CASE REPORT

Diagnostic Obscurity in a Case of Large Right Suprarenal Mass

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Abstract

Aim: Aim of this case report is to highlight the difficulties we faced in differentiating renal mass from the adrenal mass.

Background: MIBG positive uptake usually signify a neuroendocrine tumor. However, there is reported false positive MIBG uptake in the chromophobe variant of renal cell carcinoma (RCC).

Case description: A postmenopausal lady in her early 70s presented to our outpatient clinic with right-sided flank pain radiating to the back. On examination, a right hypochondriac region lump of size 10 × 8 cm was found. Contrast-enhanced computed tomography (CECT) scan done elsewhere showed a large, well-defined, smooth, lobulated, heterogeneously enhancing soft tissue lesion involving the upper pole of the right kidney measuring 15.5 × 11.4 × 11.2 cm, suspected to be an adrenal mass. The hormonal evaluation was inconclusive and the iodine-123-meta-iodobenzylguanidine (MIBG) scan showed concentration in the right suprarenal region. In view of the inconclusiveness of the findings, a repeat dedicated adrenal protocol CECT scan was done, which showed a mass arising from the superior pole of the kidney. The patient underwent open radical nephrectomy and final histopathology showed a chromophobe variant of RCC, which had shown a false positive uptake in the MIBG scan.

Conclusion: We must interpret radiological, clinical and pathological aspects of large suprarenal mass during evaluation and be aware of false positive uptakes in MIBG scan.

Clinical significance: An MIBG scan is a good tool for assessing the functionality of the neuroendocrine lesion, but we should always keep in mind the other conditions that can take up MIBG.

Keywords: Chromophobe variant of renal cell carcinoma, MIBG scan, Pheochromocytoma.

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Background

Pheochromocytoma is a neuroendocrine tumor arising from the adrenal medulla and produces mainly catecholamines. The majority proportion of these cases are sporadic, but 10–25% can be associated with genetic syndromes, which include von Hippel-Lindau disease, type 1 neurofibromatosis, and multiple endocrine neoplasia type 2, which required genetic testing to confirm.

Iodine-123-meta-iodobenzylguanidine scan has been the mainstay in the functional assessment of adrenal lesions. With the advent of positron emission tomography with newer ligands, MIBG has taken a backfoot, especially in the evaluation of hereditary or metastatic/extra-adrenal pheochromocytomas. MIBG, however, is still very frequently done in India due to its easy availability and low cost.

Case Presentation

A postmenopausal lady in her early 70s presented in the Endocrine Surgery Outpatient clinic with complaints of right flank pain radiating to the back for 1 year. She complained of having a nontender mass in the right upper abdomen for 2 months. She also complained of having passing blood mixed with stools on two occasions. She was diabetic and hypertensive for 6 years. Her blood sugar levels were under control with oral hypoglycemic agents. Her blood pressure was also under control with tablets amlodipine, 5 mg, and atenolol, 50 mg, once a day. She had undergone a hysterectomy 15 years back for a fibroid uterus and cholecystectomy 4 years back for cholelithiasis.

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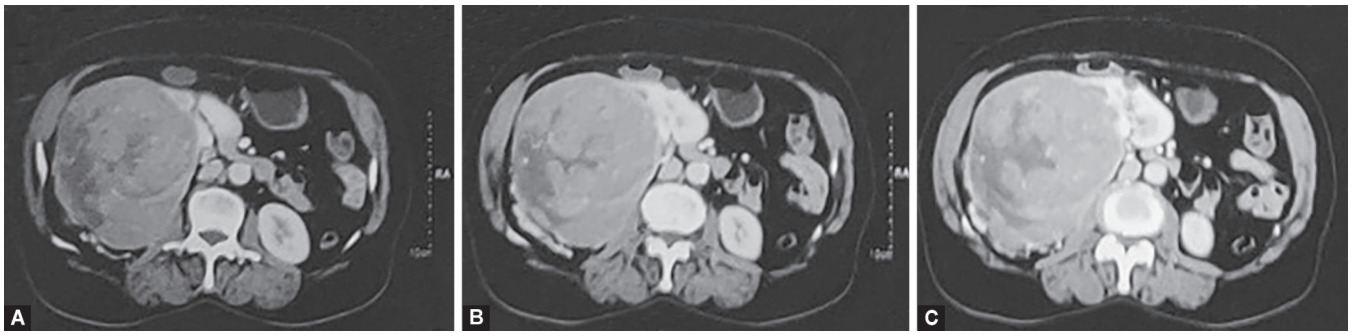
On examination, she was found to have a lump in the right hypochondriac region measuring around 10 × 8 cm extending around 10 cm below the costal margin. The mass was firm in consistency, did not move with respiration, and was ballotable. Fingers could be insinuated between the mass and the costal margins.

Investigations

She was evaluated elsewhere and underwent a CECT of the abdomen, which showed a large, well-defined, smooth, lobulated, heterogeneously enhancing soft tissue lesion involving the upper pole of the right kidney measuring 15.5 × 11.4 × 11.2 cm, suspected to be RCC (Fig. 1).



Figs 1A to C: CT scan images of the right suprarenal marge mass

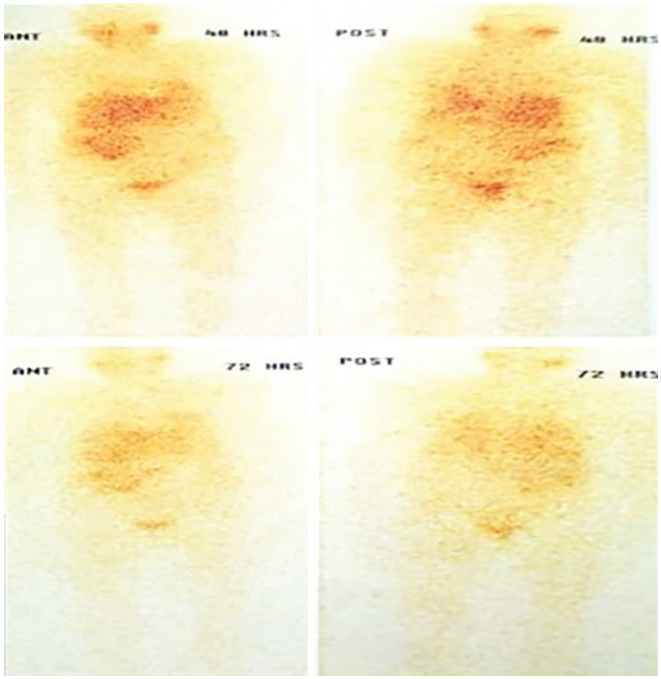


Fig. 2: MIBG scan showing uptake in right suprarenal mass

Ultrasound-guided fine-needle aspiration of the mass was attempted, but as it was inadequate for diagnosis, the patient was referred to the Department of Urology in our institute.

As the patient did not have any urinary complaints, the imaging was reviewed and there was suspicion of the lesion arising from the right adrenal gland. Hence, the patient was referred to the Department of Endocrine Surgery for hormonal evaluation.

Twenty-four-hour urine collection was carried out for measurement of metanephrines and normetanephrine and was found to be marginally elevated [urinary metanephrine: 355.85 µg (reference: <350 µg), urinary normetanephrine: 479.55 µg (reference: <600 µg)].

Serum cortisol level was normal (11.3 µg/dL, reference: 5–23) and was suppressed on overnight dexamethasone suppression (1.8 µg/dL, reference: 5–23).

An MIBG scan was performed and there was MIBG concentration in the right suprarenal region suggestive of a pheochromocytoma (Fig. 2).

Differential Diagnosis

With a history of right flank pain, without urinary complaints; hypertension which was well controlled, CECT differentials of RCC or adrenal mass; nonfunctional on hormonal assessment but with positive uptake on MIBG, the diagnostic dilemma persisted. After discussion with the patient, a dedicated adrenal protocol CECT scan was done and again discussed with radiologists. On the basis of imaging, an RCC diagnosis was made and the patient was transferred to the Department of Urology.

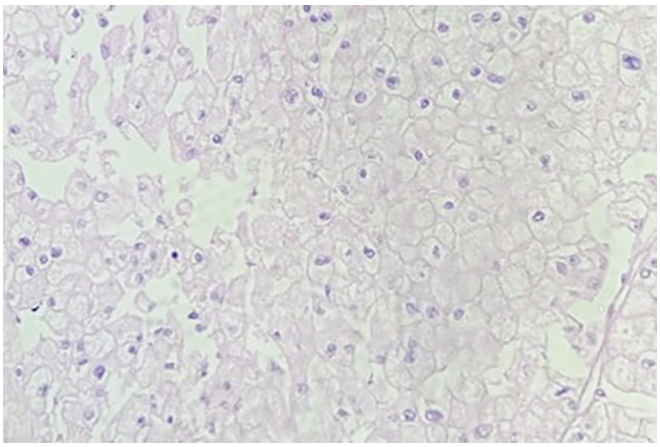


Fig. 3: Histology image of right renal cell carcinoma-chromophobe variant

Treatment

She was taken up for open radical nephrectomy. Intraoperatively, a 15 × 15 cm mass was identified arising from the upper pole of the right kidney abutting the renal vessels and inferior vena cava (IVC). On histopathologic examination, the mass was seen to have partially encapsulated tissue disposed of in sheets and nests separated by delicate fibrovascular septae (Fig. 3). Individual atypical cells were oval to polygonal in shape having plant cell-like membrane, small hyperchromic nuclei, and abundant eosinophilic vacuolated cytoplasm, and a diagnosis of chromophobe variant of RCC was made.

Outcome and Follow-up

The postoperative period was uneventful. The abdominal drain was removed on postoperative day 3, and the patient was discharged on day 5 of surgery. The patient is under follow-up in the Department of Urology and is doing well.

Discussion

Pheochromocytoma/Paragangliomas (PPGLs) account for approximately 7% (1.5–14%) of all adrenal incidentalomas. They manifest with symptoms of excess catecholamine levels such as

hypertension (sustained/paroxysmal), headache, palpitations, episodic sweating, pallor, and apprehension or anxiety.[1]

Elevated plasma or urinary metanephrine level with clinical symptoms that strongly suggest the presence of these tumors is recommended in diagnosing this condition. Plasma or urinary metanephrine levels, four times the upper reference limit, generally preclude the need for further tests, with the exception of cases where the patient is taking antidepressants or other drugs interfering with these biochemical assays, which require clonidine suppression test.[2,3]

Once the diagnosis is established with biochemical tests, localization is done by anatomical imaging. Functional imaging is indicated only in suspected PPGLs, familial/hereditary syndrome, metastatic disease at initial presentation, restaging after surgery, and if planning for targeted molecular therapy. MIBG scan is the most commonly done and widely available imaging technique for the functional assessment of PPGLs and has been the gold standard functional imaging method, until recently.[4] MIBG is a structural and functional analog of norepinephrine and guanethidine. Cytoplasmic intravesicular neurosecretory granules retain MIBG, permitting scintigraphic detection in the adrenal medulla, pheochromocytomas, extra-adrenal PPGLs, and neuroblastomas.[5] The sensitivity of 123I-MIBG ranges between 85 and 88% for pheochromocytomas and between 56 and 75% for PPGLs, whereas specificity ranges from 70 to 100% and 84 to 100%, respectively.[6–9]

False-positive findings are caused by tumors that express the neuroendocrine transporter system on the cell membrane; these include chromophobe variants of RCC, carcinoids, medullary thyroid cancer, Merkel cell carcinoma, ganglioneuroma, and composite tumors. They have also been reported in angiomyolipoma, hepatocellular carcinoma, accessory spleen, and adrenocortical carcinoma.[10–13] In this case, the patient had complaints of abdominal pain, with a history of hypertension, without urinary complaints, and an inconclusive biochemical evaluation and imaging characteristics with MIBG concentrating in the right suprarenal region; we were misled into suspecting a pheochromocytoma. However, a repeat radiological imaging with a dedicated adrenal protocol, which involves a CT washout technique using unenhanced CT (to establish benign absolute washout of >60%) or using only 70-second and 15-minute CECT (to calculate relative washout of >40%), helped us reach an appropriate decision.[14]

Learning Points/Take Home Message

An MIBG scan is a good tool for assessing the functionality of the lesion, but we should always keep in mind the other conditions that can take up MIBG.

Contrast-enhanced CT scan should be done with a dedicated adrenal protocol to reduce the delay in reaching the diagnosis especially if the patient is suspected to have a renal or suprarenal mass.

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