https://doi.org/10.22502/jlmc.v1i1.17

Case Report

Pheochromocytoma: a Rare Cause of Secondary Hypertension

Sahadeb Prasad Dhungana,[a,b] GP Acharya,[a,b] RM Piryani,[a,b] SK Shrestha[a,b]

ABSTRACT:

Introduction: Pheochromocytoma is a rare tumor, originating from the chromaffin tissue. Its frequency is approximately 1/100,000. The clinical manifestation is due to catecholamine excess, which includes high blood pressure, palpitation, headache, sweating, nausea, vomiting, trembling, weakness, irritation, abdominal and chest pain, dyspnea, red warm face, constipation, polyuria, and polydipsia. **Case Report**: We present a case of 53 years old male, hospitalized for hypertensive crisis following the manipulation of left sided frozen shoulder. He had labile blood pressure ranging from 220/120 systolic- 90/60 diastolic, profuse sweating and tachycardia. Findings of Contrast enhanced CT of abdomen was consistent with right adrenal pheochromocytoma and 24 hours urinary VMA was 17 mg /24 hr (Normal <13.6 mg /24hr). After the clinical, paraclinical investigations and radiological tests, it was proved to be a pheochromocytoma. The surgical intervention was planned. But due to unavailability of required antihypertensive drugs in Nepal (alpha-blockers like phentolamine and phenoxybenzamine), surgeons were reluctant to operate, although blood pressure was well controlled with use of sodium nitroprusside during hypertensive crisis and prazosin, a selective alpha blocker as maintainance therapy. The use of prazosin to control hypertension secondary to pheochromocytoma is limited to case report and case series. **Conclusion**: Although rare, pheochromocytoma is a treatable surgical cause of secondary hypertension.

Keywords: adrenal gland • pheochromocytoma • secondary hypertension.

INTRODUCTION:

The term "pheochromocytoma" has been classically used to describe an adrenal functioning (epinephrine and norepinephrine- secreting) paraganglioma, but it has also been applied to extra-adrenal functioning paraganglionomas.[1-2] The pheochromocytoma usually is benign, but approximately 10 per cent are malignant. The "rule of 10" used to describe pheochromocytomas is a good example: 10 percent are extra-adrenal, and of those, 10 percent are extraabdominal; 10 percent are malignant; 10 percent are found in patients who do not have hypertension; and finally, 10 percent are hereditary.[3] Familial pheochromocytoma is inherited as an autosomal dominant trait alone or as a component of the multiple endocrine neoplasia type 2 syndromes (MEN-2A and MEN-2B), von Hippel–Lindau disease or, in rare cases, neurofibromatosis type 1.[4] The remaining 90 percent of pheochromocytomas are classified as sporadic or nonsyndromic.

CASE REPORT:

A 53 years male was found to have raised blood pressure months months back and had taking tab amlodipine 10 mg OD, losartan 50 mg OD since then. He was diagnosed as having diabetes mellitus nine months back and taking tab Glimepiride 2 mg OD. He had acute kidney injury eight months back and received four sessions of hemodialysis. He

a - Lecturer
b - Department of Medicine,
Kist Medical College Teaching Hospital, Lalitpur, Nepal

Corresponding Author:
Dr. Sahadeb Prasad Dhungana,
e-mail: spdhungana@rediffmail.com

How to cite this article:
Dhungana SP, Acharya GP, Piryani RM, Shrestha SK. Pheochromocytoma: a rare cause of secondary hypertension. Journal of Lumbini Medical College. 2013;1(1):48-50. doi:10.22502/jlmc.v1i1.17.

presented to emergency department with complaints of headache, sweating, palpitation, vertigo for one day following manipulation of left frozen shoulder. Family history was insignificant. On examination, he had labile blood pressure ranging from 220/120 systolic- 90/60 diastolic, profuse sweating and tachycardia. Laboratory investigations revealed Hemoglobin 13.3 gm/dl, Total leucocytes 11,500/mm3 (Neutrophil 75%, Lymphocytes 20%, Monocytes 3%, Eosinophils 2%), Fasting blood sugar- 98 mg/dl, Urea- 37 mg/dl, Creatinine-1.1 mg/dl, sodium-138 meq/dl, potassium-4.4 meq/dl, Calcium- 8.3 mg/dl, Phosphorus- 3.6 mg/dl, Urine examination-trace sugar, Total protein- 7.8 gm/l , albumin -3.4 gm/l, Lipid profile (mg/ dl)- cholesterol 260, HDL cholesterol 42, LDL cholesterol 184, Triglycerides 170, Electrocardiogram- normal, 24 hr urinary VMA - 17 mg/24 hr (Normal <13.6 mg/24hr). 2D echocardiography - normal, Ultrasound of thyroid and parathyroid glands were normal. Ultrasound abdomen showed well defined heterogeneously echogenic mass measuring 4x5 cm with small cystic changes seen in right suprarenal area suggestive of adrenal mass ?pheochromocytoma, left renal cortical cyst. (Fig.1). Contrast enhanced CT of abdomen was suggestive of intensely and heterogeneously enhancing round to oval and iso to hypodense mass (4.8 x 4.6 cm) in right adrenal gland showing almost homogenous enhancement during delayed phase. Findings were consistent with right adrenal pheochromocytoma, bilateral renal cyst, calcification in right lobe of liver and lumbar spondylosis Fig. 2.

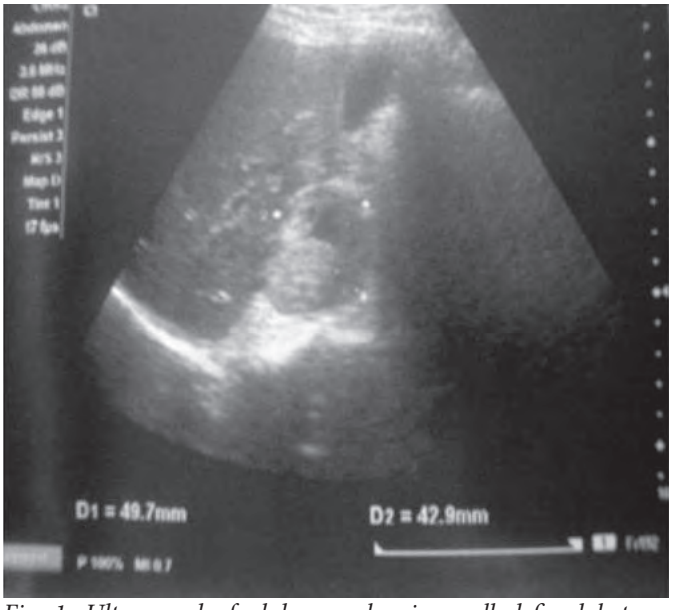

Fig. 1. Ultrasound of abdomen showing well defined heterogeneously echogenic mass measuring 4x5 cm with small cystic changes seen in right suprarenal area suggestive of adrenal mass ? pheochromocytoma.

DISCUSSION:

Pheochromocytoma, a tumor of neuroendocrine origin, is a rare tumor found in children and adults and is a cause of symptomatic hypertension. Pheochromocytoma is a catecholamine secreting tumor that arises from chromaffin cells of the sympathetic nervous system (adrenal medulla and sympathetic chain); however, the tumor may develop anywhere in the body. These tumors release catecholamines into the circulation, causing significant hypertension. In our case it was proved

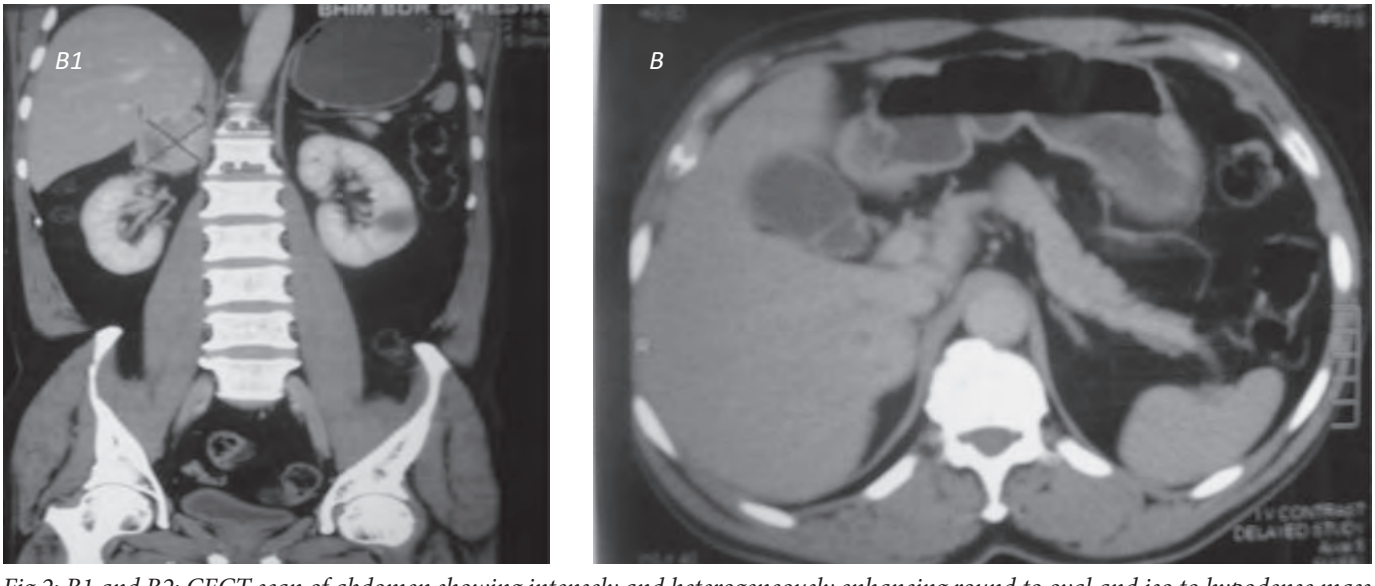

Fig 2: B1 and B2: CECT scan of abdomen showing intensely and heterogeneously enhancing round to oval and iso to hypodense mass (4.8 x 4.6 cm) in right adrenal gland showing almost homogenous enhancement during delayed phase. Findings are consistent with right adrenal pheochromocytoma.

from raised level 24 urinary VMA that tumor was releasing catecholamines.

In children, pheochromocytoma is more frequently associated with other familial syndromes, such as neurofibromatosis, von Hippel-Lindau disease, tuberous sclerosis, and Sturge-Weber syndrome, and as a component of multiple endocrine neoplasia (MEN) syndromes (MEN 2A, MEN 2B). No such association was found in our case.

Familial cases are often bilateral or multicentric within an individual adrenal gland. Adrenal pheochromocytomas are most often found on the right side and are sporadic, unilateral, and intra-adrenal. Family history was insignificant in our case.

In almost 90% of the cases, tumors are found in the abdomen- suprarenal gland or sympathetic ganglia. The extra-adrenal localization, bilateral and multiple tumors are more frequent in children than in adults and with poor prognosis.[5] Our patients had adrenal pheochromocytoma.

Hypertension appears to be uniformly present and is sustained in 80-90% of affected patients at the time of diagnosis. Occasionally, patients with sustained hypertension also have paroxysmal episodes. The paroxysms are occasionally precipitated by excitement or a particular physical activity, such as bending over or lifting a heavy object. In our case, patient developed hypertensive crises after manipulation of frozen shoulder. Two main methods for diagnosis with high sensitivity are: levels of catecholamine in urine and CT scan.[6] In our case, we trusted only the radiological imaging and catecholamine level in urine. It could not be proved histologically. The main treatment for the patient with pheochromocytoma is surgical removal of the tumor. Surgical removal is only scheduled after successful pharmacotherapy to block the effects of catecholamine excess is the key treatment. In our case blood pressure was controlled but tumor could not be removed.

CONCLUSION:

The case is interesting because of the fact, that this is a rare disease presenting with hypertensive crisis. Before the admission to the hospital, the patient had some of the typical symptoms – episodic headache, nausea, sweating and palpitation. In the presence of such complaints for several months, pheochromocytoma was not suspected in this patient. The prognosis in our case seems good. Blood pressure is well controlled with prazosin. The patient needs surgical intervention provided the availability of phentolamine and phenoxybenzamine.

REFERENCES:

1. Goodall MC, Stone C. Adrenaline and noradrenaline producing tumors of the adrenal medulla and sympathetic nerves. Ann Surg. 1960;151(3):391-8.
2. Hume DM. Pheochromocytoma in the adult and in the child. Am J Surg. 1960;99:458-96.
3. Bravo EL, Gifford RW Jr. Pheochromocytoma: diagnosis, localization and management. N Engl J Med. 1984;311(20):1298-303.
4. Eng C, Crossey PA, Milligan LM et al. Mutations in the RET protooncogene and the von Hippel-Lindau disease tumour suppressor gene in sporadic and syndromic phaeochromocytomas. J Med Genet. 1995;32:934-7.
5. Mittendorf EA, Evans DB, Lee JE, Perrier ND. Pheochromocytoma: advances in genetics, diagnosis, localization, and treatment. Hematol Oncol Clin North Am. 2007;21:509-25.
6. Davidson DF. Elevated urinary dopamine in adults and children. Ann Clin Biochem. 2005;42:200-7.