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Case report

A UNIQUE CASE OF PHEOCHROMOCYTOMA PRESENTING WITH HYPERTENSIVE RETINOPATHY

*Maji.S[1], Saha. ML[2], Kanwar KS[3], Das S[4], Bhagat P[5], Bhar P[6]

[1,3]Resident, [2]Professor, [4,6]Assistant Professor, [5]RMO, Department of General Surgery, I.P.G.M.E&R, SSKM&H, Kolkata, India

*Corresponding author email: drsuvendumaji@rediffmail.com

ABSTRACT

Pheochromocytoma is an extremely uncommon tumor of childhood and there are several features that distinguish its presentation between adults and children. The incidence of pheochromocytoma in childhood is 10% of the adult incidence, occurring in approximately 1 in 500,000 children compared with 1 in 50,000 adults. Around 10% of childhood tumors are familial which is 4times the frequency in adults. Whereas only 7% of pheochromocytomas are bilateral in adults, the reported incidence of the same in children range from 24 % to as high as &70%.These tumors are known for their great diversity in clinical presentation. Greater than 50% of children present with headaches, fever, palpitation, thirst, polyuria, sweating, nausea and weight loss. However the commonest mode of presentation is sustained hypertension. Pheochromocytoma accounts for 0.5% of children with hypertension and must be considered once other causes have been eliminated. We here in report a unique case of a 13 year old girl who initially presented with bilateral hypertensive retinopathy and later found to have a pheochromocytoma on subsequent workup. Hypertensive retinopathy secondary to pheochromocytoma is itself a rare entity whose exact incidence in children is still unknown. This case highlights the importance of routine history, physical examination and measurement of bp. Prompt surgery can reverse the effect of hypertension and lead to good outcome as was evident in our case.

Keywords: Pheochromocytoma, Hypertensive retinopathy, Metanephrines, Normetanephrines, Zellballen

INTRODUCTION

Pheochromocytomas are rare tumors with prevalence rates ranging from 0.3 to 0.95% in autopsy series, and approximately 1.9% in series using biochemical screening. Recent advances in molecular genetics have shown presence of germline mutation in up to 59% of apparently sporadic pheochromocytomas presenting at 18 years or younger and in 70% of those presenting before 10 years of age[1].They can occur at any age with a peak incidence in the fourth and fifth decades of life, and have no gender predilection[2]. Pheochromocytoma is rare in children[1].It is a catecholamine producing tumor of the sympathetic nervous system[9]. The presentation varies from vague symptoms to hypertensive emergencies. Headache, palpitations, and diaphoresis constitute the "classic triad" of pheochromocytomas. The hypertension related to this tumor may be paroxysmal with intervening normotension, sustained with paroxysms, or sustained hypertension alone. In children however hypertension often remains sustained. Presentation may not be always with the above classic triad and unique presentation as in our case needs to be kept in mind! The treatment of pheochromocytoma remains surgical excision although medical management of hypertension is an essential part of preoperative preparation[3].

Case report

A 13 year old girl presented with blurring of vision since a week .She underwent ophthalmic assessment in a private eye clinic where she was found to have bilateral hypertensive retinopathy. She was then admitted to our SSKM hospital, initially in the general medicine ward where she underwent full work up and physical examination. She was later transferred to the surgical unit. Her history revealed that she had complaints of palpitation, throbbing headache and occasional sweating for the last 7 month. She also had associated weight loss. However her appetite, bowel and bladder habits were normal. On admission she had a bp of 180/110 mm of Hg and a pulse rate of 130/mint. On abdominal examination no lump was found. On USG a large (3.92x 4.21) cms hetrogenous mass with cystic degeneration above superior pole of left kidney was found. CECT abdomen revealed (4.5 x 3.9) cms cystic sol with enhancing thick walled mass in the left adrenal gland. Biochemical tests showed high value of 24 hour urinary metanephrine level (14.36) microgram/litre and elevated level of normetanephrine (1445.1) microgram/litre. There was no drug history of ephedrine, amphetamines, methlxanthines etc use in this patient which could have lead to false elevated metanephrine level. Serum levels of Ca^{2+},PTH, phosphate, calcitonin were all normal. Preoperatively she received prazosin & propnanolol. She underwent left adrenalectomy under GA following which she was shifted to ICU where constant monitoring of her vitals was undertaken. She was hypotensive and had to be put on noradrenaline drip for 3 days. She was started on oral diet by 5^{th} postop. day and was discharged on the 7^{th} day. She was followed up with a repeat test of serum and urinary markers which showed normal results. Her pathological report showed a well encapsulated tumor composed of large polygonal cells, vesicular nuclei, small nucleoli and abundant eosinophilic granular cytoplasm, arranged in Zellballen, surrounded by elaborate vascular network which was consistent with a diagnosis of benign pheochromocytoma [fig 1&2]. On 6 months follow up she has been healthy and enjoying a good quality of life without any visual problem.

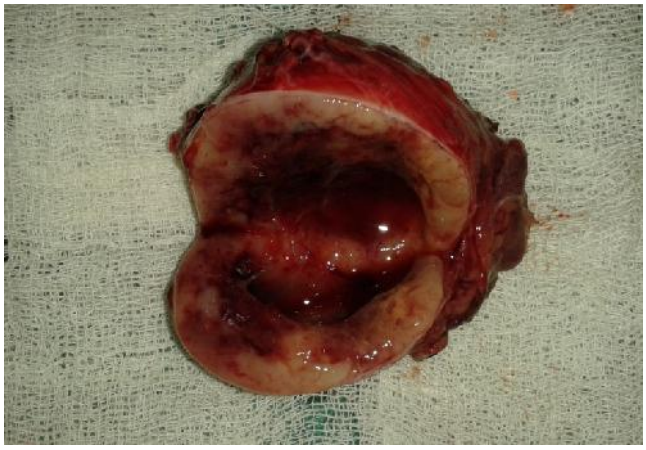

Fig1: Pathology specimen showing the cystic adrenal tumor.

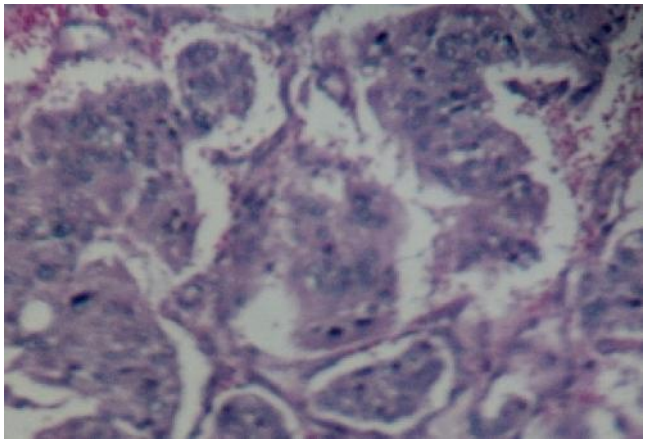

Fig 2: Microscopic appearance showing tumor cells arranged in Zellballen

DISCUSSION

Pheochromocytomas are catecholamine-secreting neoplasms. Due to its variable presentation, they have been called the “masquerader” .The clinical spectrum, ranges from completely asymptomatic (10%) to a sustained stable hypertension (50%), or to frequent life-threatening hypertensive crises (30%). Majority of the patients present with the classical triad of episodic headache, palpitations, diaphoresis, and a feeling of impending doom. Several series have reported that 19-76% of pheochromocytomas are not diagnosed until after death[2] & the incidence of asymptomatic tumors being is 4.4 to 17 %[3]. Atypical symptoms described in literature include abdominal pain, vomiting, polyuria, polydipsia, heart failure, cerebrovascular hemorrhage. A pheochromocytoma presenting initially with hypertensive retinopathy is rare and its incidence is unknown. About 10% of pediatric pheochromocytoma is thought to be familial[4]. Studies of families with pheochromocytoma occurring across several generations suggest a dominant autosomal mode of inheritance with high

penetration[4,5].Familial cases of pheochromocytoma also carry a higher risk of malignancy than sporadically occurring varieties[6].The diagnosis of this tumor relies on the demonstration of blood and urinary catecholamine's and their metabolites. A 24 hr urine measurement of catecholamine's, metanephrine and vanillyl mandelic acid is the best diagnostic test. Once the chemical diagnosis is established the tumor must be localized. Radiological imaging modalities like USG, Contrast Enhanced CT scan and MRI are useful in determining the origin and extent of tumour. Prompt diagnosis and complete excision are the most important treatment for childhood pheochromocytoma[7,8].Pre-intra-and postoperative medical management is as important as the surgical procedure itself. All patients should undergo follow up to confirm normalization of catecholamine levels. [9,10]

This case was unique and interesting due to following reasons:

1. Hypertensive retinopathy as an initial presentation of pheochromocytoma is extremely rare. Data about this is limited and is mainly in form of few case reports. Most of these are from western literature[9,10] and very few from the Indian subcontinent.
2. Though childhood pheochromocytomas are frequently multiple, bilateral and frequently have familial associations our case was unique being unilateral, single lesion and sporadic in presentation.
3. It highlights the importance of blood pressure measurement in the clinical diagnosis of this rare condition. Early diagnosis is crucial not only because it is a curable cause of severe hypertension but also since unrecognized tumor may provoke fatal hypertension crisis during surgery, some diagnostic procedures or other stresses.

CONCLUSION

A high index of suspicion and early diagnosis is key to successful management in pheochromocytomas .A high blood pressure in a child should prompt thorough search for this condition. Preoperative stabilization of blood pressure is crucial in preventing intraoperative catastrophe of uncontrolled hemorrhage.

Conflict of Interest: Nil

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