupils normally reacting to light. By 36 hours, she was extubated and breathing spontaneously. Histopathology was confirmatory of pheochromocytoma and at 6 months follow-up her BP was 130/80 mmHg.

All the events of morbidity are represented graphically in Figure 2.

DISCUSSION

Pheochromocytoma is known as great mimic featuring in many differential diagnoses.[3] Clinical course of pheochromocytoma is characterized by fluctuating BP with frequent paroxysms, though with alpha receptor blockade, their frequency is reduced.[4] Preoperative preparation of a pheochromocytoma patient can range from no alpha blockade to complete alpha blockade.[4,5] A majority of clinicians opt a protocol of antihypertensive medications with phenoxybenzamine or prazosin with or without other medications like calcium channel blockers or ACE inhibitors for a period of 1 week to 3 weeks. The criteria of adequate preparation is based on patient's improvement in symptoms with adequate hydration and hematocrit increase. A stringent BP control is not mandatory as intraoperative BP fluctuations can occur even with complete blockade. A short course of preoperative preparation depending on patient's comorbid conditions suffices before sugery.

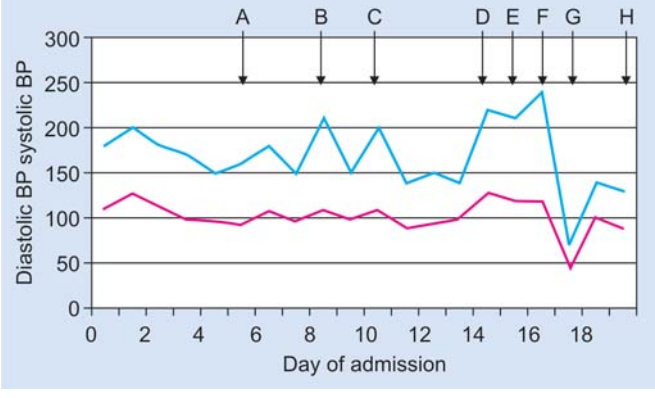

Fig. 2: Curve showing disease course with clinical events: (A) cardiac event; (B) gastrointestinal event; (C) renal, fluid and electrolyte; (D) pulmonary event; (E) cerebral event; (F) surgery; (G) postoperative hypotension; (H) normalization of BP

Pheochromocytoma crisis can be defined as a paroxysmal attack of hypertensive crisis usually presenting with nonspecific hyperadrenergic clinical features and often associated with a major organ system involvement, which is mostly reversible with timely treatment. If multiple organ systems are involved with impending doom simulating septicemia with lactic acidosis, it is called PMC (Pheochromocytoma multisystem crisis), which carries very poor prognosis.[6] Clinical manifestations are protean and highly nonspecific affecting any organ system.[2] Accordingly, this patient had repeated hypertensive crises involving an organ system. She had dilated cardiomyopathy, which is reversible in early stages being a specific entity of catecholamine induced cardiomyopathy.[7] She suffered from abdominal distension on day 8, due to $\alpha_{1,2}$-receptor mediated sphincteric contraction and β_2-receptor mediated gut muscle relaxation.[8] It is a self-limiting morbidity with no organic pathology, as seen in our case with complete improvement within 24 hours.

On day 10, she was dehydrated due to vomitings, excess sweating with cold, clammy, cyanosed extremities (Raynaund's phenomenon) leading to oliguria, but improved with parenteral fluids. Cold and cyanosed extremities may be due to intense vasospasm caused by excess catecholamines.[9] On day 14, she had pulmonary edema, probably due to pulmonary vascular instability caused by rapid and wide BP fluctuations. On day 15, the patient lost consciousness with pupillary dilatation due to hypertensive encephalopathy, but improved with mannitol and hydrocortisone. Hypertensive encephalopathy with reversible multifocal cerebral edema in pheochromocytoma is already reported.[10] Inspite of adequate treatment, repeated self-limiting paroxysms and labile hypertension could not be prevented. Only surgical removal of tumor lead to alleviation of paroxysms.

At surgery, experienced anesthetic team with all vasoactive drugs are crucial for success. Open surgery is usually preferred for familial, multifocal, large (> 6 cm) and malignant pheochromocytomas. Majority of other pheochromocytomas are operable laparoscopically. Opposite adrenal and retroperitoneum should be inspected and palpated to rule out multifocal tumors and lymph nodes, even in apparently sporadic tumors.

The important lesson learnt from this patient apart from protean manifestations of multiple organ systems was that most of them are reversible, if monitored and treated symptomatically, as they have functional basis rather than organic pathology in the end organ.

We highlighted the typical and striking feature of pheochromocytoma through this case, i.e. a clinical course characterized by unpredictable events involving multiple organ systems at various intervals, yet completely reversible with proper and timely treatment.

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