

## Case Report

# A CASE REPORT OF EXTRA-ADRENAL PHEOCHROMOCYTOMA: RAREST OF THE RARE CHILDHOOD TUMOUR

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## ABSTRACT

Pheochromocytoma is a rare tumour of childhood and a rare cause of hypertension in children. It arises most commonly from catecholamine releasing chromaffin cells of adrenal medulla but can also arise in extra adrenal sites from paraganglionic chromaffin tissue of sympathetic nervous system extending from the base of skull to urinary bladder. Herein we highlight the case of a 14 year old male child who presented with this rare tumor in an extra-adrenal location. The child at presentation had classical triad of headache, dizziness and palpitation associated with episode of micturation since one year. His blood pressure was noted to be very high (>95<sup>th</sup> percentile) and considering the clinical suspicion of this tumor a CT abdomen and urinary VMA levels were done. The CT abdomen revealed an extra adrenal pheochromocytoma involving the bladder wall and surrounding peri-pancreatic and aorto-caval tissue. Extra adrenal pheochromocytomas comprise around 10% of all such tumors. They need to be clinically suspected and picked up early on basis of classical symptoms as otherwise they may precipitate life threatening hypertension or cardiac arrhythmias due to high catecholamine release. Surgical removal is the treatment of choice with prior adequate alpha receptor blocking with various drugs to prevent complications during the surgery.

**Keywords:** Pheochromocytoma, Extra- Adrenal, VMA, CT Abdomen

## INTRODUCTION

Pheochromocytoma is a rare tumour of childhood. It affects boys more than girls. It arises most commonly from catecholamine releasing chromaffin cells of adrenal medulla but can also arise in extra adrenal sites from paraganglionic chromaffin tissue of sympathetic nervous system extending from the base of skull to urinary bladder Neuman HP (2008). Due to their tendency to release catecholamines in body, the patients typically have high blood pressure and hence the tumor is considered in clinical differential diagnosis in every young patient presenting with high blood pressure symptoms. The presenting symptoms of pheochromocytoma are well defined in literature as five P's i.e. Pressure (hypertension), Pain (headache), Perspiration, Palpitation and Pallor. It is also called the "10% tumour as 10% are extra adrenal, 10% occur in children, 10% are familial, 10% are bilateral or multiple, 10% are malignant and 10% are discovered incidentally. Common locations of extra adrenal pheochromocytoma however include urinary bladder wall, heart, mediastinum and carotid and glomus jugulare bodies Young L (2007). Radiological imaging modalities like ultrasound, computed tomography and MRI are useful in localizing the origin and extent of tumour. <sup>131</sup>I-metaiodobenzylguanidine radioisotope study is indicated when above imaging studies are inconclusive or to detect metastatic sites Bouloux PM (1997). Plasma metanephrine testing though has high sensitivity as a screening test but it lacks specificity. For biochemical confirmation 24 hour urinary levels of metanephrines and catecholamine's are done as these have a high sensitivity of 87.5% and specificity of 99.7% De Jong WH (2009).

## CASES

A 14 year old male child presented to the pediatric ward of our hospital with complaints of sudden episodes of intense headache, dizziness, palpitations and sweating associated particularly with micturation. These symptoms had been present since one year. The child had been evaluated outside by various practitioners, without any relief in symptoms or any conclusive diagnosis. He had a report of an

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ultrasound abdomen from outside which showed some mass in the epigastric region. On further physical examination in our unit, no specific abnormality was noted. However, the child was noted to have a pulse rate was-110/min, respiratory rate-18/ min, blood pressure-150/100mmHg with visible evidence of sweating while he passed urine and experienced episodic symptoms. The flow, colour and consistency of urine were normal. With a clinical suspicion of pheochromocytoma on a repeat ultrasound in our hospital, child was further evaluated with CT-abdomen and plasma and urinary examination for metanephrines and catecholamine's (VMA).

The plasma free metanephrine levels were 376.8 pgm/ml (Reference range 12-61 pgm/ml) and normetanephrine levels 1378.49 pgm/ml (Reference range 18-112 pgm/ml). Urinary VMA levels were also found to be very high 17.68mg/g (reference values- 1.5-3.6mg/g) and CT abdomen showed a significant polypoidal bladder growth (3.8cmx2.3cmx2.7cm) with another similar lesion in the peri pancreatic and aorto-caval region (2.5cmx2.2cm). Considering the characteristic clinical, biochemical and radiological findings, a diagnosis of extra adrenal pheochromocytoma was made. The child was further referred to higher tertiary care centre for surgical management. We however, managed the child's high blood pressure and other symptoms with propranolol drug.

## DISCUSSION

Pheochromocytoma is mostly seen in adrenal gland but extra adrenal sites are not also very uncommon. As in this case, the primary site was extra adrenal in nature involving primarily the urinary bladder and the sympathetic chain in the peripancreatic and aorto-caval area. Diagnosis of pheochromocytoma typically requires confirmation by several tests, perhaps the most important being biochemical evidence of excessive catecholamine production by the tumor. This is usually achieved from measurements of catecholamines and certain catecholamine metabolites (metanephrine and normetanephrine) in urine or plasma. The levels of catecholamine metabolites in the plasma and urine in our index case were more than five times the reference range and it has been well highlighted in literature that such high levels are quite diagnostic of a pheochromocytoma De Jong WH (2009). In few studies on extra adrenal pheochromocytoma, the incidence of such tumors has been found to be ranging from 12-42% and the extra adrenal sites are more commonly described in pediatric age group than adults. A CT abdomen is the radiological investigation of choice to note the site of origin and extent of tumor, but MRI and radionuclide scanning are also employed in certain special cases Shimazui T (1989).

Surgical resection of the tumor is the treatment of choice and usually results in cure of hypertension. Careful pre operative preparation is however required with combined alpha and beta blockade to control blood pressure and to prevent hypertension Wong C (2010). The preferred drug of choice before surgical preparation is phenoxybenzamine, an alpha blocker, in a dose of 20mg initially until dose of 100-160mg is achieved and the patient reports symptomatic postural hypotension. Additional beta blocker is required if tachycardia or arrhythmias develop and they however should not be introduced until the patient is alpha blocked because unopposed alpha adrenergic receptor stimulation can precipitate hypertensive crisis.

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