

CASE REPORT

CLINICALLY SILENT GIANT PHEOCHROMOCYTOMA: A CASE REPORT

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ABSTRACT: INTRODUCTION: Adrenal masses discovered by imaging techniques for reasons unrelated to adrenal diseases are documented as incidentalomas with pheochromocytoma accounting for about 20% of adrenal masses. Clinically silent, biochemically negative Pheochromocytomas are rare. Close intra and postoperative monitoring is the key to success while operating on a mass lesion in suprarenal area.

CASE REPORT: A 38 years female patient presented with dull aching pain & lump in left hypochondrium and left lumbar region of four months duration. Patient was normotensive and did not have any comorbid conditions. Abdominal examination revealed a smooth surfaced, firm, ballotable lump of 14 X 10 cm in the left lumbar region. Adrenal function tests including urinary and plasma catecholamines and metanephrine were within normal limits. CT scan of abdomen revealed a left retroperitoneal mass arising from left adrenal gland of benign nature. Left adrenalectomy was performed. Per operatively patient had unexpected hypertensive episode which was managed. Histopathological examination was reported as Benign Pheochromocytoma.

CONCLUSION: Clinically silent pheochromocytoma is a rare condition and with normal catecholamines and metanephrines is even rarer. Possibility of pheochromocytoma should be kept in mind while operating on a mass in the suprarenal region even in a normotensive patient with normal hormonal parameters so that operating team can deal with an unexpected hypertensive crisis. Adrenalectomy should be done for such patients because of concern of malignant transformation and or development of subsequent functional abnormality.

INTRODUCTION: Adrenal masses discovered by imaging techniques for reasons unrelated to adrenal diseases are documented as incidentalomas with pheochromocytoma accounting for about 20% of adrenal masses.^[1] The most frequent symptom of the pheochromocytoma is hypertension and about 90% of cases exhibit it. The literature indicates that incidental pheochromocytoma cases that are smaller than 1 cm have no clinical symptoms. It is extremely rare for large pheochromocytoma case, not to show any clinical symptoms. No single or combination of biochemical parameters can exclude or establish presence of pheochromocytoma with certainty.

CASE REPORT: A 38 years female patient presented with dull aching pain in left hypochondrium and left lumbar region of four months duration. Simultaneously she also noticed a lump in left lumbar region. Patient had no dyspeptic symptoms or urinary complaints. She was not a known hypertensive and did not have any comorbid conditions. She had three children, youngest being 12 years old. Patient was afebrile, pulse rate was 76 per minute and regular and blood pressure was 130/80 mmHg. Pallor was detected. Abdominal examination revealed a 14 x 10 cms smooth surfaced, firm, ballotable lump in left hypochondrium, extending into left lumbar region. All haematological

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investigations were normal except for Hb%, which was 5 gm%. Adrenal function test including urinary and plasma catecholamines and metanephrine were within normal limits. Contrast Enhanced CT Scan of abdomen showed a large, well defined solid retroperitoneal mass in left suprarenal region measuring 10.5 x 9.3 x 8.4 cms with heterogeneous density, showing heterogeneous enhancement in contrast with multiple central nonenhancing hypodense areas of necrosis. The mass was seen to extend from D₁₂ upper margin upto lower margin of L₂, abutting the left lateral margin of abdominal aorta and extending anteriorly upto abdominal wall. It was seen to displace and rotate the left kidney inferiorly and laterally with preserved intervening fat plane of cleavage. Left adrenal was not seen separately from the mass. There was no invasion of adjacent organs. Splenic vein and left main renal artery were displaced superiorly and inferiorly respectively without evidence of invasion. Radiologically it was a left retroperitoneal mass arising from left adrenal gland of benign nature. [Fig. 1 & 2].

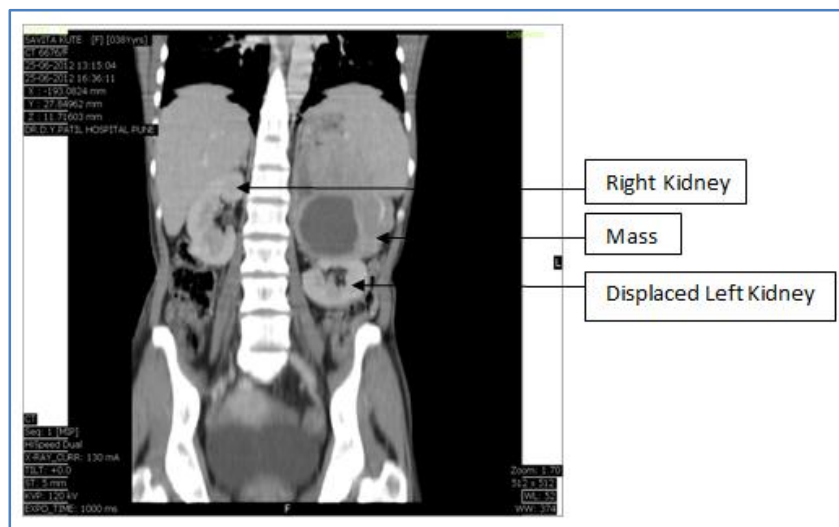


Fig. 1: Coronal reformatted image of post contrast CT Scan

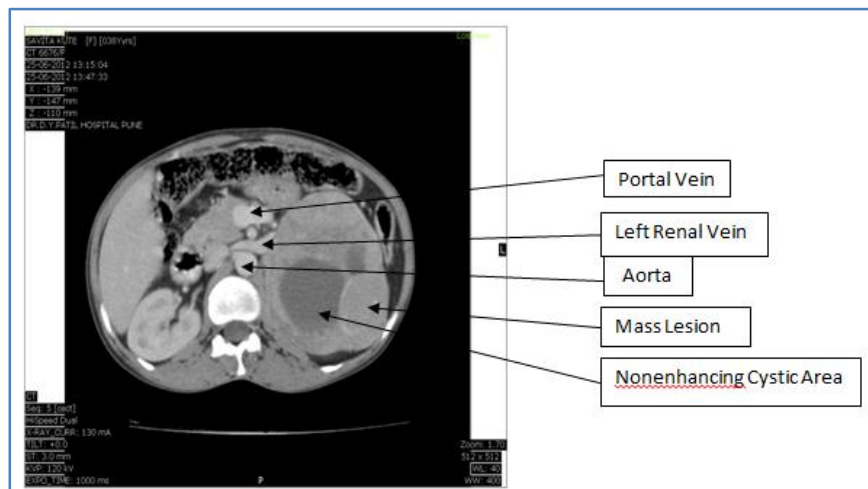


Fig. 2: Axial image at the level of transpyloric plane showing epicenter of the mass

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On exploratory laprotomy a well encapsulated tumour, measuring 12cms x 10cms was seen in left supra-renal gland region, pushing left kidney down and displacing body and tail of pancreas upwards. [Fig. 3].

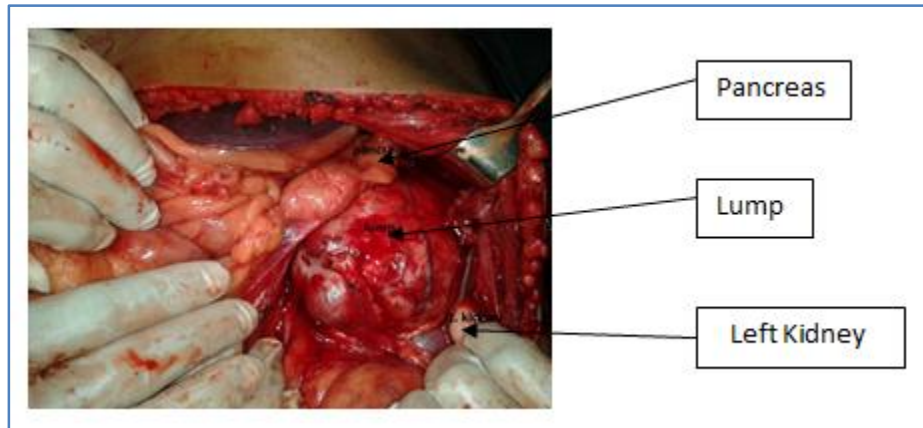


Fig. 3: Intraoperative photograph showing adrenal mass

Left adrenalectomy was performed. Weight of the mass was 146 grams.

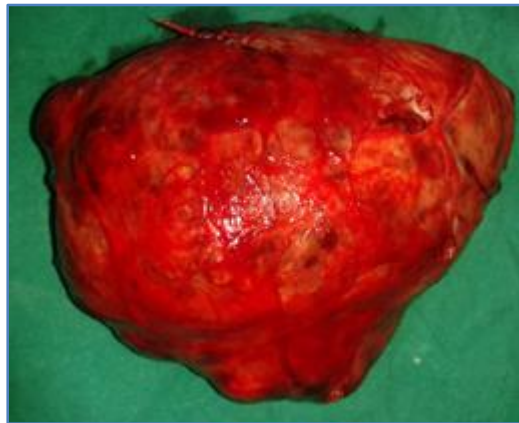


Fig. 4: Excised adrenal mass

Patient had sudden episode of hypertension as soon as the tumour mass was handled. Blood pressure went up to 200/110 mmHg, which was controlled by inj. Esmolol and inj. Labetolol. After excision of mass, there was sudden drop in blood pressure to 80/50 mmHg. Inotropes were used to maintain normal B.P, which took 3 hours to come to normal. Inotropic support was gradually tapered and withdrawn after 48 hours as the B.P. was maintained at normal level after that. Patient was discharged on tenth post-operative day. Histopathological examination was reported as Benign Pheochromocytoma.

Patient had been followed up for six months so far and is asymptomatic.

DISCUSSION: Pheochromocytomas are catecholamine-producing tumors of neuroectodermal origin identified by the presence of cells with positive chromaffin stain.^[2] The tumor may occur in patients of any age and with equal frequency in both sexes.

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Pheochromocytomas may be classified as sporadic or familial. Most pheochromocytomas are sporadic. Our patient had no features suggestive of it being part of familial MEN2A or MEN2B syndromes.

In general, 10% of these tumors are extra-adrenal, 10% are malignant, 10% occur bilaterally, 10% are inherited as an autosomal dominant pattern and 10% occur in pediatric patient.^[3]

Patient with pheochromocytoma are typically symptomatic. The classical triad of symptoms of pheochromocytoma consist of headache, palpitation and diaphoresis. Hypertension is the most common sign of pheochromocytoma and 90% to 100% of cases exhibit it. None of the above features were demonstrable in our patient, who was normotensive. The incidence of unsuspected pheochromocytoma in absence of classical symptoms has been reported to be about 30%.^[4]

Literature indicates that incidental pheochromocytoma cases that are smaller than 1cm have no clinical symptoms but rarely some large pheochromocytoma cases do not show any clinical symptoms, as observed in our case.^[5,6]

Persistent hypertension is seen in half, paroxysmal hypertension in third and normal blood pressure in less than fifth of patients. More than 90% of patient presented with at least two of the three symptoms in the classical triad.^[7] Completely silent pheochromocytoma are extremely rare.⁷ Our case although presented with an abdominal mass was normotensive with no feature from amongst the classical triad of symptoms.

Patient did not exhibit any abnormality of adrenal cortical or adrenal medullary function. Sometimes pheochromocytomas do not secrete enough catecholamines to produce positive test results or typical sign and symptoms. Our patient remained normotensive throughout the course of her hospitalization except during Intraoperative period hypertensive episode followed by hypotensive episode, extending briefly into immediate postoperative period. It resolved with immediate appropriate treatment.

Functional tumors are usually small when detected because of dramatic symptoms whereas nonfunctional tumors may be large.^[8] Crout and Sjoerdsma found that pheochromocytomas 50gm. or larger are often asymptomatic because secreted catecholamines are metabolized within the tumor.^[9]

In contrast, tumors small than 50 gm. have slow turnover rates and release free catecholamines into the circulation, exhibiting persistent symptoms and signs.^[9] In our case, patient's adrenal tumor was large in size, weighed 146 grams and was clinically silent.

Biochemical tests are diagnostic in most of the cases but in a small number they can be normal despite the presence of pheochromocytoma.^[10]

Diagnosis of pheochromocytoma is usually based on increase in plasma and urinary levels of catecholamines and its metabolites. According to Poraco et al, measurement of metanephrines in the urine is the most useful test.^[11] Urinary and plasma metanephrines levels were found to be normal in our case.

Recent studies have shown that plasma metanephrines are the most reliable tests to identify pheochromocytoma with sensitivity approaching 100%.^[3]

In asymptomatic patients, normal values of plasma catecholamines do not rule out the disease.^[11] A multicentre cohort study examined the diagnostic utility of several biochemical tests in 214 patients with proven pheochromocytoma (out of which 138 were sporadic cases) and in 644 patient in whom diagnosis was suspected but excluded.^[12] In this study measurement of plasma free metanephrines was clearly superior to plasma catecholamines in the diagnosis of sporadic and

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hereditary pheochromocytoma. However the test specificity of plasma free metanephrines was only 82% in sporadic pheochromocytoma. In sporadic pheochromocytoma urinary catecholamines had excellent sensitivity (97%) and total metanephrines the best specificity (89%). These authors recommended against use of multiple biochemical tests to exclude pheochromocytoma and favoured single test of plasma free metanephrines.

However in 109 confirmed cases in whom all tests were performed, authors observed that assays of plasma catecholamines and urinary metanephrines have the lowest false negative rates (7%) and urinary norepinephrine and epinephrine the next higher (14%).^[11] Urinary VMA have high false negative rates(41%) and should not be used for screening purposes. Contrary to above mentioned observations the authors recommend that for 100% diagnostic accuracy multiple tests be performed.^[11] The availability of tests in any given center will necessarily determine the nature of the investigation in an individual patient, and debate over the relative merits of various tests will continue.^[11]

Van Heerden and his associates reported that the abdominal CT scan is a highly accurate and non-invasive tool for localisation of pheochromocytoma.^[13] CT and MRI are equally sensitive (98% and 100% respectively) but have lower specificities of 70% and 67% respectively.

Patients with pheochromocytoma, even when clinically silent, are at risk for a hypertensive crisis and should be treated theoretically in the same way as that with symptomatic disease. Adrenalectomy is treatment of choice.

CONCLUSION: Clinically silent pheochromocytoma is a rare condition and with normal catecholamines and metanephrines is even rarer. Possibility of pheochromocytoma should be kept in mind while operating on a mass in the suprarenal region even in a normotensive patient with normal hormonal parameters so that operating team can deal with an unexpected hypertensive crisis. Adrenalectomy should be done for such patients because of concern of malignant transformation and or development of subsequent functional abnormality.

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