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Case Report

PHEOCHROMOCYTOMA WITH CRANIAL NERVE PALSIES

***S.A. Kareem and Palani**
Dept. of Ophthalmology
Sree Balaji Medical Collegeand Hospital, Chennai-600044
Bharath University, Chennai-600073
**Author for Correspondence*

ABSTRACT
Pheochromocytoma is the tumour of sympathetic and parasympathetic systems, and important cause of curable secondary hypertension. Cases present with signs and symptoms depending upon the complications produced by the hypertension. Proper diagnosis and managements can save the life of the patient .We report a case that had neuro ophthalmic manifestations in addition to features involving other systems. This case was managed in co ordination with other specialties.

Key Words: *Pheochromocytoma, Paraganglionoma, Secondary Hypertension, Vanillymandelic Acid (Vma), Cranial Nerve Palsies*

INTRODUCTION
Pheochromocytomas are tumors producing catecholamine, which are usually adrenaline and nor adrenaline. Patients may present with variable symptoms and signs due to hypertensive crisis with associated cerebrovascular or cardiac complications.
Classical triad: Headache, Palpitation and profuse sweating associated with features of hypertension.

Ophthalmic Features
The ocular abnormalities are related to secondary hypertension. They include focally or generally narrowed, retinal hemorrhages, soft and hard exudates, papilloedema and blurring of vision. Cerebro vascular complications can give rise to cranial nerve palsies with diplopia and lagophthalmos and exposure keratopathy. Orbital paraganglioma may present as slow growing tumor with progressive proptosis, diplopia and loss of vision. Neuroblastoma is the commonest cause of metastatic orbital tumor of childhood. The orbital metastasis can be diagnosed even before the adrenal primary in about 3% of cases.

CASES
A 22 year old male patient was admitted in our hospital with complaints of recurrent headache, diplopia, inability to use upper limb and lower limb in left side and dribbling of saliva from right side of mouth. He had similar problem 2 years and also 7 years before. The details of previous medical diagnosis and management were not available. Nobody in the family suffered from similar episodes.
B.P was 220/120 and pulse 120/min. CVS, RS and Abdomen were normal. There were no neurocutaneous markers. Neurological exam showed right side 6^{th} nerve palsy, LMN facial palsy and left sided hemiplegia with left 3^{rd} nerve palsy. Patient also had cerebellar symptoms like Ataxic gait, hypertonia, Broca's aphasia and dysdiadochobkinesis. Ocular exam showed RE. 6^{th} nerve palsy, lagophthalmos, with xerosis, due to Facial nerve palsy RT along with LE 3^{rd} nerve palsy. nystagmus present. Vision be 6/6, BE FUNDI normal.
Because of his high B.P., in this young age with nerve palsies, the following investigations were done.

Investigations
Haemogram, blood sugar, lipid profile, renal function tests, electrolytes were normal,except urine VMA..
24 HR URINARY (VMA) Vanillymandelic Acid 18.88mg/24hrs (normal value <6-8 mgm/ dl in adults**)**

MRI Abdomen
Multiple relatively defined discrete lesions of varying sizes noted in the bilateral para aortic region. ? Extra adrenal pheochromocytomas. Shrunken Right kidney

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CT Brian

Showed multiple infarcts in brain.

Based on these findings diagnosis of **Extra adrenal pheochromocytomas was made.**

Medical Management

High B.P. was controlled with multiple anti hypertensive drugs like alpha blockers, beta blockers, calcium channel blockers, ACE inhibitors **,** Mannitol, Nitroglycerine(NTG), phenytoin, atorvastatin, aspirin , clopidogrel, pantaprazole, ,antibiotics , physiotherapy, catheterization and bladder care. The B.P. was gradually reduced to 150/90 with episodes of wide fluctuations.

Surgical Management

The pt was taken up for surgery. A small portion of the tumor near the left renal vein was left because of difficulty in removing it without damaging the left renal artery. Thspecimen was sent for histopathology report. His B.P. was stabilized to 130/100 within a week and and was discharged on with T. Aldomet 250 mg BD**.**

Ophthalmic Management

Xerosis of the eye was taken care off with liberal application of artificial tears n lubricants.

After surgical removal of the tumor, his hemiplegia and cranial nerve palsies gradually started recovering and the patient is being followed up. The xerosis of the eye is much less now.

Histopathology Report

Small nesting growth pattern- Zellballen appearance. Tumor cells are fairly uniform with finely granular cytoplasm; Stroma shows several large blood vessel sinusoids Confirms extra adrenal pheochromocytoma.

DISCUSSION

Pheochromocytoma occur .1) Sporadic and 2) inherited as a part of Multiple Endocrine Neoplasia (MEN). The mean age is about 40 years, although the tumors can occur from early childhood until late in life. They arise from the chromaffin cells of the adrenal medulla (intra adrearenal) and also the extra adrenal sites. The most common extra adrenal locations are para aortic (75%), bladder (10%), thorax (10%), and head and neck (5%). The term Paraganglioma is used to describe catecholamine-producing tumors in the head and neck, as well as tumors that arise from the parasympathetic nervous system, which may secrete little or no catecholamines. Neuroblastoma, also from adrenal or sympathetic chain producing Catecholamine, is common malignancy in children with rapid growth & widespread metastasis.

Most of the patients of Pheochromocytoma have Headache, palpitation and profuse sweating associated with features of hypertension. Other features are anxiety, pallor, nausea, abdominal pain, weight loss, increased blood sugar, hypocalcaemia, hypercalcemia, polyuria, polydypsea, constipation, orthostatic hypotension, dilated cardiomyopathy, erythrocytosis and paradoxical response to anti hypertensives. Episodic HyperTension is a characteristic feature.

Hypertension is Difficult to Control with Routine Hypertensives

This patient presented with high B.P. along with multiple cerebral infarcts giving rise to hemiplegia with multiple cranial nerve palsies involving the BE..Once the tumor was removed surgically, his B.P. was brought under control and good improvement in his cerebro vascular status.

Probally the dignosis was missed in earlier occasions, when patient had similar episodes. . When the pt comes with symptoms of headache with or without other symptoms like diplopia, the ophthalmologist in addition to doing routine ocular exam and, neuro-ophthalmic exam, should do simple test of checking the blood pressure of the patient irrespective of age, which will help us to diagnose such cases for proper management in coordination with other specialties

Case Report

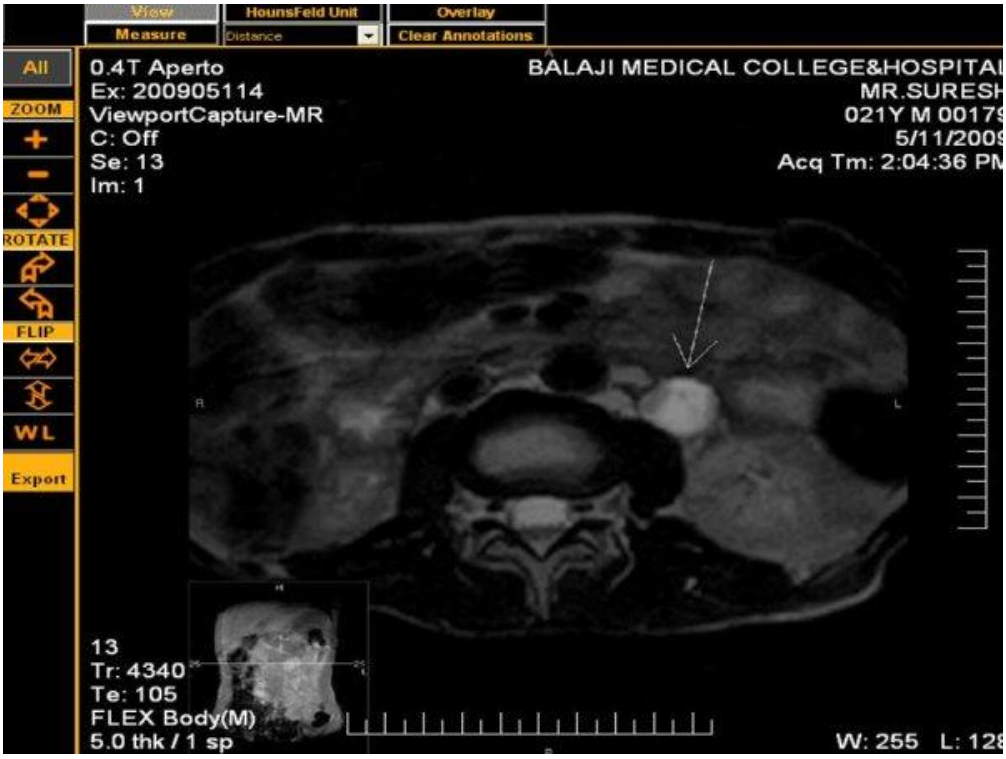

Figure 1: MRI Abdomen PHEO MRI ONE

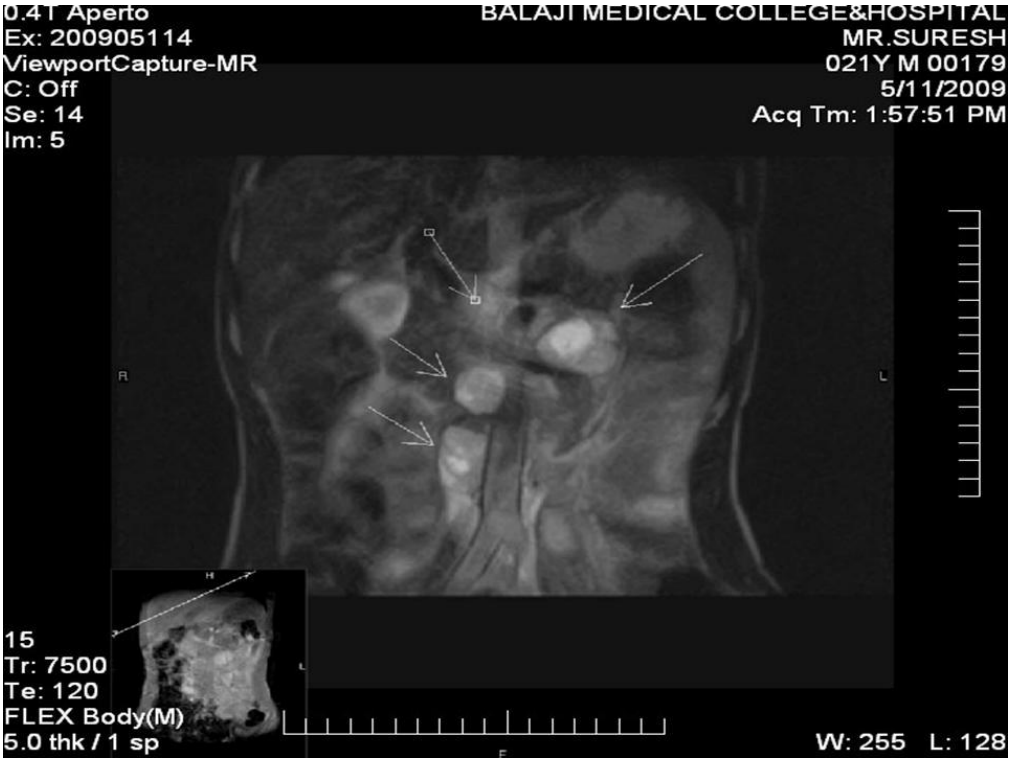

Figure 2: MRI Abdomen PHEO MRI TWO

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