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CASE REPORT

Pheochromocytoma: A Case Report

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ABSTRACT

Pheochromocytoma refers to the intra adrenal chromaffin neoplasm which is derived from neuroblast from neural crest. Signs and symptoms associated with pheochromocytoma are due to excessive secretion of catecholamines. 65% to 70% cases are sporadic and 30% to35% are manifestation of inherited tumor syndromes. There is germline mutation in gene which encode B, C and D subunit of Succinate dehydrogenase (SDH). Here, we present a case of a 50 year old male patient presenting with paroxysmal attacks of hypertention, palpitation, dizziness, blurring of vision and headache for six month. Clinically he had high systolic and diastolic blood pressure. Radiological imaging studies (CT scan) and abdominal ultrasonography suggested suspected left adrenocortical carcinoma. However histopathological (HP) and immunohistochemical (IHC) examinations confirm the diagnosis of pheochromocytoma. Appropiate diagnosis of pheochromocytoma is important to avoid fatal concequences due to excessive secretion of catecholamines. Moreover, it is a cause of surgically correctable hypertention.

***Keywords:** Pheochromocytoma, hypertention, histopathology, immunohistochemistry*

INTRODUCTION

Pheochromocytoma is an uncommon tumor originating from catecholamine secreting chromaffin cells that are derived from the ectodermic neural system and mostly situated within the adrenal medulla (0.005% to 0.1% of unselected autopsies).[1] Described by Poll in 1905 as having cut surface with dusky [pheo] color [chromo]. Because of excessive catecholamine secretion, Pheochromocytoma may precipitate life-threatening hypertension or cardiac arrhythmias. Pheochromocytoma has been called the 10% tumor-approximately 10% are bilateral, 10% are extra adrenal (i.e., they are paragangliomas), 10% occur in children and 10% are malignant.The susceptibility genes for Pheochromocytoma include RET, VHL, NF1 and SDHD, SDHC and SHCB.[2]

CASE REPORT

A fifty years old male patient admitted in the Endocrinology Department of GMCH with the complaints of paroxysmal attacks of hypertension along with palpitation, dizziness, blurring of vision and headache for last 6 months.

On systemic and general examination of the patient, no abnormal findings noted except high Blood Pressure (BP) during paroxysmal attacks (Systolic BP varies from 140 to 190mmHg and diastolic BP varies from 100 to 140 mmHg, with tachycardia). Complete blood count and other Biochemical investigations were normal except raised Random blood sugars (269mg/dl).

Serum catecholamine or 24 hour urinary catecholamine levels were not done because facilities were not available locally. **Abdominal USG shows** retroperitoneal mass lesion. **CT (computed Tomography) scan of abdomen report was given as** suspected left adrenocortical carcinoma. Later on Left radical adrenelectomy was done in GMCH and specimen was sent to Pathology department of GMCH for histopathological examination.

Grossly: Received one nodular light brown soft tissue mass weighing 100 gm, measuring 10x10x6 cm^3 involving the whole adrenal gland. Cut surfaces were yellowish dusky red brown in colour, consistency was soft (**Figure 1**).

Figure 1 Gross picture of Pheochromocytoma

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Microscopically: Multiple random sections from the tumor mass showed capsulated tumor with predominantly well defined nested pattern (zellballen appearance) with focal areas of solid architecture separated by delicate fibrovascular network. Focal areas showed rim of sustantacular cells at periphery of some nests. Tumor cells have varying size and shape with round nuclei, prominent, nucleoli and granular amphophilic to basophilic cytoplasm. No evidence of regional or distant metastasis noted. Patient is doing well on follow up and his Blood pressure came to normal level after operation. Occasional intracytoplasmic hyaline globules also noted. Mitotic count 0-2/50 hpf. Necrosis and hemosiderin pigment deposition also noted in some areas. There was no evidence of capsular invasion and lymphovascular invasion (**Figure 2, 3**).

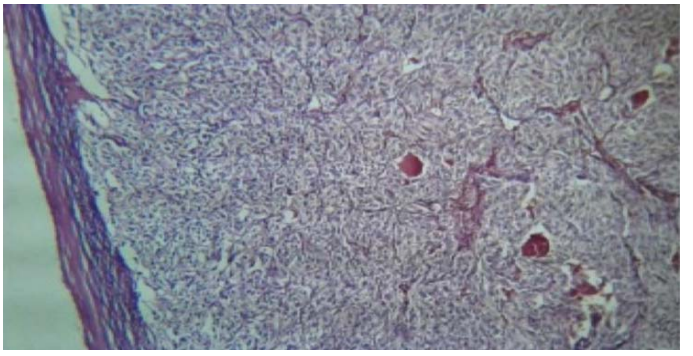

Figure 2 Photomicrograph of pheochromocytoma with capsule (H & E,10x10)

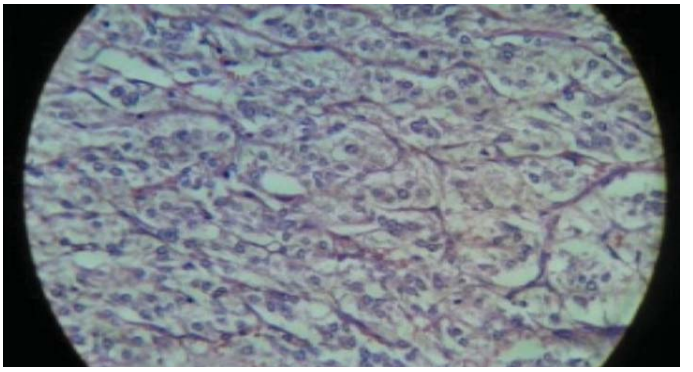

Figure 3 Photomicrograph of Pheochromocytoma, zellballen pattern (H & E, 10 x40)

Based on clinical findings, gross pathology and histopathological features diagnosis of Pheochromocytomawas made and advised for IHC confirmation. On **immunohistochemistry** the tumour was positive for chromogranin and vimentin and negative for inhibin, (**Figure 4, 5, 6**) calretinin and cytokeratin.

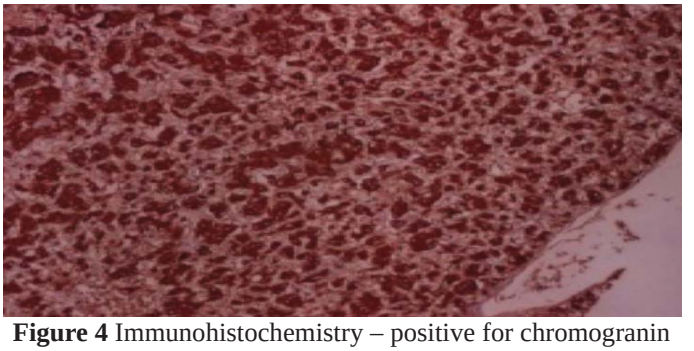

Figure 4 Immunohistochemistry – positive for chromogranin

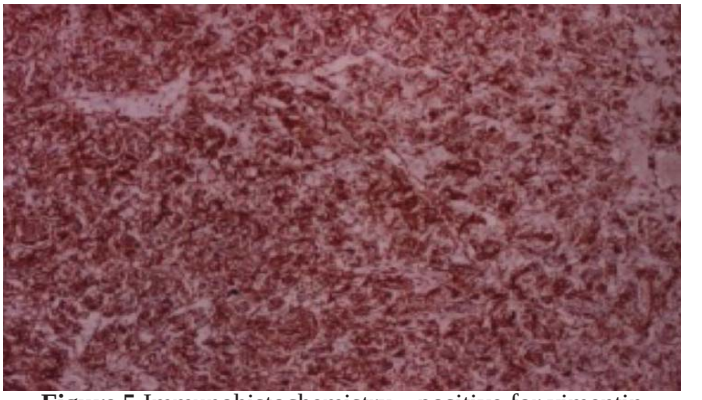

Figure 5 Immunohistochemistry – positive for vimentin

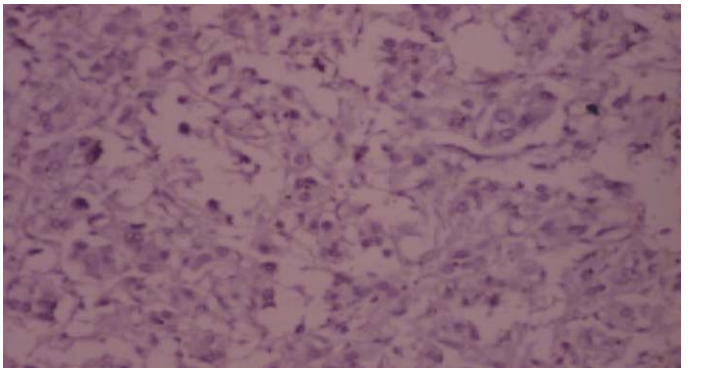

Figure 6 Immunohistochemistry – negative for inhibin

Postoperative period was uneventful with normal systolic and diastolic blood pressure levels. Patient was discharged on 7th postoperative day and now he is on regular followup without any evidence of recurrence or complication.

DISCUSSION

Pheochromocytoma is an uncommon tumor. It is also called paraganglioma of adrenal medulla (extra-adrenal tumors are called extra-adrenal paragangliomas). Common locations of extra adrenal pheochromocytomas include the organ of Zuckerkandle (close to origin of the inferior mesenteric artery), urinary bladder wall, heart, mediastinum and carotid and glomusjugulare bodies.[3] Mean age of pheochromocytoma is 47 years in one series, range is 3-81 years. In children, usually extra-adrenal, bilateral and associated with MEN 2a/2b. The clinical manifestation of pheochromocytoma results from excessive catecholamine secretion by the tumor, it represents 0.1% of patients with hypertension, but may be fatal. It occurs in equal frequency in male and female. Only the presence of metastases defines malignancy.[4] Main differential diagnoses are adrenocortical carcinoma, adrenocortical adenoma, neuroblastoma.[5] In adrenocortical carcinoma, histopathologically there is marked nuclear pleomorphism and hyperchromasia , diffuse pattern of growth and high mitotic count. In IHC-cells are immunoreactive for inhibin, calretinin and cytokeratin. Adrenocortical adenomas are negative for chromogranin. Neuroblastomas are found in children and composed of small round blue cells often with pseudorosette formation.

Surgical resection of the tumor is the treatment of choice and usually results in cure of hypertension. Although it is the causative factor of hypertension in about 0.1% to 0.6% of the hypertensive

population detection and proper diagnosis of pheochromocytoma is mandatory, not only for the potential cure of hypertension but also to avoid the hazardous effects of the undiagnosed tumor.[6] The massive release of catecholamines in pheochromocytoma can cause damage to heart cells. This damage may be due to either compromising the coronary microcirculation or by direct toxic effects on the heart cells.[7]

CONCLUSION

Although Pheochromocytoma is a rare cause of hypertension, early diagnosis is necessary to prevent the fatal outcome of hypertension. Most of the time radiology cannot give confirmatory diagnosis. Hence, histopathology and immunohistochemistry play important roles in confirmatory diagnosis of Pheochromocytoma, which helps in proper management of patients.

Conflict of interest: None.

Due consent from the patient for publication: Taken.

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LETTER TO THE EDITOR

Sir,

Casuistry in defense of Forensic Medicine

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Casuistry is defined as "the interpretation of moral issues, using procedures of reasoning based on paradigm and analogies, leading to the formulation of expert opinion about the existence and stringency of certain particular obligation, framed in terms of rules or maxims that are generated but not universal or invariable, since they hold good with certainty only in the typical conditions of the agent and circumstances of action". [1] The usefulness of the maxim is that it provides cash value in making a quick defensible decision respected by professionals and the law. By direct application it favors doing well to others and in case it meets an obstacle, it seeks analogy by extension. Some people are of the opinion that modern medical ethics is casuistry.

Forensic Medicine has got several sub-disciplines namely, Forensic Pathology, Forensic Serology, Forensic Anthropology, Forensic Psychiatry and Clinical Forensic Medicine. By implication casuistry would be more in tune with clinical aspect of Forensic Medicine. However, this doesn't mean that other sub-disciplines would receive an unfair treatment.

Casuistry like is an arrow ready to pierce the phenomenon of "shared insanity". In shared insanity two or more siblings go insane with the same disease. One is known as the primary insane and the other is known as secondary or inducted insane. Let it be assumed that a crime is committed by primary insane under partnership with secondary insane. Both are suffering from delusional ideas. The primary insane is the one who got the disease first. By virtue of closed proximity this disease got rubbed off on the other sibling. However detailed examination may reveal that the inducted insane have some episodes of sanity or clarity of consciousness.

In the above situation casuistry as a tool of redemption would be employed in order to do well to the deserving and identify the non-deserving. It would be useful while evaluating insanity, competence and dangerousness of the parties involved. The case is not as simple as it appears to be. Therefore by extrapolation the analogy would be sought.

The analogy quotable can be: two persons were raping a woman in secluded country side. An unknown person was walking through that way. He saw the rape being committed. Without a second thought he joined the party although he didn't have malicious thought beforehand. The trail court would seek to clarify the objective meeting action. The first two persons involved in sexual assault definitely had common intention. .The third man who joined later did it on the spur of the moment. Therefore imposing punishment would be different. [2]

Finally, the authors would venture to say that casuistry has a definite place in Forensic Medicine too. May be the area of extended analogy be small but judicious application of mind can bring relief and succor to the aggrieved as well as definite party.

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