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RESEARCH ARTICLE

A CASE REPORT OF A GIANT ADRENAL TUMOR; A DELAYED DIAGNOSIS OR MISSED DIAGNOSIS

*Kunal Singh

India

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ABSTRACT

Pheochromocytoma is a rare catecholamine secreting tumor originating usually from adrenal medulla and produces signs and symptoms due excessive catecholamine secretion from tumor. A young male patient of 32 year age presented with febrile episodes, dullache on left upper quadrant of abdomen, episodic headache and weakness for last 2 months. Clinical suspicion of pheochromocytoma was confirmed by vitals monitoring and CT scan of abdomen. After having two weeks of preoperative preparation with alpha blocker and beta-blocker, open surgical removal of pheochromocytoma was done. Preoperative fluctuation of BP was well managed by IV fluid overload, intravenous Phentolamine, intravenous Esmolol and intravenous Ephedrine. Postoperative recovery was uneventful and BP regains to normal range from 1st postoperative day. Pheochromocytoma is a rare cause of hypertension. If the diagnosis of pheochromocytoma is delayed or missed, the consequences could be disastrous, even fatal; however, if a pheochromocytoma is identified, it is potentially curable, as being one of the causes of surgically correctable hypertension.

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INTRODUCTION

Pheochromocytoma is a rare tumor originating from catecholamine secreting chromaffin cells that are derived from the ectodermic neural system and mostly situated within the adrenal medulla (Neumann, 2008). It was suggested that most doctors will meet only 1 patient with a pheochromocytoma in their working life time and a large general hospital will admit on an average 1 patient per year (Karet, 1947). Because of excessive catecholamine secretion, pheochromocytoma may precipitate life-threatening hypertension or cardiac arrhythmias. Pheochromocytoma is fascinating and challenging to clinicians because it combines lethal potential if untreated with possible long term cure in the majority if diagnosed and treated surgically. We present a case report of adrenal pheochromocytoma of 32 year aged young male patient presenting with febrile episodes and paroxysmal attacks of hypertension, which was treated successfully by open surgical removal of pheochromocytoma.

Case Presentation: A 32 years old male, height: 152 cm; weight: 74 kg, admitted to our Hospital with the complaints of febrile episodes, dullache in upper quadrant of abdomen, episodic headache and weakness, anxiety for last 5 months (Figure 1).

Initial evaluation and management done in primary centre. During evaluation B.P elevated (Systolic BP 150 mmHg and diastolic BP varies from 100 mmHg) in multiple recording and ultrasound suggested abscess cavity in supra renal region . Patients symptoms persisted and finally referred to our centre . During hospital stay vitals charting suggested variable and fluctuating reading, tachycardia (range 110 to 140), Blood Pressure (BP) is high (Systolic BP varies from 160 mmHg to 180 mmHg and diastolic BP varies from 100 mmHg to 120 mmHg), afebrile. . On examination, patient had no palpable mass, mild tenderness in left upper quadrant, flank within normal limit . Complete blood count had elevated ESR 125, Random Blood Sugar raised 191 mg/dl, HB1AC 8.3%, PT/INR raised 19.08/1.32. KFT, LFT, routine urine examination, Chest X-rays and ECG reports were within normal limit. CT (Computed Tomography) scan of abdomen shows left sided adrenal mass of a size about 10.2 cm x7.6 x7.8 cm heterogeneously enhancing without calcifications and cystic degenerations, that shifts tail of pancreas, superior pole of right kidney and spleen, countor and flat plane intact without suggestive of invasion. We made a provisional diagnosis of secondary hypertension due to adrenal pheochromocytoma but for confirming this diagnosis we need to do biochemical investigations. LD DST serum cortisol (8.65ug/ml) and 24 hour urinary cortisol (16.2 ug/24 hr) were normal but serum free metanephrine raised (192.6 pg/ml) and 24 hour fractionated metanephrine (189.45 ug / 24 hr). Plasma aldosterone were (13.3 ng/dl), aldosterone and plasma renin ratio (2.10).

*Corresponding author: Singh Kunal
India

DISCUSSION

Pheochromocytomas occur in people irrespective of races, equal incidence in male and female (Neumann, 2008). The peak incidence, however, is between the third and the fifth decades of life.

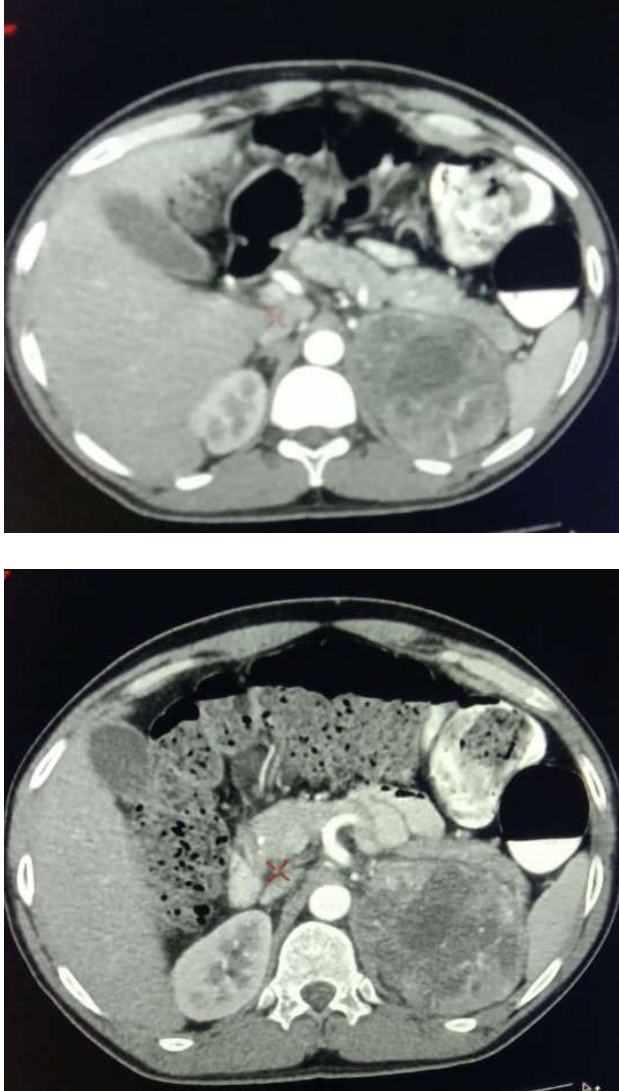


Fig. 1. Axial /Transverse image of left adrenal tumor

Approximately 10% occur in children. The majority of cases are sporadic, with only 16% having a history of associated endocrine disorder such as Multiple Endocrine Neoplasia type II (MEN IIA and IIB), Neurofibromatosis 1 (NF1) and von Hippel- Lindau disease (VHL) (Karet, 1994). Approximately 10% of pheochromocytomas are malignant (Karet, 1994). Direct invasion of surrounding tissue or the presence of metastases determines malignancy. Unfortunately, no reliable clinical, biochemical or histological features distinguish a malignant from a benign pheochromocytoma. The clinical manifestations of a pheochromocytoma results from excessive catecholamine secretion by tumor. Catecholamine typically secreted, either intermittently or continuously, includes norepinephrine and epinephrine; rarely dopamine is secreted. The biological effects of catecholamines are well known. Catecholamine secretion in pheochromocytoma is not regulated in the same manner as in healthy adrenal tissue. Relative catecholamine levels also differ in pheochromocytoma.

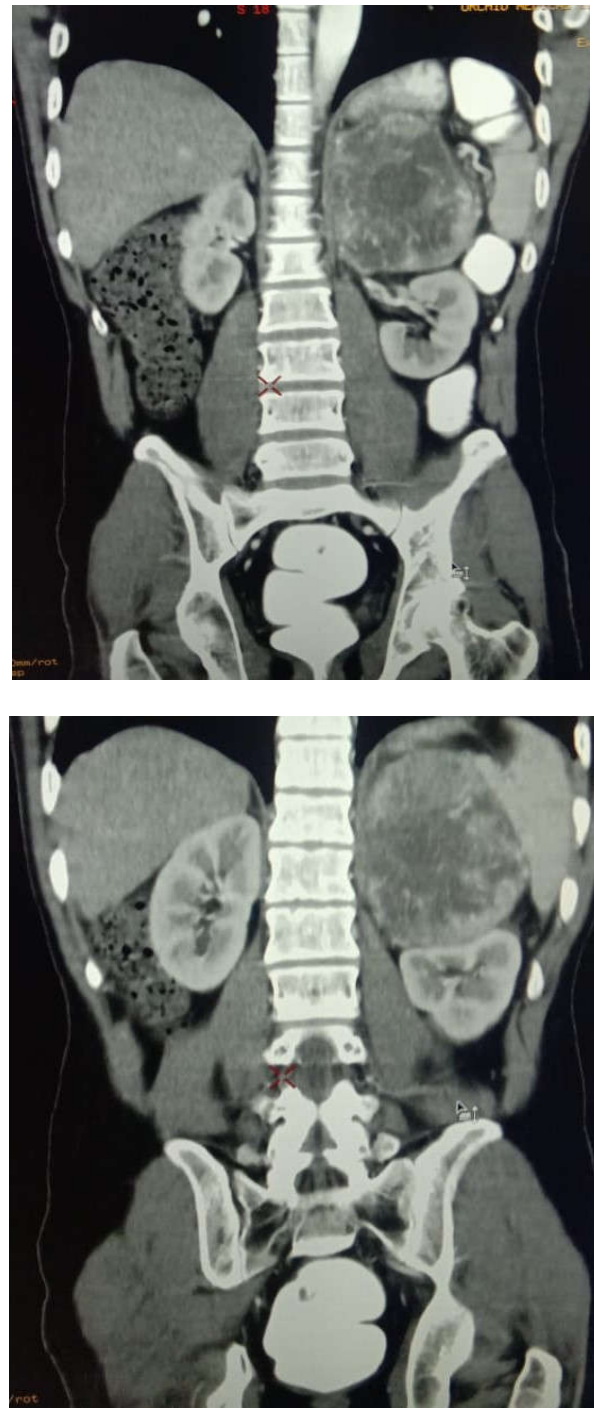


Fig 2. Longitudnal/coronal image of left adrenal tumor

Most pheochromocytomas secrete norepinephrine predominantly, whereas secretions from normal adrenal medulla are composed of 85% epinephrine (Yong, 2007). The classic history of a patient with pheochromocytoma includes spells (Paroxysms) characterized by headaches, palpitations and diaphoresis in association with severe hypertension. The spells may vary in occurrence from monthly to several times per day and the duration may vary from seconds to hours. Giant pheochromocytomas (>7 cm in size) are rare entities with around few cases reported in the literature (Bentrem, 2002). They do not present with the classical symptoms of pheochromocytomas (Bravo, 1993). Most patients present with vague discomfort while others may complain of a palpable abdominal mass. Operative surgery is the ideal management option (Sheps, 1990).

There needs to be a multidisciplinary approach while managing such cases. The first step in the diagnosis of a pheochromocytoma is the biochemical confirmation of catecholamine excess. Plasma metanephrine testing has the highest sensitivity (96%) for detecting a pheochromocytoma, but it lower specificity (Rothmund *et al.*, 2008). In comparison, a 24 hour urinary collection for catecholamines and metanephrines has a sensitivity of 87.5% and a specificity of 99.7% (Gifford, 1964). The biochemical diagnosis is followed by the localization of the pheochromocytoma and/or metastases. Magnetic Resonance Imaging (MRI) is preferred over Computed Tomography (CT) scanning because contrast media used for CT scans can provoke paroxysms. In addition MRI has a reported sensitivity of up to 100% in detecting adrenal pheochromocytoma (Neumann, 2008).

I-MIBG (Iodine-131 labeled metaiodobenzylguanide) scanning is reserved for cases in which a pheochromocytoma is confirmed biochemically but CT scan or MRI does not show a tumor (Neumann, 2008). Surgical resection of the tumor is the treatment of choice and usually results in cure of hypertension. Careful preoperative preparation requires with combined alpha and beta blockade to control blood pressure and to prevent intraoperative hypertensive crisis (Drenou, 1995). Alpha-adrenergic blockade, in particular, is required to control blood pressure and prevent a hypertensive crisis. Phenoxybenzamine is the preferred alpha blocker in preparation for surgery. A dose of 20 mg of phenoxybenzamine initially, should be increased daily by 10 mg until a daily dose of 100 mg to 160 mg is achieved and the patient reports symptomatic postural hypotension (Sarveswaran, 2015). Others alpha blocker such as Doxazosin, Prazosin and Terazosin are only rarely used because of their incomplete alpha blockade. Additional beta-blocked is required if tachycardia or arrhythmias develop; this should not be introduced until the patient is alpha blocked because unopposed alpha adrenergic receptor stimulation can precipitate hypertensive crisis (Karet, 1994). Non cardioselective beta blockers, such as Propranolol or Nadolol are often used; however, cardioselective agents, such as Atenolol and Metoprolol, also may be used.

Conclusion

Pheochromocytoma is often called 10% tumor because 10 percent are bilateral, malignant, extra adrenal, multiple, familial and occur in children (Duh, 2010). Pheochromocytoma is one of the few causes of hypertension that can be treated surgically.

Although it is the causative factor of hypertension in about 0.1% to 0.6% of the hypertensive population, detection is mandatory, not only for the potential cure of the hypertension but also to avoid the potentially lethal effects of the unrecognized tumor (Lo *et al.*, 2000).

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