BILATERAL PHEOCHROMOCYTOMA

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INTRODUCTION

Although Pheochromocytoma represents only a rare cause of Hypertension comprising less than 1% of cases, excluding this diagnosis is critical. Pharmacological mismanagement can be disastrous and contribute to high mortality rate.[1,2,3]

The index of suspicion for pheochromocytoma is increased by the presence of paroxysmal symptoms such as palpitation sweating and headache[2] in hypertensive patients.

10% of pheochronocytoma are associated with a variety of familial syndromes, like Multiple Endocrine Neoplasia, neuro fibromatosis and von Heppel Lindau disease.[4,5]

CASE REPORT

We present a case of bilateral pheochromocytoma in a 12 year old boy from Afghanistan. Two and half years ago, the patient had a sudden attack of severe headache, associated with palpitation and drenching sweats. He complained of pain abdomen for the same duration. He consulted a local hospital in Afghanistan, where he was found to have high blood pressure. He was put on treatment and improved temporarily. Since the onset of symptoms his headache, palpitations and hypertension persisted with periodic exacerbation. The attacks were precipitated by stressful conditions.

For the same complaints, he was admitted in Japan-Afghan Medical Centre, Peshawar. His B.P. was 160/140 mm Hg. on admission. He remained hospitalized for 10 days. IVP revealed delayed clearance on left side. He was labelled hypertensive secondary to left renal artery stenosis. Later he was admitted in Medical "B" unit, Postgraduate Medical Institute, Lady Reading Hospital Peshawar.

His haemogram was normal. Renal functions, blood glucose and serum calcium were normal. VMA were 24.8 mg/24 hours. (Normal 1.9–9.8). Thyroid tests were normal. ECG showed left ventricular hypertrophy. Excretory urogram revealed delayed clearance on left side.

Ultrasound revealed a 5.2 x 4.2 cm right suprarenal hypoechoic mass with pressure effect on inferior V. cava a left side 2.9 cm lobulated hypoechoic mass in portal region. There was no paraortic lymphadenopathy; liver, spleen, pancreas and thyroid were normal.

C.T. scan abdomen showed appearances consistent with bilateral pheochromocytomas, the right measuring 5x5 cm and left 3x2.4 cm. No evidence of secondaries in the liver or enlarged lymph nodes was seen.

This patient's blood pressure was controlled with Phenoxy benzamin initially, later Beta blocker (Inderal) was added. After a week of Alpha blockade followed by Beta-blockade this boy under-went surgery by paediatric surgeon and vascular surgeon. An anterior approach was made.

During operation while manipulating right adrenal, blood pressure went up which was controlled with Nitroprusside infusion. In view of the big size and some suspicion of the adjuscent lymphnode enlargement, bilateral adrenalectomy was performed.

Post operative period was uneventful. His blood pressure came to normal. VMA. level after a week was 5 mg/24 hours. He was discharged on Prednisolone 7.5 mg daily.

DISCUSSION

Pheochromocytomas are defined as Catecholamine secreting tumours, usually derived from adrenal medulla. But they may develop from chromaffin cells in or around ganglia the sympathetic. The term pheo is greek word indicating dusky appearance of the cut surface of the tumour.6

Types. A. Adrenal pheo

B. Extra adrenal pheo or paragangliomas.

A. Adrenal pheo. It is common. 90% arise from adrenal medulla. Usually benign, only 10% are bilateral or multiple.[7] In children pheo are usually bilateral or multiple.[8]

B. Extra adrenal pheo or Paraganglioma: as the name indicate, these tumours arise from chromaffin located along the sympathetic ganglia.

Sites - I. Cervical region II. Posterior mediastinum III along the Aorta. IV In the organ of Zukerkandl[9] ventral to Aorta, near the origin of inferior mesentesic artery. This is the common site for Ext. adrenal pheo. V. In the pelvis VI. in the urinary bladder VII. in the carotid body (chemodietoma). Overall 99% of pheo - are abdominal or abdomino - pelvic. Only 1% cervical or thoracic.

Incidence

Equally affects male and female. It occurs from child hood to 7th decade of life (peak age 30-42 years).[10] In adults 80% are unilateral and solitary. 10% are bilateral and 10% are extra-adrenal.

Children

Occurrence is rare, frequently bilateral and renal familial incidence is high. Inheritnace is autosomal dominant gene. This inherited disorder may occur alone or in association with other disorders like multiple endocrine neoplasia syndrome. (MEN)

MEN Syndrome are of two types

I. MEN I Syndrome (Wermers syndrome). It is association of Parathyroid Adenoma, pituitary Adenoma and Panceatic endocrine adenoma (insulinoma or Gastrinoma). This syndrome has nothing to do with Pheochromocytoma.

II. MEN II ..... MEN IIa (Sipples syndrome).[11]

..... MEN IIb (Mucosal neuoromal syndrome).

MEN IIa (Sipples syndrome) — This is association of medullary Ca. of the thyroid, parathyroid adrenoma and bilateral pheo. This syndrome is relatively common than MEN IIb.[12]

The expression of this disorder varies from family to family, any component of the disorder may occur first and may proceed the others by years.

MEN IIb

This is association of the medullry Ca. of thyroid, bilateral pheo and neuro-Ectodermal syndromes like;

- Neuro fibromatosis (Von- Rechlen-gansun)
- Von-Hippel Lindna disease.
- Sturge Weber disease.
- Tuberous Sclerosis.

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