Scholars Journal of Medical Case Reports
Sch J Med Case Rep 2017; 5(11):770-771
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(An International Publisher for Academic and Scientific Resources)

ISSN 2347-6559 (Online)
ISSN 2347-9507 (Print)

Pheochromocytoma of left Adrenal gland

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Article History
Received: 08.11.2017
Accepted: 15.11.2017
Published:30.11.2017

DOI:
10.36347/sjmcr.2017.v05i11.023

Abstract: Pheochromocytomas are uncommon neoplasms composed of chromaffin cells, which synthesize and release catecholamines and in some instances peptide harmones. These tumours are important because they give rise to surgically correctable forms of hypertension. Although only about 0.1% to 0.3% of hypertensive patients have an underlying pheochromocytoma, the hypertension can be fatal when the pheochromocytoma goes unrecognised. Pheochromocytoma can occur at any age but most patients are 20 - 60 years old. Most pheochromocytomas are slow growing and benign but about 10% of the tumours are malignant, invasive and metastasising. These tumours are commonly sporadic but 10% are associated with familial syndromes of multiple endocrine neoplasias like medullary carcinoma of thyroid, hyperparathyroidism.
Keywords: Pheochromocytoma, hypertension, adrenal gland**.**

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]. Because of excessive catecholamine secretion, pheochromocytoma may precipitate life-threatening hypertension or cardiac arrhythmias. Pheochromocytoma is fascinating and challenging to clinicians because it combines lethal potential if untreated with possible long term cure in the majority if diagnosed and treated surgically. We present a case. Report of adrenal pheochromocytoma of 20 year aged young female patient presenting with paroxysomal attacks of hypertension, palpitation which was treated successfully.

CASE REPORT

A 20 years old female came with the complaints of paroxysomal attacks of palpitation, dizziness, and headache for last six months. Each attack persists for few minutes to half an hour and occurs irregularly once within two to three days to 2 to 3 times a day. On examination, patient had no abnormal physical findings except Blood Pressure (BP) is high during paroxysomal attack (Systolic BP varies from 130 to 200 mmHg and diastolic BP varies from 110 to 130 mmHg). Complete hemogram and other routine investigations were normal. Provisional diagnosis of secondary hypertension due to adrenal pheochromocytoma was made, biochemical investigations i.e. serum catecholamine level or 24 hour urinary catecholamine levels were raised. Abdominal USG and CT scan shows left sided adrenal mass of 7cm in diameter. Surgery was done and sent for Histopathological examination. We received tumour of size 7 cm in diameter, externally yellowish in colour. Finally it was diagnosed as Pheochromocytoma on microscopy.

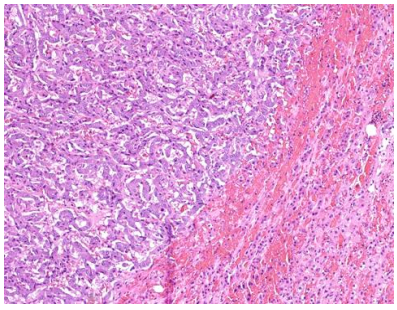

Fig-1: Section showing tumour tissue arranged in zellballen pattern with adjacent capsule (H&E,x10)

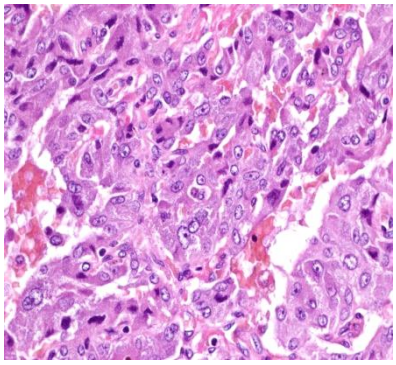

Fig-2: Section shoing round to polygonal tumour cells arranged diffuesely with prominent nucleoloi (H&E, x40)

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 paraganglion chromaffin tissue of the sympathetic nervous system extending from base of skull to the urinary bladder.

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 [2, 3]. The dominant clinical feature in patients with pheochromocytoma is ypertension. Classically, this is described as an abrupt, precipitous elevation in blood pressure, associated with tachycardia, palpitations, headache, sweating, tremor and a sense of apprehension. These episodes may also be associated with pain in the abdomen or chest, nausea and vominting. In practise, isolated paroxysomal episodes of hypertension ocur in fewer than half of patients as seen in our case. In about two thirds of patients, the hypertension occurs in the form of chronic, sustained elevation in blood pressure, although an element of labile hypertension is also present. The paroxysms may be precipitated by emotional stress, exercise, changes in posture and palpitation in the region of the tumour.

The elevation of pressure is induced by the sudden release of catecholamines that may acutely precipitate congestive heart failure, pulmonary edema, myocardial infarction, ventricular fibrillation and cerebrovascular accidents. The cardiac complications have been attributed to what has been called catecholamine cardiomyopathy, or catecholamine induced myocardial instability and ventricular arrhythmias.

Microscopically the tumour is composed of polygonal to spindle shaped chromaffin cells or chief cells, clustered with sustentacular cells into small nests or alveoli (Zellballen pattern) with abundant cytoplasm.[Figure 1 &2]

The laboratory diagnosis of pheochromocytoma is based on the demonstration of increased urinary excretion of free catecholamines and their metabolites, such as vanillylmandelic acid (VMA) and metanephrines. Isolated benign tumours are treated with surgical excision, after preoperative and intra-operative medication of pateints with adrenergic blocking agents to prevent a hypertensive crisis. Multifocal lesions require long term medical treatment for hypertension [4].

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