

Adrenal Pheochromocytoma - A Case Presentation with Review of Literature

Nanda Patil¹, Shubhang Monpara², Pranjal Shah², Nidhi Goswami²

¹Professor, Department of Pathology, Krishna Institute of Medical Sciences, deemed to be University, Karad, Maharashtra

²Tutor, Department of Pathology, Krishna Institute of Medical Sciences, deemed to be University, Karad, Maharashtra

Corresponding Author: Pranjal Shah

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ABSTRACT

Pheochromocytoma is a rare catecholamine secreting tumor which arises from adrenal medulla and extra-adrenal sites. Its clinical presentation is sustained or intermittent hypertension often with paroxysmal symptoms. Imaging studies and estimation of catecholamines and metanephrine in a 24 hours urine sample helps to arrive at definite diagnosis. Correct diagnosis is very important because resection of tumor dramatically reverses clinical symptoms. We present a case of adrenal pheochromocytoma in a 34-year-old female patient to highlight its rarity, clinical presentation and histopathological features.

Key Words: Adrenal medulla, Catecholamine, Hypertension, Pheochromocytoma.

INTRODUCTION

Pheochromocytoma is a rare tumor of adrenal medulla which produces catecholamines. Its clinical hallmark is sustained or intermittent hypertension often in spells (1,2). This tumor can produce unmanageable hypertension. Hypertensive crisis is a life-threatening complication of pheochromocytoma. We present a case of pheochromocytoma in a 34 of year old female patient who presented with flank pain, episodes of headache and sweating as well as palpitations.

CASE REPORT

A 34 year old female patient presented with right sided flank pain since one month and episode of headache, sweating and palpitations after doing physical exertion. On physical examination, blood pressure was 170/98 mm of Hg. Ultrasound examination of abdomen revealed large, well defined, lobulated and heterogeneously

enhancing hypodense lesion in the right suprarenal region.

Urinary Vanilyl Mandelic Acid (VMA) levels were 57.15 mg/24 hours (Normal level: <6.8 mg/24 hours). VMA to creatinine ratio was 127.56 mg/g creatinine (reference value: <8 mg/g creatinine). Adrenal tumor was excised and sent for histopathological examination.

Histopathological findings – Received a right adrenal gland tumor totally measuring 11.5 x 9 x 6 cm and weighing 200 grams. The tumor was grey brown, globular mass cut section of which was yellowish soft with areas of hemorrhages. (Fig. 1 ,2). Microscopy revealed a capsulated tumor composed of round to polygonal neoplastic cells showing round regular vesicular nuclei with fine chromatin and ample amount of eosinophilic granular cytoplasm. Neoplastic cells were arranged in the form of cords and nests separated by fibrous septa and blood vessels showing Zellballen pattern. There

was no evidence of nuclear atypia or capsular infiltration (3, 4, 5). Considering the features, the tumor was diagnosed as

pheochromocytoma arising from the right adrenal gland. Postoperative follow-up of the patient is uneventful.



Fig. 1



Fig. 2

Gross examination (Fig. 1, 2, 3): grey brown globular mass. Cut section showing yellowish tumor with areas of hemorrhages.

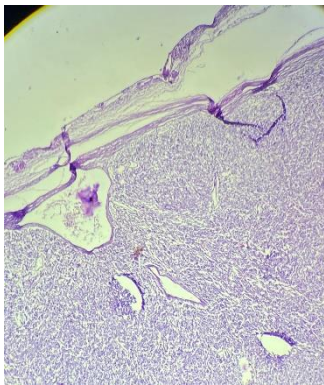


Fig. 3

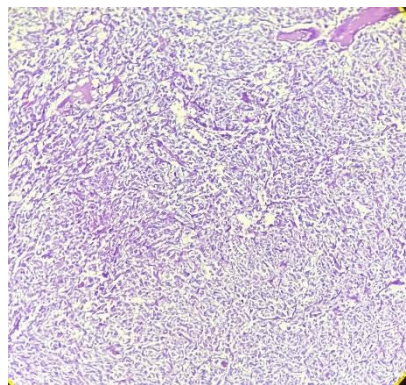


Fig. 4

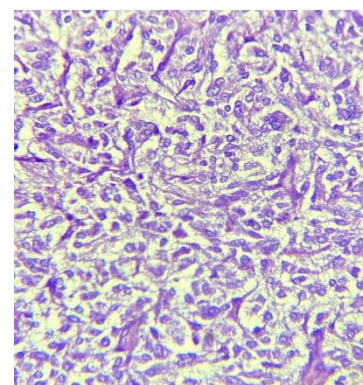


Fig. 5

Microscopic examination (Fig. 3, 4, 5): Encapsulated tumor with round to polygonal neoplastic cells showing Zellballen pattern. (40x, 100x and 400x HPE)

DISCUSSION

Pheochromocytoma is a rare tumor derived from chromaffin tissue cells mostly situated within adrenal medulla. Pheochromocytoma was first recognized in 1926 by Casor Roux and Charles Mayo (3,4). Annual incidence is eight cases per million persons. (5,6). Only 15% pheochromocytoma develop from extra-adrenal chromaffin tissue which lies in the sympathetic nervous system extending from base of skull to urinary bladder. The tumor can occur at any age. The peak incidence is between third and fifth decade of life. Majority of them are sporadic, while

16% cases present with associated endocrine disorders such as multiple endocrine neoplasm (MEN) type I, neurofibromatosis I and Von Hippel – Lindau disease. (7,8). The tumor has no sex predilection. There are no reliable clinical, biochemical or histopathological criteria to distinguish a malignant from benign pheochromocytoma (8). Clinical features of pheochromocytoma are because of increased secretion of catecholamines either intermittently or continuously (9).

Classic symptoms of this tumor is spell of headache, palpitations and diaphoresis in

association with severe hypertension precipitated by physical exertion, induction of general anesthesia and drugs like contrast media, antidepressants, opiates. These symptoms worsen as the tumor progresses (10). Similar presentation was seen in our case. Diagnosis of pheochromocytoma depends on biochemical confirmation of excess catecholamine secretion. Plasma metanephrines testing gives 96% sensitivity but it has low specificity (11). 24 hours urine catecholamine and metanephrines have 87.5 % sensitivity and 99.7% specificity. Localization of pheochromocytoma and metastasis is done with magnetic resonance imaging (12). Histopathological examination of the tumor confirms the diagnosis. The tumor varies in size and weight, average weight is 100 grams. Histology shows polygonal to spindle shaped neoplastic cells arranged in nesting, solid or alveolar (Zellballen) pattern with round nuclei and stippled chromatin. Immunohistochemistry (IHC) and electron microscopy demonstrate membrane bound secretory granules and are helpful when histology is equivocal (13,14,15). Surgical resection of the tumor is treatment of choice and results in cure of symptoms. An experienced anesthesiologist and surgeon are required, as there is risk of hypertensive crisis (7). Biochemical cure should be confirmed by assay of 24 hours urinary catecholamines 2-3 weeks after surgery.

CONCLUSION

Pheochromocytoma is a rare tumor which arises from adrenal medulla. Correct diagnosis is important because resection of the tumor drastically reverses the clinical symptoms and cures hypertension.

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Conflict of Interest: None

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