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A RARE CASE OF PHEOCHROMOCYTOMA PRESENTED WITH HYPERTENSIVE ENCEPHALOPATHY

Medicine

Dr. S. K. Gadhavicharan Associate Professor, Department Of Medicine , P.D.U. Medical College , Rajkot

Dr. Vidhi Patel 3rd Year Medicine Resident, Department Of Medicine, P.D.U. Medical College, Rajkot.

Dr. Raj Sachde 2ND Year Medicine Resident, Department Of Medicine, P.D.U. Medical College , Rajkot.

ABSTRACT

Pheochromocytoma is a type of neuroendocrine tumor that grows from cells called chromaffin cells. These cells produce hormones needed for the body and are found in the adrenal glands. The adrenal glands are small organs located in the upper region of the abdomen on top of the kidneys. Pheochromocytomas account for 0.05% of total cases of hypertension. Hypertension mainly occurring due to circulating levels of catecholamines. Approximately 20% of cases of pheochromocytomas are familial with autosomal dominant inheritance. Inherited pheochromocytomas are associated with MEN 2A and 2B, Von Hippel Lindau disease and Neurofibromatosis. Pheochromocytomas are diagnosed with urinary metanephrines and fractionated plasma free metanephrine levels. Cerebral blood flow remains unchanged over a wide range of mean arterial pressures (MAP 50 – 150 mm Hg) through a process of autoregulation of blood flow. In patients with the clinical syndrome of Malignant Hypertension, encephalopathy is related to failure of autoregulation at upper pressure limit, resulting in vasodilation and hyperperfusion. Hypertensive encephalopathy may present with severe headache, nausea and projectile vomiting, altered mentation process. If left untreated may progress to stupor, coma and death. We report an uncommon case of pheochromocytma presenting with hypertensive encephalopathy.

KEYWORDS

Hypertension, Pheochromocytoma, Catecholamines, Metanephrines

INTRODUCTION

Pheochromocytoma are rare neuroendocrine tumor that arise from cells derived from sympathetic or parasympathetic paraganglia. Prevalence is 2-8 per million. The mean age of diagnosis is 40 years. This designation is used to describe symptomatic catecholamine producing tumor including those in extra adrenal, retroperitoneal, pelvic & thoracic sites. It is due to germline mutation in RET, VHL, NF1, Subunits of SDH gene.

CASE REPORT

25 year old male present with complain of 2 episodes of GTCS & Altered sensorium since 1 hour, severe headache with intermittent vomiting & abdominal pain since 1 month prior to medical attention. Associated with excessive thirst, polyuria, sweating & palpitation. Family history – His mother is hypertensive.

ON EXAMINATION

Temp.- normal
Pulse- 130/min
Blood pressure- 230/120 mmhg
RBS- 209 mg/dl
RS/CVS- normal
CNS- conscious,irritable, not oriented to time, place, person(postictal)
Pupil- bilateral equally reactive to light
Plantar- bilateral absent
Power & tone- normal tone with all 4 limbs moving
Neck rigidity & kerning's sign- absent
Grade 2 hypertensive retinopathy changes in both Eye.

INVESTIGATION

CBC, Renal function test, Liver function test, serum Electrolytes, Thyroid function test – normal

Urine albumin- +2

USG(Abdo+ KUB+ renal artery doppler) / CT BRAINNAD

24 hour urine metanephrin- 1203 mcg/24 hour(elevated)

Metanephrines creatinine ratio- 652 mcg/creatinine(elevated)

CT Abdo with pelvis – well defined soft tissue enhancing lesion of size 62* 52*53 mm with central necrotic foci noted at left paraaortic region abutting left psoas muscle p/o PHEOCHROMOCYTOMA.

MANAGEMENT

Patient was acutely managed with injectable Labetalol. Hypertension was then controlled by tab Prazopress XL and tab Labetalol. Surgical resection was planned. Pre operative hypertension is controlled with phenoxybenzamine. Intraoperatively hypertension is controlled by phentolamine. Now patient is asymptomatic and on Tab Prazopress XL.

CONCLUSION

A high index of suspicion is required for causes of secondary hypertension in young hypertensives because pheochromocytoma carry significant morbidity & mortality if untreated. 50% to 75 % of Pheochromocytoma patients develop hypertension. Hypertensive encephalopathy is a rare but serious complication of uncontrolled hypertension, which can occur in some patients with pheochromocytoma. It is characterized by symptoms such as headache, nausea, vomiting, seizures, confusion, and altered mental status, and can lead to cerebral edema and potentially life-threatening complications.

The percentage of pheochromocytoma patients who develop hypertensive encephalopathy is not well established, as it is a rare event and can depend on various factors such as the severity and duration of hypertension, the size and location of the tumor, and the presence of comorbidities.

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