Incidental pheochromocytoma presenting with sublaboratory findings in asymptomatic suprarenal masses: a case report

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Abstract

Adrenal incidentaloma are described as adrenal lesions that are diagnosed incidentally during ab- dominal laparotomy or any other abdominal screening without prior suspicion of adrenal disease. It is important to diagnose adrenal lesions to know whether they are hormonally active or
malignant .A small group of 5–7% of the functional ones can exist as pheochromocytoma. Pheochromocytomas (PCC) are rare type of neuroendocrine tumors of the adrenal medulla that arise from chromaffin cells. These cells are derived from neural crest and are innervated by the splanch- nic nerve of the sympathetic nervous system which releases acetylcholine that in turn binds to nico- tinic acetylcholine receptors of the adrenal medulla causing the release of catecholamines. The dopamine, norepinephrine, and epinephrine released from these tumors are causing the episodic hy- peradrenergic symptoms seen in these cases such as hypertension, palpitations, and headaches.

This case report discusses about the incidental finding of a unilateral PCC in a 21-year-old woman who initially presented to our emergency department complaining of epigastric pain and hyperten- sion .

INTRODUCTION

Adrenal incidentaloma are described as adrenal lesions that are incidentally diagnosed during ab- dominal laparotomy or any other abdominal screening without prior suspicion of adrenal disease. It is important to

diagnose adrenal lesions to know whether they are hormonally active or malignant .A small group of 5–7% of the functional ones can exist as pheochromocytoma. 3

Pheochromocytomas are rare neuroendocrine tumours originating from the adrenal medulla1,2. Classically, these catecholamine-secreting tumours have been termed the !10% tumour," in which 10% are malignant, 10% are bilateral, 10% are paediatric, 10% are familial, and 10% are extra- adrenal. 1. Extra-adrenal pheochromocytomas are usually described as paragangliomas.Familial causes include multiple endocrine neoplasia type IIA, multiple endocrine neoplasia type IIB, neu- rofibromatosis 1 and von Hippel-Lindau disease.1

They may present in a number of ways from an incidental finding to that of the classical triad of episodic headache, tachycardia and sweating due to hypersecretion of cate- cholamines resulting in episodic hypertension[2)

Malignancy in pheochromocytoma is very difficult to diagnose microscopically. Therefore, malig- nant pheochromocytomas are diagnosed by the presence of local invasion or metastatic disease. When they are metastatic, the most frequent locations are bone, lymph nodes, liver, lung, and brain.1

Ten percent of pheochromocytoma cases are diagnosed incidentally during computed tomography (CT) or magnetic resonance imaging (MRI) screenings for other causes.3

The literature suggests that incidental pheochromocytoma cases that are smaller than 1 cm have no clinical symptoms. Rarely, some of the large pheochromocytoma cases do not show any clinical symptoms, and it is difficult to diagnose very small pheochromocytoma cases.

This case is reported to increase the suspicion index of malignant pheochromocytoma in the case of epigastric pain with hypertension in young women.

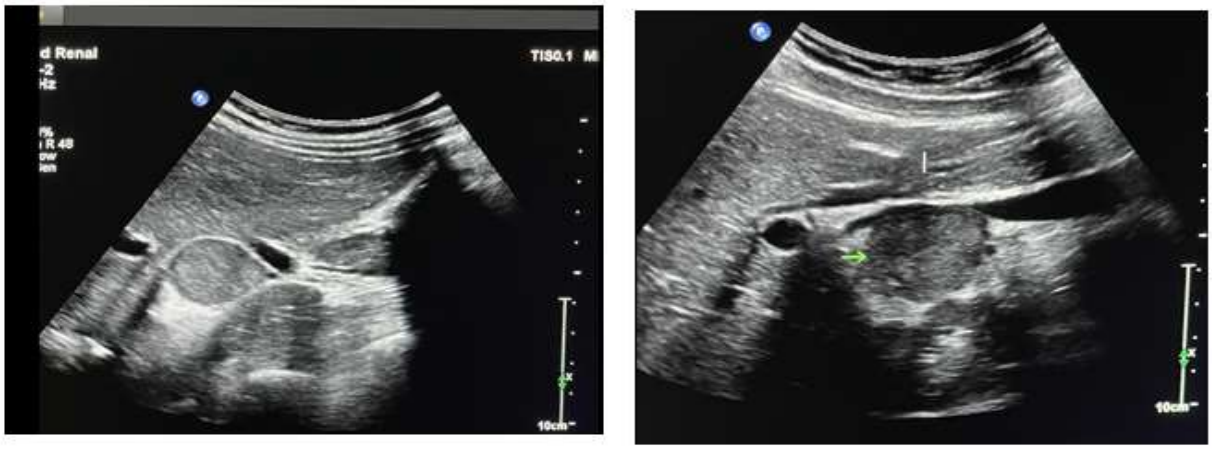

Fig 1A and Fig 1B. Sagittal and axial view: ultrasonography of right hypogastrium showing hypoe- choic mass lesion .(green arrow)

PRESENTATION OF CASE

An 21 year old female presented with a history of epigastric pain ,dyspnea, constipation , abnormal uterine bleeding and weighty loss from the last 5-6 months. She had no nausea, vomiting or bladder abnormalities. On physical examination tenderness in right hypogastrium was present. Exam- ination of the urogenital system was unremarkable. Blood tests revealed 7.3mg% haemo- globin. She was known hypertensive with blood pressure was recorded upto170/100 mmhg, no other relevant past medical history was present . An ultrasound scan was per- formed which revealed a well defined heterogeneously hypoechoic mass lesion in the right

suprarenal location measuring approximately 2.6X2.1X2.2 cm showing internal vascularity and few anachoic cystic areas .[Figure 1A,B].Based on the ultrasound appearance, our first differential diagnosis was of adrenal incidenteloma with paraganglioma bring the sec- ond possibility.CECT findings corroborated with USG and showed a small well defined oval shaped relatively hypodense lesion with HU value corresponding to 20-25 HU on plain scan in the retroperitoneum The mass mass was seen replacing the lateral limb of the the right adrenal gland while medial limb and body were normal . It showed avid en- hancement in the arterial phase (180HU) with washout in delayed phases (70 HU) .No in- ternal calcification or fast density was noted . The mass was abutting the ivc and right re- nal vein anteriorly .

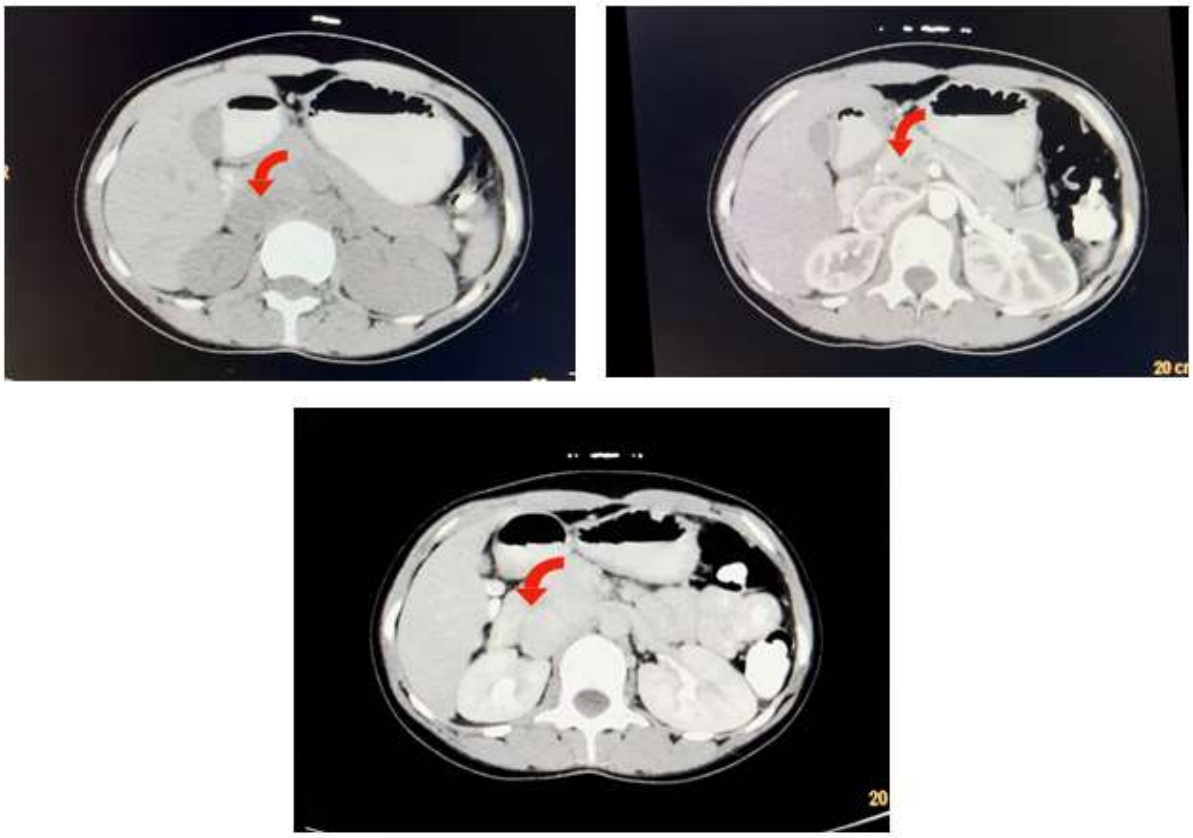

Fig 2A, 2B and 2C.Axial non contrast ,arterial phase ,delayed phase images: showing a well circumscribed, right adrenal mass

Few(5-6) small focal lesions showing hyper enhancement on arterial phase were noted in both lobes of liver measuring upto 5mm. They were not visualised on venous/delayed phase-possibility of metastasis was made.

The uterus and paramaterium showed increased enhancement .Extensive endocrinology workup was completed prior to surgical planning. Plasma normetanephrine and 24-hour urine normetanephrine levels were found to be 2170.9 pg/mL (0.0-136.8) and 7520 ug/24 hour (131-612) respectively. Plasma metanephrines resulted within range at 84.9 pg/mL (0.0-88.0) while 24-hour urine metanephrines were elevated at 864 ug/24 hour (36-209).The patient was started on oral prazosin 1mg to take twice daily for a duration of 10 days for preoperative alpha blockade prior to undergoing a right-side adrenalectomy.

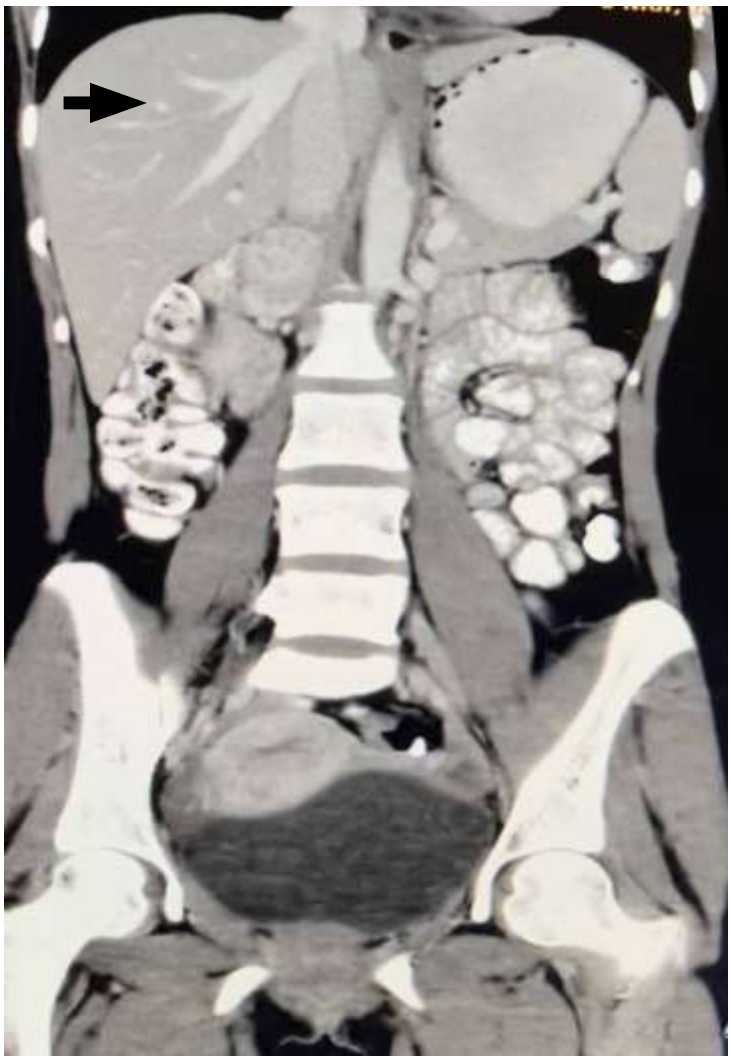

Fig 3A

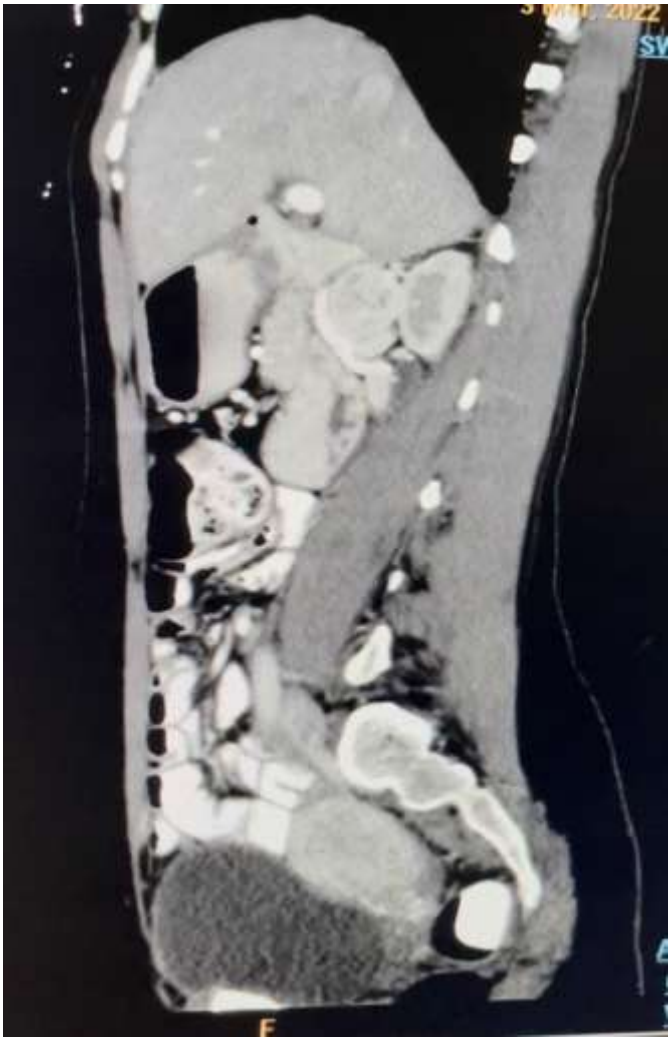

Fig 3 B

Coronal and saggital contrast arterial phase images : showing a well circumscribed, right adrenal mass and few focal hyper enhancing lesions in liver

DISCUSSION

Adrenal masses are mostly discovered incidentally. The differential diagnosis for unilateral adrenal masses are PCC, adrenal cortical carcinoma, adrenal myelolipoma, adrenal metastasis , adrenal adenoma, and more.

Biochemical testing and radiological studies help to differentiate between the possible differentials and help in guiding physicians to make the appropriate diagnosis.

In cases of PCC, Plasma levels and 24-hour urinary excretion of normetanephrines and metanephrines are typically elevated, as evidenced by our patient.

Initial imaging for evaluation of adrenal masses include CT and MRI studies with and without IV contrast. The sensitivity of CT scans for adrenal masses is upto 100%, however it is lacking the specificity of MRI and functional imaging for PCC [4].

The standard treatment of managing a PCC is surgical removal of the tumour. Surgical approach varies case by case, but include a transabdominal laparoscopic adrenalectomy or posteriorly retroperitoneoscopic adrenalectomy. According to Dickson et al., the posterior method has proved to be of shorter operation, decreased complications, decreased blood loss, and hospital stay when compared with the transabdominal approach [5].

In patients with PCC , before undergoing an adrenalectomy, we usually start them on a course of alpha-blockers for preventing intraoperative hypertensive crisis. Sole use of beta-blockers is contraindicated in patients with PCC because hypertensive crisis can be precipitated by an unopposed alpha agonism can. Our patient was discharged and was given a two-week course of prazosin for preoperative alpha blockade.

CONCLUSIONS

This case report discusses a diagnostic workup for an incidental adrenal mass and the requirement for a high clinical suspicion of PCC when a patient is presenting with hypertension and epigastric pain . Correlating the clinical presentation of our patient with the imaging findings from the Ultrasound and CT directed us to pursue further biochemical testing to confirm our suspected diagnosis at that time. Plasma and 24-hour urine normetanephrines and metanephrines were found to be elevated and the diagnosis was confirmed.

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