Brief Communication

A case of normotensive pheochromocytoma with management dilemma

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ABSTRACT

Introduction: We report an unusual case of normotensive pheochromocytoma detected incidentally, presenting a pre-operative management problem. **Case Note:** A 40-year-old lady with vague abdominal symptoms was initially discovered with a left adrenal incidentaloma by ultrasound abdomen, which was also revealed in computed tomography (CT). After exclusion of all the causes with possible necessary investigations, pheochromocytoma was confirmed with elevated 24 hour urinary metanephrine and normetanephrine. Her blood pressure was in low to normotensive range all throughout. She was attempted to be prepared with combined alpha and beta blockade but could not tolerate this regimen due to symptomatic hypotension. Subsequently, surgical preparation was planned cautiously with alpha-adenergic blockade only. With intensive monitoring, she underwent uneventful left adrenalectomy, and surgical pathology was consistent with pheochromocytoma. **Conclusion:** This case illustrates an unusual presentation of normotensive pheochromocytoma as adrenal incidentaloma. Pre-operative preparation in these patients can be achieved with alpha-adrenergic blockade, adequate hydration, and liberal salt intake.

Key words: Normotensive pheochromocytoma, adrenal incidentaloma, preoperative preparation

Introduction

Although hypertension, sustained or paroxysmal, is usually a cardinal feature of pheochromocytoma, normotensive presentation is unusual.[1] Symptoms of orthostatic hypotension may dominate the presentation in patients with epinephrine- or dopamine-predominant tumors.[1,2] In this case report, we describe a normotensive patient with pheochromocytoma who initially presented as adrenal incidentaloma and the management dilemma regarding this case.

Case Report

A 40-year-old, menstruating, mother of one child, was referred to the outpatient department of Endocrinology of N.R.S Medical College and Hospital, Kolkata, with an ultrasound abdomen report suggestive of enlarged left adrenal gland. On enquiry, she gave history of vague abdominal discomfort for prolonged duration without any other positive complaints. Her blood pressure (recorded on multiple occasions) was around 110/70 mmHg with pulse rate 76-84/min in sitting and 106/70 mmHg with pulse rate 82-90/min in standing position. She did not have any features of marfanoid habitus or any evidence of mucosal neuroma, café-au-lait spots, or any features of hyperandrogenism. General physical and systemic examinations were non-contributory.

Her routine blood investigations (complete hemogram, blood sugar, urea, creatinine, urine routine examination, E.C.G, chest X-ray) were within normal limits. Subsequently, computed tomography abdomen also confirmed a left adrenal adenoma with dimension of 48 mm × 47 mm, round shape, smooth contour with sharp margin [Figure 1]. All necessary biochemical investigations were done to rule out the differential causes. Her thyroid function tests were within normal limits. Serum cortisol (8am) and dexamethasone suppression test did not reveal any abnormality. Her electrolytes levels were: Sodium-140 meq/l, potassium-4.6

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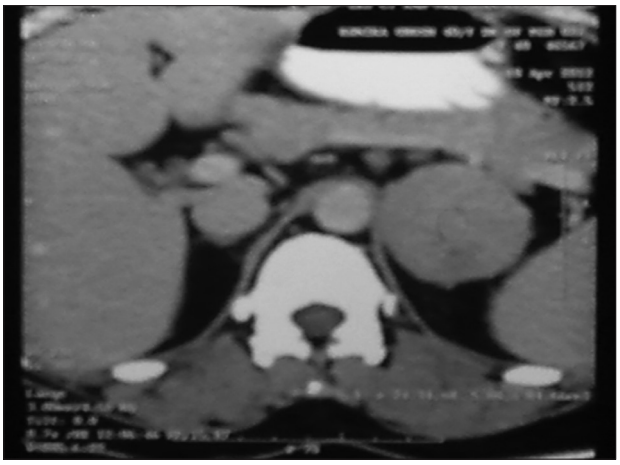

Figure 1: Computed tomography abdomen showing left adrenal adenoma (48 mm × 47 mm)

meq/l, calcium-9 meq/l. Total protein level was -7 gm%, and serum albumin level was 4.6 gm%. Urinary metanephrine and normetanephrine levels were: 6986.56 μg/24 hrs (8963.33 μg/gm of creatinine) and 7496.50 μg/24 hrs (9617.56 μg /gm of creatinine), respectively. Having thus diagnosed, left adrenal pheochromocytoma, the patient was being prepared for surgery with volume repletion and administration of extended release prazosin (2.5 mg) at night (since phenoxybenzamine was not available) and propranolol (30 mg) in divided doses.

However, she had symptomatic hypotension with this combination so much so that cardiologist was keen on stopping both the anti-hypertensives, and the anesthesiologist wanted to withhold prazosin. She was cautiously put on extended release prazosin (2.5 mg) only with intensive monitoring as a preparation for surgery. Subsequently, she underwent an open left adrenalectomy under general anesthesia, which she tolerated well. The tumor was well-encapsulated, and histopathology also confirmed the diagnosis of pheochromocytoma.

Discussion

All patients with adrenal incidentaloma should be investigated for pheochromocytoma, even when asymptomatic.[3] Though hypertension (attributed to norepinephrine secretion) – episodic or sustained is common, hypo to normotensive presentation, as a possibility, should also be kept in mind. In 2008, Gonzalez-Pantaleon and Simon presented a case of normotensive pheochromocytoma along with its challenges in diagnosis and management.[4]

The intra-operative mortality in patients with unsuspected pheochromocytoma may approach -50%. As a result, extreme care in the pre-operative preparation and the intra-operative management should be taken.[5] In normotensive patients, pre-operative management is more controversial.[5] For example, our patient developed symptomatic hypotension during combination (alpha- and beta-blocker) therapy but tolerated well the low-dose alpha-blocker therapy only.

The spectrum of the presentation of pheochromocytoma continues to expand. Hypertension may be absent despite excess norepinephrine secretion. Again, in absence of hypertension, use of combination (alpha- and beta-blocker) therapy may cause precipitous fall in blood pressure and, therefore, these agents should be used cautiously in such patients.

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