

A giant malignant pheochromocytoma of the adrenal gland: biologic time bomb

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Citation

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Abstract

BACKGROUND: Pheochromocytoma is a rare tumor of adrenal medulla and only 5-10% of pheochromocytoma are malignant. Surgical resection is the only mode of treatment. **Methods:** A case report with review of literature is presented in brief. **Results:** Overall prognosis in benign case: 5 yr survival rate is >90% and in malignant case: about 40%

CASE REPORT

A 40 yr old female came to the outpatient department with complain of upper abdominal pain since 1 year and vomiting since 10 days. She was a known case of hypertension and diagnosed to have pheochromocytoma at a local place and referred to our hospital for further management. She had a history of headache. No other significant history available. She had been advised to undergo routine blood investigations plus investigations to rule out other endocrine problem. At the time of admission her blood pressure was 170/100 mm hg in supine and 160/100mmhg in standing position. Tender vague lump was palpable in the right hypochondriac region. All biochemical investigations were normal except VMA level in the urine was 55mg/24hr(normal:0.7-6.8mg/24hr).CT scan abdomen shows fairly large heterogeneously enhancing mass lesion of size 12.8 X 8.0 X 13.0 cm in the region of Right adrenal mass is displacing surrounding structures aorta to left, IVC anteriorly and causing compression Liver, pancreas, are displaced anteriorly Findings suggestive of Right adrenal mass lesion possibly pheochromocytoma .ECG shows T inversion in antero lateral leads. ECHO was normal. Patient was kept on alpha blocker followed by addition of beta blocker (beta blocker was added after 1 week of prazosin).total 14 days treatment was given along with improvement of general nutritional status. Patient was optimized for surgery. On the day prior to surgery, Antibiotics, MgSo4 powder (20 g) orally, IV fluids were started, on the day of surgery, fresh ECG, electrolytes were repeated, which was normal. Epidural catheter, CVP line, arterial line was taken for monitoring. After induction, abdomen was opened through midline incision, abdomen

was checked for evidence of metastases .on exposing the tumor, it was found to be dark brown in color, very vascular and densely adherent to surrounding major vessels like aorta, IVC (photo 1). The tumor was freed from the surrounding structure (photo 2).

Figure 1

Figure 1 tumor surrounding major vessels

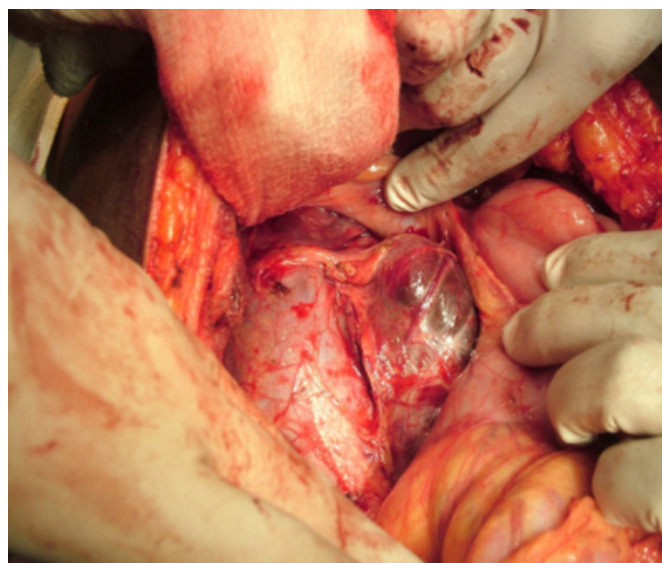
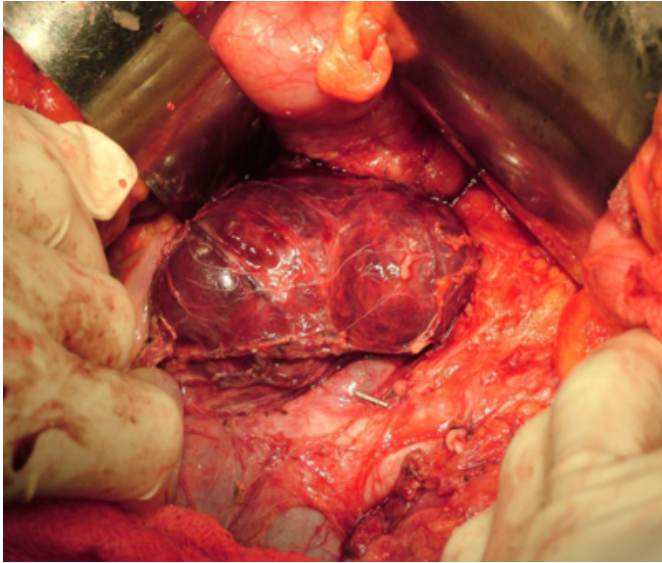


Figure 2

Figure 2 tumor freed from major vessels



During manipulation of the tumor, BP shot up to 240/ 140 mm Hg Na Nitroprusside @ 10 - 20 µg/kg/min , MgSO₄ 1.5g bolus followed by infusion of 1g/hr, Metoprolol 0.5mg was administrated intermittently to control tachycardia . During ligation of the veins, the patient develop sudden hypotension (BP: 50 systolic) , IV Phenylephrine 1mg was given. After removal of tumor, the patient develop profound hypotension (systolic BP 40 mm Hg) , increments of IV Phenylephrine and Dopamine infusion@10µ/kg/min. After the tumor resection, all the vasodilators, beta blockers, MgSO₄ were stopped. The size of the tumor was 18cm×12cm in size (photo 3). On cut section tumor shows areas of hemorrhage and necrosis and cyst (photo 4).

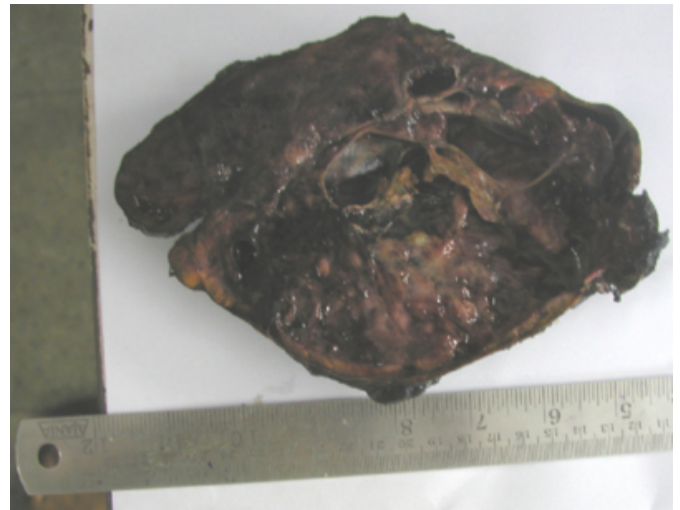
Figure 3

Figure 3 gross macroscopic view



Figure 4

Figure 4 cut section



Depending on the size of the tumor, gross macroscopic appearance and histopathological finding diagnosis of malignant pheochromocytoma was made. Post-operative course was uneventful. post-operative catecholamine level was normal and blood pressure was under controlled.

DISCUSSION

Pheochromocytoma is a rare catecholamine secreting tumor of adrenal medulla derived from chromaffin cells. The term derived from Greek word phaeos means dusky , chroma means color and cytoma means tumor .tumor may be found anywhere in the body from neck to pelvis. The tumor which develops outside adrenal medulla but anywhere in the sympathetic chain from chromaffin cells is known as extra

adrenal pheochromocytoma. Most common site for extra adrenal pheochromocytoma is Organ of Zuckerkandl. César Roux and Charles Mayo performed first successful surgical resection of pheochromocytoma in 1926¹. incidence is up to 0.2% in hypertensive individual. It can affect any age group but peak incidence in 30-50 yrs of age group. They occur with roughly equal frequency in both sexes. 80-90% of pheochromocytomas are found in the adrenal medulla and 3% are extraabdominal, 10-20% are bilateral and 10-20 are familial. 5-10% are malignant and found to be associated with familial variety. They are associated with MEN type IIa, MEN type IIb, von Recklinghausen's disease². familial variety usually present in childhood and likely to be multiple and bilateral. Symptoms are attributable to effect of circulating catecholamines. Classical presentation includes triad of headache, diaphoresis, and tachycardia in a patient with known case of hypertension. The presence of all three symptoms, in presence of hypertension, makes pheochromocytoma diagnosis likely. Hypertension is the most consistent feature and is present in more than 90% case. It may be sustained or paroxysmal. Paroxysms may be precipitated by physical training, surgery, pregnancy, urination, and numerous drugs and agents (tricyclic antidepressant, opiates, dopamine antagonist etc), child birth, induction of general anesthesia. Episode may be associated with anxiety, pallor, tachycardia, palpitation. Most attacks will not last for more than hour. The choice of diagnostic biochemical test is plasma metanephrine (sensitivity: 95%, specificity: 85%)³. Therefore primary role of plasma metanephrine level is to exclude pheochromocytoma when test is normal. When positive, confirmation with 24 hr urinary catecholamine and their metabolite is required. Two 24-hour urine collections for catecholamine and their metabolites are sufficient to make (or exclude) the diagnosis of pheochromocytoma in almost

all cases. Provocative and suppression test are rarely performed now a days. Biochemical diagnosis should be followed by radiological diagnosis. CT and MRI are equally sensitive but MRI is preferred because contrast media used for CT can provoke episode. Tumor can also be localized by ¹²³I MIBG scan, can be performed when multifocal disease is suspected. Only 10% of tumors are malignant. The diagnosis is based on clinical or macroscopic evidence of local invasion or the presence of distant metastases. In most patients' surgical resection is the most appropriate treatment after controlling blood pressure with alpha and beta blockers to prevent cardiovascular complications including hypertensive crisis. Intra operative use of alpha blockers followed by aggressive volume replacement and alpha adrenergic agonist in the immediate post-operative period is recommended⁴. In case of malignancy when ever possible surgical resection or debulking should be tried as it is the only chance for cure. Even in absence of cure, resection in malignant pheochromocytoma is of significant palliative benefits⁵. Overall 5 yrs survival rate for benign is >95% and for malignant is about 45%.

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