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Bilateral adrenal pheochromocytoma: a case report

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ABSTRACT

Pheochromocytoma (PCC) is a tumor of catecholamines producing cells of adrenal medulla. Approximately 1 to 2 per 100,000 Individuals are diagnosed annually with PCC. Individual with PCC presents with signs and symptoms of sympathetic overstimulation. Only 10% of PCC's cases were found to be bilateral. Though bilateral PCC is more often a part of a familial syndrome, in this case, we report bilateral PCC with no evidence of familial disorders that correlates with this presentation. We report a case of a 13-years-old female presenting with adrenoceptor overstimulation manifested as uncontrolled hypertension which progressed to hypertensive encephalopathy precipitated by Bilateral Adrenal PCC.

KEYWORDS Pheochromocytoma(PCC), Bilateral pheochromocytoma, Uncontrolled hypertension

INTRODUCTION:

Pheochromocytoma, (PCC) is a tumor of catecholamines producing cells of adrenal medulla. The reported incidence of PCC is about 2 to 8 cases per million [1,2]. Individuals with PCC, present with signs and symptoms of sympathetic overstimultation. Awareness of the classic triad of tachycardia, episodic headache and sweating, is a key to early recognition of this condition [3]. In this article, we report a case of a 13-years-old female presenting with hypertensive encephalopathy precipitated by Bilateral Pheochromocytoma.

CASE STUDY:

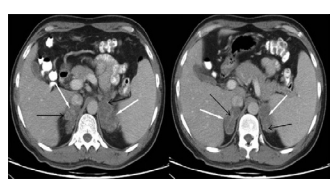
A 13-years-old female patient presented to the Emergency Department at Vadilal Sarabhai General, Hospital Ahmedabad with history of headache, abdominal pain, vomiting and tiredness since 1 month. There was no history of photophobia or trauma. On physical examination, it was found that patient had blood pressure readings around 180/90mmHg. Full Septic screen was negative. Complete Blood Count, Serum Calcium, Phosphate, Parathyroid Hormone, Thyroid Function tests, Serum Urea & Electrolytes were within normal limits. While, high blood pressure was managed by nifedipine and IV infusion of labetalol.

Ultrasound Abdomen: 49x35 mm sized mixed echogenic lesion with calcification within it was noted in left hypochondrium region in retroperitoneum.Suggesting possibilty of Adrenal origin.

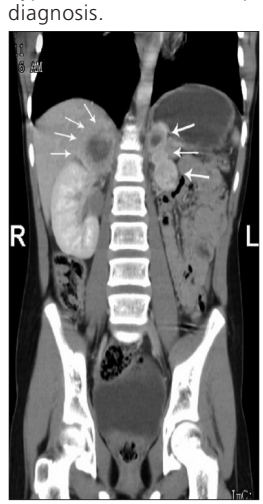
CT Abdomen: Neoplastic lesions involving bilateral Suprarenal glands

(left: 44x48x36mm and right :22x27x19mm)

Possibility of Bilateral Pheochromocytoma.



CT- Abdomen (Transverse section of the abdomen): Heterogenously enhanced adrenal lesions with central hypodensities and with giving clinical history of uncontrolled hypertension, bilateral pheochromocytoma is most likely diagnosis.



CT Abdomen showing Bilateral enlarged suprarenal masses, the right suprarenal mass measuring 22x27x19mm, the left suprarenal mass measuring 44x48x36mm. Both kidneys has normal size and cortical density.

MRI Brain : Suggestive of Hypertensive Encephalopathy and Demyelination.

With the given clinical history of uncontrolled hypertension, a provisional diagnosis of Hypertensive Encephalopathy secondary to Adrenal Pheochromocytoma was made. For confirmation 24 hour urinary collection for catecholamines revealed high levels compatible with pheochromocytoma. MIBG Scan showed high uptake in supra-renal glands in both sides with absence of metastasis. Surgery was scheduled and patient was given Prazosin (an alpha blocker) preoperatively, starting with 2.5 mg Q6 hours, Amlodipine 10 mg once daily and Atenolol (a beta blocker) 20 mg twice daily till the morning of the surgery. Blood pressure was recorded around 110/70mmHg. Bilateral Adrenal Cortex sparing Adrenalectomy was done via laparotomy and was sent for hisopathological examination .

Histopathology report revealed well circumscribed neoplastic growth composed of well-defined nets bounded by delicate fibrovascular stroma. The cells were polygonal with oval to round nuclei, prominent nucleoli and granular cytoplasm. The histopathology report confirmed the diagnosis of Bilateral Pheochromocytoma. In addition, no steroids were required.

Regular follow up showed fast recovery with complete resolution of hypertension. No steroids were administerd.

DISCUSSION:

Pheochromocytoma (PCC) is a rare tumor, originating from the suprarenal medulla and sympathoadrenal neuroendocrine system. The reported incidence of PCC is about 2 to 8 cases per million [1,2]. 10 to 20% of these cases occur in pediatric age group [3]. Recently it was found that about (6.7%) of patients with PCC were associated with familial syndromes [5]. Patients with positive genetic mutation were likely to be young at presentation, have bilateral disease [7]. In pediatric age group, bilateral pheochromocytoma is more often part of a familial syndrome [8,9], such as neuro-fibromatosis, von Hippel-Lindau disease and as a component of multiple endocrine neoplasia (MEN 2). Adrenal pheochromocytomas are most often found on the right side and are unilateral, sporadic and benign. 10% of PCC's cases were found to be Bilateral.

These tumors produce and secrete catecholamines (epinephrine, nor epinephrine and dopamine). Due to the significant rise in catecholamine levels, patients having PCC present with various symptoms and signs of adrenoceptor overstimulation. Therefore, sustained or paroxysmal Hypertension, palpitation, diaphoresis and headache strongly suggest the presence of PCC, with a 94% specificity and 91% sensitivity [4]. Malignant hypertension characterized by symptomatic elevations of blood pressure can be a clinical feature of malignant pheochromocytoma [6]. Most PCC are localized within the abdomen and in about 75% to 90% of the cases it's adrenal in origin [4].

Around 25% of all PCC cases are discovered incidentally during imaging studies for unrelated disorders [2]. In addition to clinical presentation, preoperative diagnosis is usually made by significantly high levels of catecholamines and their metabolites in blood and urine. 24-hour urine collection for urine catecholamines is both sensitive and specific.[3] Recent studies show a higher sensitivity for the determination of 24-h urine normetanephrine [5].

Computed tomography scan and magnetic resonance imaging are used in diagnosing and localizing PCC, with a sensitivity of 75% to 100%, though with low specificity. The (131-I-MIBG), 131-I- Metaiodobenzylguanidine gammagraphy, in combination with platelet normetanephrin, has (100%) sensitivity. In comparison to 131-I-MIBG, PET scan, Positron emission tomography scan has a better image resolution and using tracer (18-fluoro-dihydroxyphenylalanine), its sensitivity reaches about 100% [6].

Definitive treatment of PCC is surgical resection of the mass which is considered to be curative in 90% of the cases [7]. Anesthesia preparation can be difficult in terms of maintaining stable hemodynamic in patients with PCC. The patient's status should be optimized, blood pressure controlled and blood volume repletion are accomplished in order to prepare the patient for the operation. In order to prevent hypertensive crisis, Alpha-adrenergic blockade, in particular, is used for BP control. An initial dose of 20 mg of phenoxybenzamine is given to the patient with daily increase by 10 mg until 160 mg is reached, aiming to orthostatic hypotension [8]. In cases where the patient has tachycardia or arrhythmias, a beta blocker drug should be added but it should not be started until the patient has received an alpha blocker in order to avoid hypertensive crisis caused by unopposed alpha adrenergic receptor stimulation [9].

Nowadays, laparoscopic adrenalectomy, has been utilized frequently for the surgical treatment for both adrenal and extra-adrenal. After total bilateral adrenalectomy, patients will require prolonged steroid replacement. To avoid morbidity associated with steroid treatment, adrenal sparing surgery has been utilized successful in the management of bilateral PCC.

As in the case we are reporting, adrenal cortex was spared, bilaterally so no steroids were used in the postoperative period. Adrenocortical function was preserved and patient did not require steroid in post-operative period following the operation, Biochemical tests (24-h urinary catecholamine) should be performed and lifelong follow up of urinary catecholamine levels should be performed to identify recurrent or metachronous pheochromocytoma.

CONCLUSION:

Although PCC is a rare neoplasm, it should be considered as a possible diagnosis in children presenting with malignant hypertension. Genetic tests should be done in patients with pheochromocytoma who are young, have a positive family history, and with bilateral disease. Patients must be treated properly with antihypertensive

medications in order to stabilize blood pressure before surgery. Adrenal sparing surgery has been utilized successfully and most patients will not need steroid treatment afterward.

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