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A Rare Presentation of Pheochromocytoma

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Abstarct
Pheochromocytomas and paragangliomas are catecholamine-producing tumors derived from the sympathetic and parasympathetic nervous system. These tumors may arise sporadically or be inherited as features of multiple endocrine neoplasia type 2, von hippel-Lindau disease, or several other pheochromocytoma-associated syndromes. The diagnosis of pheochromocytomas identifies a potentially correctable cause of hypertension, and their removal can prevent hypertensive crisis that can be lethal.The clinical presentation is variable, ranging from an adrenal incidentaloma to a hypertensive crisis with associated cerebrovascular or cardiac complications.
Keywords: *PRES, Pheochromocytoma,Vonhippel Lindau.*

Introduction

Pheochromocytoma is a catecholamine producing tumour[4] that is often considered as a differential diagnosis of secondary hypertension but rarely diagnosed .The tumor is composed of chromaffin cells responsible for producing catecholamines.The classic traid[5] is episodic headache, palpitations, diaphoresis can easily recognised by most of clinicians,but the catecholamines excess can present with variety of different context. Here we are discussing a case of female presented with Posterior Reversible Encephalopathy Syndrome (PRES) [PRES, a clinical nuero radiological entiy of central nervous system due to acute severe hypertension, which has been described in Patients with Chronic Renal Failure, Eclampsia,drugs like Tacrolimus, Cyclosporine] and malignant hypertension ready to blindness

Case Report

A 18 yr female presented with gradual onset bilateral (b/l) persistent progressive painless visual loss (2 days duration) and 2 episodes of non projectile vomiting, h/o vague abdominal discomfort over right side & h/o decreased appetite.

Clinical Examination

Patient was pale with b/l pitting pedal edema . She was conscious & coherent. Her BP-210/160,PR-142/min CVS-heaving apex with ejection systolic murmur. CNS- hyperreflexia with b/l plantar extensor. No perception of light in both eyes. Fundus-cotton wool spots, hard exudates, papilledema S/O malignant hypertension, rest of the physical examination was normal.

Investigations

Hb-4.2gm%, TLC-10,900cells/cu.mm, ESR-70mm/1sthr, RBS-268mg/dl, Bun-40mg/dl,

serum.creat-1.0mg/dl, se Na+ 136,K+ 2.4,Ca+ 0.71, Hco3-24.1. Urine colour reactions positive for tyrosine and tryptophan (millon' s test and aldehyde test are positive). ECG – LVH STRAIN PATTERN. 2Decho-severe concentric LVH, turbulence across all valves,EF-65%

Ultrasound abdomen-7.7*6.6*5.3cm hypoechoiclobulated retroperitoneal mass over superior pole of right kidney MRI abdomen-right adrenal lesion,displacing IVC anteriorly. MRI orbits-normal, MRI BRAIN-ill defined T2/T2FLAIR-hyperintensities in left frotoparietal, mid corona radiata,right temporal lobe and periventricular white matter suggestive of PRES (posterior reversible encephalopathy syndrome). VEP-anterior visual pathway dysfunction. PLASMA free metanephrine levels-501pg/ml (normal-65pg/ml) confirmed it as pheochromocytoma

Treatment

Pt was started on IV nitroglycerin in acute phase and later continued with Prazosin 20mg bd, Propranolol 40mg bd.herbp was maintained at 140/90mmHg. After 8days she had normal neurological examination except for loss of vision The patient was referred to higher centre for surgical management pheochromocytoma

Discussion

This is a case of pheochromocytoma presented with PRES with malignant hypertension with persistent blindness with hyperglycemia, which is a rare presentation.The classical triad of symptoms was not found. Though Prevalence of pheochromocytoma as a cause of secondary hypertension is less (0.1 to 0.6%) it should always be kept in mind especially in young patients with secondary hypertension .the complications can be devastating which can be prevevnted by early diagnosis .

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