

Phaeochromocytoma of Adrenal Gland

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Abstract

Pheochromocytoma is a rare catecholamine secreting tumour originating usually from adrenal medulla and produces signs and symptoms due excessive catecholamine secretion from tumour. Pheochromocytoma is a rare cause of hypertension. If the diagnosis of pheochromocytoma is overlooked, the consequences could be disastrous, even fatal; however, if a pheochromocytoma is identified, it is potentially curable, as being one of the cause of surgically correctable hypertension [1,2,3,5,6,9,10,11].

Keywords: Pheochromocytoma; Catecholamine; Hypertension.

Introduction

Pheochromocytoma is a rare tumor originating from catecholamine secreting chromaffin cells that are derived from the ectodermic neural system and mostly situated within the adrenal medulla [1,2,3].

Also called paraganglioma of adrenal medulla (extra-adrenal tumors are called extra-adrenal paragangliomas). Called "10% tumor": 10% bilateral (probably higher), 10% outside adrenal medulla, 10% metastasize (probably higher), 10% in children [1,2,4,6,7].

Case Report

31 years old housewife admitted into our hospital with the complaints of paroxysmal attacks of palpitation, dizziness, blurring of vision and headache for last 06 months.

On examination, patient had no abnormal physical findings except Blood Pressure (BP) is high during paroxysmal attack (Systolic BP varies from 140 to 210 mmHg and diastolic BP varies from 100 to 140 mmHg). Complete Blood Count, Random Blood Sugar, Blood urea, Chest X-rays and ECG reports were within



Fig. 1: Gross: Chromaffin test (Potassium dichromate test): fresh tumour changes the to dark brown (due to presence of chromaffin cells)

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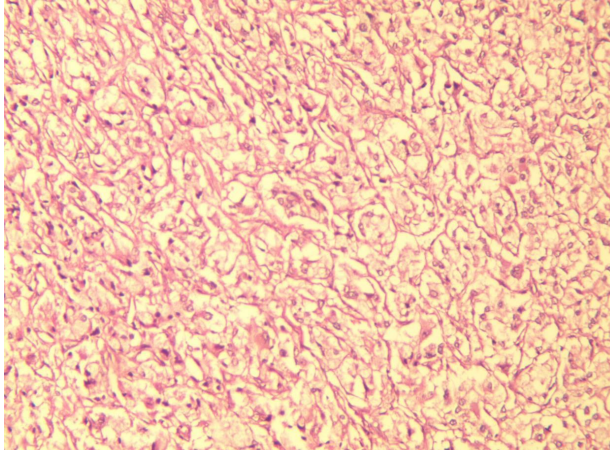


Fig. 2: Legends to figures: Zell ballen pattern (H & Estain , 10X)

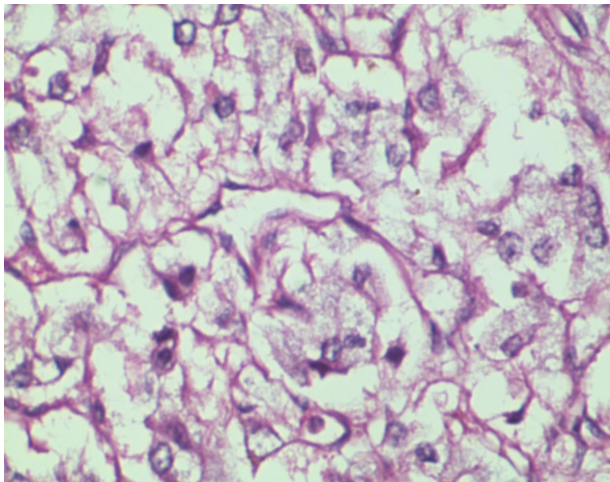


Fig. 3: Legends to figures: Zell Ballen Pattern (H & E stain , 40 x)

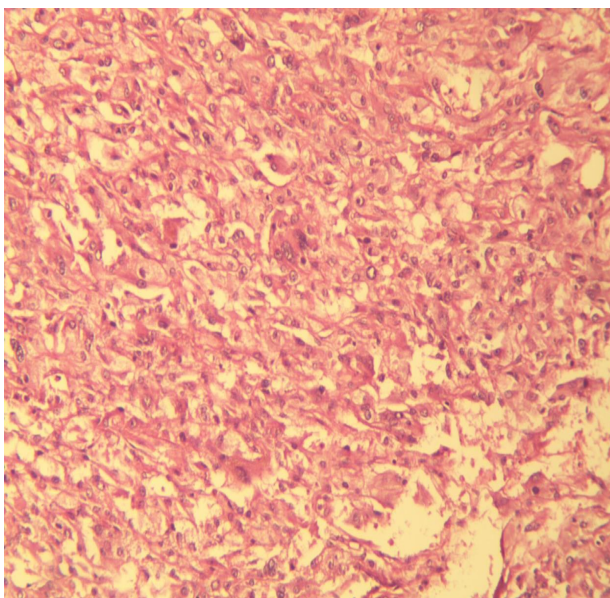


Fig. 4: Legends to figures : Few scattered large ganglion like cells (H & E stain, 10 x)

normal limit.

Provisional diagnosis of secondary hypertension due to adrenal pheochromocytoma was done and to confirm that biochemical investigations i.e. serum catecholamine level or 24 hour urinary catecholamine level were advised Abdominal USG shows right sided adrenal mass of 8 cm in diameter.

Patient underwent surgical removal of right adrenal gland (Rt. Adrenalectomy) Operative procedure was uneventful Blood pressure becomes normal (Systolic 110 to 90 mmHg and diastolic 80 to 60 mmHg) from 1st POD without any drug Finally, Histopathology report further confirmed the adrenal tumour was pheochromocytoma.

Discussion

Pheochromocytoma is a rare neoplasm, which are derived from cells of the chromaffin tissue and mostly situated within adrenal medulla. Only approximately 15% Pheochromocytoma develops from extra-adrenal chromaffin tissue which lies in the chromaffin tissue of the sympathetic nervous system extending from base of skull to the urinary bladder [1,2,3,4,6,7,8]. Common locations of extra-adrenal Pheochromocytomas include the organ of Zuckerkandl (close to origin of the inferior mesenteric artery), urinary bladder wall, heart, mediastinum and carotid and glomus jugulare bodies Pheochromocytomas occur in people of all races, although they are diagnosed less frequently in blacks and equal frequency in male and female [1,7,10,11]. Pheochromocytomas may occur in persons of any age. The peak incidence, however, is between the third and the fifth decades of life [1,4,6,7,8]. Approximately 10% occur in children. The majority of cases are sporadic, with only 16% having a history of associated endocrine disorder such as Multiple Endocrine Neoplasia type II (MEN IIA and IIB), Neurofibromatosis 1 (NF 1) and von Hippel-Lindau disease (VHL) [1,2,4,5,10].

* Approximately 10% of pheochromocytomas are malignant

* Direct invasion of surrounding tissue or the presence of metastases determines malignancy. Unfortunately, no reliable clinical, biochemical or histological features distinguish a malignant from a benign pheochromocytoma [1,2,6,7,10].

The clinical manifestations of a pheochromocytoma results from excessive catecholamine secretion by tumour. Catecholamines typically secreted, either intermittently or continuously, includes norepinephrine and epinephrine; rarely dopamine is secreted The

classic history of a patient with pheochromocytoma includes episodes characterized by headaches, palpitations and diaphoresis in association with severe hypertension. (1,2,7,10,11) Paroxysms may be precipitated by physical training, induction of general anaesthesia and numerous drugs and agents (contrast media, tricyclic antidepressive drugs, metoclopramide and opiates) [1,2,4,5,6,7,8,11].

Plasma metanephrine testing has the highest sensitivity (96%) for detecting a pheochromocytoma, but it has a lower specificity [8,9,10].

Conclusion

Pheochromocytoma is often called '10% tumour' because 10 percent are bilateral, malignant, extra-adrenal, multiple, familial and occur in children¹⁸. Pheochromocytoma is one of the few causes of hypertension that can be treated surgically. Although it is the causative factor of hypertension in about 0.1% to 0.6% of the hypertensive population, detection is mandatory, not only for the potential cure of the hypertension but also to avoid the potentially lethal effects of the unrecognized tumor.

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