A 43 YEARS OLD LADY WITH SILENT PHEOCHROMOCYTOMA: A CASE REPORT

Abstract

Improvements in imaging studies have resulted in an increase in incidentally discovered adrenal tumors. The adrenal incidentaloma was found in at least 2% on abdominal CT imaging and pheochromocytoma was reported to be about 5.1 to 6.55% among these patients. Here we report a case of silent pheochromocytoma presented as an incidentally discovered adrenal mass on abdominal sonogram. In our case patient had no apparent symptom and sign of pheochromocytoma except mild hypertension. Incidental findings in USG reveal retroperitoneal mass located in right adrenal gland. Finally CT scan of abdomen confirmed adrenal mass. Surgical excision the right adrenal gland was performed with pathological report as a benign pheochromocytoma. Majority of incidentally discovered adrenal masses are non hypersecretory benign adenomas; but a hormone screening evaluation is necessary to reveal cases with clinically unsuspected hypersecretory nature. For prevention of possible lethal outcome, patients with silent pheochromocytoma scheduled to undergo surgery should be treated according to the hemodynamic parameters.

Key Words: Incidentaloma, Pheochromocytoma, Adrenal tumor

Introduction

With the advancement of imaging like high-resolution ultrasonography (USG), computerized tomography (CT) and magnetic resonance imaging (MRI), incidentally discovered adrenal tumors are more frequently reported including pheochromocytoma. Due to the limitation of USG imaging, CT has been used for better technique to diagnose the adrenal masses and it is reported to identify adrenal masses as small as 0.5 cm or small in diameter. The prevalence of adrenal masses is at least 2% on CT scan in comparison to other suspected adrenal pathology. Among patients with adrenal incidentalomas, approximately 5.1% to 6.5%, even up to 23%, proved to have pheochromocytomas. And 10% of adrenal pheochromocytomas present as adrenal incidentalomas, even when clinically silent.

Pheochromocytomas are rare tumors found in less than 1% of the populations with hypertension. Although the majority of patients are symptomatic, 10-30% of pheochromocytomas are clinically silent. Identification of the hormone activity and malignant potential among patients with adrenal incidentalomas are clinically important. Failure to recognize and treat a pheochromocytoma could prove a fatal oversight.

Case Report

A 43-year-old woman, hypertensive but with well controlled blood pressure on drug amlodipin for 10 yrs came to outpatient department of Sarkari Karmochari Hospital, Dhaka with mild discomfort in abdomen and back pain for 2 months. On examination- there was no oedema, no apparent body weight change, not
much hair loss or hirsutism, no purple striae on abdomen or limbs were noted, except mild tenderness in abdomen. At OPD blood pressure (BP) was 125/75 mmHg, heart rate (HR) was 80/min and respiratory rate (RR) was 18/min. On routine investigation USG revealed a fairly large echogenic retroperitoneal mass located in the region of right Adrenal gland. The mass is measuring about 5×4 cm in size with central cystic changes and internal necrosis at the suprarenal area, displacing and compressing the adjacent right kidney and right adrenal tumor was suspected. Abdominal CT was preformed and identified an right adrenal tumor (5×5 cm), large lobulated mixed dense lesion having soft tissue and fluid density with central necrosis, compressing and displacing the right kidney inferomedially and IVC anteriorly. On post contrast scan the lesion shows mild inhomogenous enhancement having large central non enhancing area represent necrosis and left adrenal gland appears normal (Fig 1 and Fig 2).

On the other hand, a series of biochemical tests and hormone study were performed. Laboratory data revealed RBC: 3.45×10^6/L, Hb: 11.2 g/dL, Hct: 28%, WBC: 8200/μL, platelet count 284000/μL, ESR 74 in 1st hour. BUN: 7.5 mg/dL, creatinine: 0.81 mg/dL, blood sugar (FBS): 92 mg/dL, sodium: 141 mg/dL, potassium: 4.38 mg/dL, calcium: 8.0 mg/dL, 24 hours urinary V.M.A 7.6 mg/24 hrs (normal < 13.6 mg/24 hours), plasma renin cortisol level at 8 am: 4.57 μg/dL (normal 4.4 to 22.6); 24-hour urine metanephrine and normetanephrine were not done. Thyroid hormone tests revealed euthyroid status.

Under the impression of silent pheochromocytoma right sided laparoscopic adrenalectomy was done. No apparent complication was noted per and post operatively. After operation gross pathology of the mass demonstrated a large tumor (20×16.0×8 cm) which was well-demarcated, brownish and hard, cut surface was spongy with areas of necrosis.

Microscopic examination revealed a neoplasm showing marked fixation artifact, composed of polygonal cells having moderate to abundant cytoplasm. Some of the cells showed clear cytoplasm and some amphophilic cytoplasm. The stroma was fibrovascular. There was well...
delineated surrounding capsule. A few cells showed intranuclear pseudo inclusions consistent with pheochromocytoma of the Adrenal Gland (Fig 3) and a Scaled Score was used (PASS <4), to confirm its benign fashion.7

**Fig 3:** The tumor cells are polygonal with finely granular and eosinophilic cytoplasm. Nest of large uniform cells with rounded nucleoli arranged in Zellballen pattern was noted. The nuclei have a salt and pepper chromatin, which is characteristic of neuroendocrine tumors. Also of noted is the relative lack of mitotic activity and nuclear pleomorphism.

A focus of capsular invasion was noted while there was no further evidence of malignancy. Besides, for screening of the possibility of multiple endocrine neoplasia (MEN syndrome) type II, serum level of calcium and thyroid sonography were performed which showed normal serum calcium level and no evidence of thyroid nodule or hyperplasia. There was no family history of pheochromocytoma or MEN type II. Two days after operation, she was discharged.

**Discussion**

Pheochromocytomas are catecholamine-producing tumors of neuroectodermal origin identified by the presence of cells with positive chromaffin stain. The tumor may occur in patients of any age with equal frequency in both sexes. In general, 10% of these tumors are extra-adrenal, 10% are malignant, 5% occur bilaterally, and 10% are inherited as an autosomal dominant pattern.8-9. Diagnosis of pheochromocytoma is usually done by its clinical symptoms. Hypertension is the most common clinical manifestation of pheochromocytoma and is present in 90-100% of patients. Sustained hypertension is seen in half, paroxysmal hypertension in a third, and normal blood pressure in less than a fifth of patients.10 The classical triad consisted of headache, palpitation and diaphoresis. Less common symptoms include tremor, angina, nausea, and mass effect from the tumor. Completely silent pheochromocytomas are reported rarely.9-11 In our case, she was hypertensive controlled on single drug (tablet amlodipine) but there was no symptom suggestive of pheochromocytoma except occasional abdominal discomfort for last 3 months. Ultrasogram reveal there was a mass in the region of right adrenal gland which was supported by CT scan and histopathology report. Therefore, even when clinically silent, patient with an incidentally discovered adrenal mass, should be carefully screened for the possibility of functional activity of pheochromocytoma, the diagnosis is made with the demonstration of elevated level of circulatory urinary catecholamines or metabolites. Typically, a measurement of urinary catecholamines or metabolites that is two or three times above the upper limit of normal is considered diagnostic of pheochromocytoma.11 Because of the dramatic symptoms; functional tumors are usually small when detected, whereas nonfunctional tumors may be large.12 For pheochromocytomas, the mean diameter of incidental tumors was also significantly larger than that of symptomatic tumors.13,14 The size and appearance of an adrenal mass on CT or MRI may help distinguish between benign and malignant tumors. Furthermore, 113 metaiodobenzyl guanidine (MIBG) can be useful in determining the functional character of a tumor and locating occult secondary or metastatic lesion.15 Crout and Sjördsm found that pheochromocytomas 50 gm or larger are often asymptomatic because secreted catecholamines are metabolized within the tumor.13 In contrast, tumors smaller...
than 50 gm have slow turnover rates and release free catecholamines into the circulation, exhibiting persistent symptoms and signs. As in our case, her adrenal tumor is large in size, though weight is less than 50gm, and clinically silent. The imaging of adrenal tumors may help us to distinguish the pheochromocytoma from other tumors. In our case, to determine whether the patient with adrenal incidentaloma has subtle evidence of presence of functional tumor such as Conn’s disease, Cushing’s syndrome, pheochromocytoma or virilizing or feminizing tumors, serum cortisol level with diurnal rhythm were checked. Combining the history, physical finding, and the biochemical study, the possible diagnoses of Conn’s syndrome, subclinical Cushing’s syndrome or virilizing or feminizing tumors were initially excluded. With the improvement and more frequent use of imaging study, the incidentally discovered pheochromocytoma may not be a rare finding as we previously thought. Although majority of adrenal incidentalomas are nonhypersecretory benign adenomas, a substantial percentage among these incidentalomas are hormonally active tumor and less frequently, adrenocortical carcinomas, in which early diagnosis and treatment is critical. For prevention of possible lethal outcome, patients with silent pheochromocytoma scheduled for surgery should be closely evaluated and treated according to the hemodynamic parameters and cardiac function.

References