TAJ June 2010; Volume 23 Number 1

ISSN 1019-8555
The Journal of
Teachers Association
RMC, Rajshahi

Case Report

Adrenal Mass- A Case Report

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Abstract

Phaeochromocytomas are the rare tumors of the adrenal gland arising from the chromaffin cells of the adrenal medulla. These patients usually presents with triad symptoms of hypertension, headache and palpitation although there are some other presenting features like sweating, pallor, weight loss, hyperglycemia, nausea and psychological effects. 5 Ps- indicates Pressure (blood pressure), Pain (headache), Perspiration (sweating) Palpitation and Pallor can also be the presenting features. It is one of the surgically correctable hypertension. Diagnosis is made biochemically and localization is done by various imaging techniques such as USG, CT, MRI and I-MIBG. Extra adrenal tumors can be detected by-IMIBG. Localization is very much essential for planning of the surgery. Cytology or histopathology can not differentiate malignant from benign Phaeochromocytoma as both reveals similar features of pleomorphism, mitosis and atypical nucleus. Malignant Phaeochromocytoma can be distinguished by invasion of the adjacent structures, documenting nodal and distant metastasis. Treatment is essentially surgery either open or laparoscopic. Preoperative care is the most important part of treatment which include control of hypertension and alpha blockade adequate hydration with IV fluid. Per operative monitoring of cardiac and haemodynamic status is also very important part of treatment. HDU or ICU management may be necessary in the early post operative days.

TAJ 2010; 23(1): 67-70

Introduction

Phaeochromocytomas are catecholamine secreting adrenal tumors that arise from chromaffin cells of adrenal medulla. Extra-adrenal Phaeochromocytomas may also occur in the various sympathetic ganglia which are named as functional paragangliomas. In 1886, Frankel had reported Phaeochromocytoma first in a 18 yrs old lady with bilaterial adrenal tumor. First successful resection of Phaeochromocytoma had been done by Roax and Mayo independently in 1926 and 1927 respectively [1,2]. It is a rare tumor occuring in 2 to 8 per million people [9,7]. These tumors may occur in familial or sporadic manner and are found with increased rate in screened hypertensive populations. Peak incidence of Phaeochromocytoma occurs during the fourth and fifth decade of life and males and females are affected about equality. It is known as the 10% tumor as 10% are inherited, 10% are bilateral, 10% malignant, 10% are extra adrenal and 10% occur in children. Hereditary Phaeochromocytoma occur in several tumor syndromes like MEN-2, VHL syndrome, NF type-1 and Familial Paraganglioma syndrome.

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Case report

A55 yrs old farmer, hypertensive, smoker, non-diabetic, hailing from Meherpur, Bangladesh, presented with recurrent attacks of intractable headache, vomiting, palpitation, sweating and hotness for about 1 ½ yrs.

The headache was compressive in nature, radiates to the neck and associated with vomiting. Vomiting is non projectile, bitter in taste, contains recently taken foods and no history of induced vomiting. Severe palpitation, sweating, flushing and hotness usually accompany the headache and vomiting during each episode which make the patient restless. Hotness was so severe that it makes the patient bound to remove his clothes and roll over the ground. He has given history of about 25 such episodes in last $1^{1}/_{2}$yrs. Some of these episodes were so severe that the patient became frightened for impending death. He has given history of taking NSAIDs, anxiolytics prescribed by local quacks but nothing seems to be useful to remove the symptoms completely.

None of his family members or relatives are suffering from such type of illness. With this complaints patient admitted in Rajshahi medical college hospital for better management.

General examination was essentially normal except tachycardia and hypertension. His pulse was 112/m regular and BP was 200/100 mm Hg.

An ill defined mass was palpable in the right lumbar region which did not move with respiration.

Other systemic examinations were normal.

USG and CT scan was done which suggest, “Supra Renal Mass” in the right side.

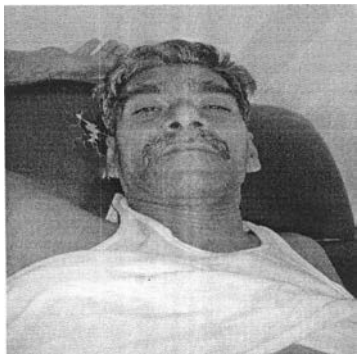

Fig. Picture of the Patient

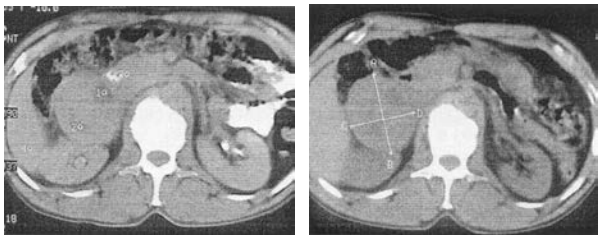

Fig. CT Scan of abdomen showing Rt adrenal mass

Urinary VMA (24hrs) reveals 32.62mg/day which was 4 times of normal. At this stage, we had assessed the operability & decided to do surgery.

After control of hypertension and adequate alpha-blockade, preoperative rehydration was done with IV fluids. Then right open adrenalectomy done on 22/10/2010.

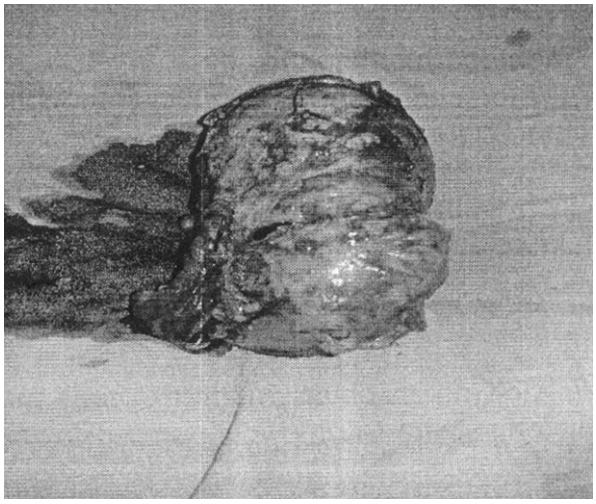

Fig: Resected Rt Adrenal gland.

During operation cardiac and haemodynamic complications (extreme tachycardia-180/min, sudden rise and fall of BP-210/120 mmHg to 90/60 mmHg) occurred and managed carefully. Postoperative period was eventless and patient was discharged on 10^{th} POD after doing 24 hrs urinary VMA (7.58mg/day) which was within normal range. At the time of discharge, patient was normotensive and free of symptoms.

Discussion

Phaeochromocytomas are catecholamine secreting adrenal tumors which causes elevation of the blood pressure which may range from mild hypertension a dramatic hypertensive crisis which

is the most consistent manifestation of this disorder. Hypertension is sustained in 50% of the patients, is paroxysmal in 30%, and absent in 20% of the patients[3]. Other symptoms include headache (60-90%), sweating (50-70%), palpitation (50-70%), pallor (40-45%), weight loss, hyperglycaemia, nausea and psychological effects.

Phaeochromocytomas may be hereditary which occur in association with several tumor syndromes such as MEN-2, VHL syndrome, NF type-1, Familial paraganglioma syndrome. So whenever family history is positive one should search for other hereditary tumors.

Demonstration in creased plasma or urinary levels of catecholamines and their metabolites is the sine qua non for the diagnosis of Phaeochromocytoma. More than 90% of the patients with phaeochromocytoma have distinctly elevated levels of catecholamines in 24 hrs urine. Urinary VMA is the least specific test because false positive finding may result from ingestion of coffee, tea, raw fruits or drugs such as alpha methyl dopa.

Recently, the estimation of plasma fractionated metanephrines (metanephrine and normetanephrine) has become available as a screening test for detection of phaeochromocytoma[4,5]. It is highly sensitive for detecting phaeochromocytoma and is simple to perform.[6] False positive results are common with this test.

CT scan and MRI are the two radiologic modalities of choice to localize phaeochromocytoma. CT usually detects tumors of 1 cm and larger with reported sensitivity of 77% to 87% and a specificity of 96% to 100%. This test is most useful in localizing extra adrenal tumors not seen in the conventional imaging and a maligant phaeochromocytomas.

Preoperative management of the patients with phaeochromocytoma centers on:

1. Adequate control of hypertension.
2. Adequate alpha blockade to prevent intra-operative hypertensive crisis due to tumor manipulation and release of catecholamines.
3. Fluid resuscitation to prevent circulatory collapse after removal of the tumor.

Alpha adrenergic blockade is achieved with phenoxybenzamin starting at a dose of 10mg twice daily. The dose increased by 10 to 20mg per day until hypertension and symptoms are controlled and the patient demonstrates mild postural hypotension. Beta adrenergic blockade is indicted in patients who develop tachycardia with an alpha blockade or who have tachyarrhythmia.

Patients with phaeochromocytoma can be expected to have blood pressure volatility and high fluid volume requirements during and immediately after surgery. All patients should have additional hydration by pre-operative I/V fluid administration on for 12 to 24 hrs before surgery to avoid cardio-vascular collapse after removal of the tumor.

Now a days, CT, MRI & I0MIBG permit preoperative localization of the tumor in >95% of cases. In most cases, phaeochromocytomas re laparoscopically resectable[9,10]. Regardless of approach, important common principles include-

1. Minimal handling of the tumor.
2. Early isolation of the adrenal vein and
3. Avoidance of capsular rupture.

Immediate after resection of phaeochromocytomas, profound hypotension as a result of vasodilation may occur. Aggressive fluid resuscitaitonh is necessary to expand intravascular volume. HDU or ICU support may be necessary in the early post operative period. Monitoring of serum glucose concentration should also be carried out because of rise of hypoglycemia owing to rebound hyper-insulinemia from catecholamine induced suppression of insulin secretion.

Biochemical cure should be confirmed by assessment of serum catecholamines 2-3 weeks post operatively. Life-long yearly biochemical test should be performed to identify recurrence, metastasis or metachronous phaeochromocytomas.

Conclusion

Experienced surgeon, experienced anesthesiologist & preoperative preparation of the patient by adequate α-blockade & rehydration is the most important pre-requisite in the surgical treatment of phaeochromocytomas. Early post operative period

is also very crucial for the development of circulatory collapse and hypoglycemia. So, HDU or ICU management may sometimes be necessary at this period.

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