Case Report

Primary extra-renal clear cell renal cell carcinoma masquerading as an adrenal mass: A diagnostic challenge

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Abstract

We present the first case of a nonmetastasizing renal cell carcinoma (RCC) masquerading as an adrenal mass, in the presence of normal bilateral native kidneys, in a young adult. The possibility of this mass developing in a supernumerary kidney was ruled out, since no identifiable renal tissue, pelvis or ureters was seen within the mass, nor was any separate systemic arterial supply to the mass seen. The diagnosis of extra-renal clear cell RCC was based on cyto-morphological features, further confirmed by immunohistochemistry findings. The origin of this extra-renal clear cell renal cell is proposed to be from the mesodermal embryonic rests.

Key Words: Adrenal, clear cell, extra-renal, metastasis, renal cell carcinoma

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INTRODUCTION

Renal cell carcinoma (RCC) is the most common renal tumor in adults with the clear cell being the most common histologic type.[1] Extra-renal localization of RCC, with intact bilateral kidneys, is an exceptionally rare occurrence and has been reported only once before in English literature.[2] Herein, we describe an interesting and rare case of extra-renal RCC, camouflaging as an adrenal mass.

CASE REPORT

A 28-year-old female presented in the outpatient department of a tertiary care hospital with left upper quadrant pain of 1 month duration. The pain was constant, dull aching in nature and did not migrate. It was not associated with vomiting, fever, hematuria, dysuria or bowel disturbance. On physical examination, her abdomen was soft and no palpable lump was found. Rest of the systemic examinations were within normal limits. Computed tomography (CT) scan of the abdomen revealed a 6 cm × 5 cm × 5 cm, heterogeneously enhancing, necrotic, left suprarenal mass inseparable from the lateral limb of the left adrenal gland. No calcification or fat density was seen within the lesion. Left kidney was inferiorly displaced by the mass lesion, however intervening fat planes were maintained. Medially the lesion was abutting the crus of left hemi-diaphragm and laterally the spleen respectively, with maintained fat planes [Figure 1]. CT Hounsfield units (HU) within the solid posterior portion of the lesion were as follows: Plain scan - 47 HU, postcontrast enhanced scan - 104 HU, delayed scan - 73 HU, absolute washout - 54.3%, relative washout - 29.8%. Both kidneys showed normal size, location and enhancement. No mass lesion was seen in either kidney. Plasma and urine catecholamine levels, 24 h urinary vanillylmandellic acid (VMA) and serum cortisol levels were within normal limits. Routine hematological investigations were normal except for mild leukocytosis (white blood cell count – 11.8 × $10^3/\mu l$). Based on clinical examination

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and radiological assessment a diagnosis of left adrenal mass was made and the differentials entertained were an adrenal carcinoma, metastasis, and pheochromocytoma. Subsequently, laparoscopic left adrenalectomy by way of transperitoneal approach was done. Intraoperatively a 5.5 cm × 5.5 cm mass arising from left adrenal gland was found, and a single adrenal vein was seen draining into the renal vein. The mass was easily mobilized all around from the spleen, kidney, and stomach. The adrenal vein was ligated and divided and wound closed in layers.

Gross examination of the surgical specimen sent as adrenal gland weighed 119 g and measured 8 cm × 7.5 cm × 5.5 cm. Externally it appeared as a nodular lesion, which was enlarged and congested. Cut section shows cystic areas, haemorrhagic areas, gray brown necrotic areas and gray yellow areas filled with hemorrhagic fluid. Peripheral portion showed separate preserved adrenal gland, totally separate from the tumor [Figure 2].

Histologically, the partially encapsulated tumor comprised of alveolar nests of polygonal cells with distinct cell borders, abundant clear to pale eosinophilic cytoplasm, oval to angulated irregular nuclei some with prominent nucleoli, increased mitotic activity, few dispersed large bizarre cells separated by fibrous septa with blood vessels and luminal fibrin along with areas of hemorrhage, fibrin deposition, areas of necrosis and hemosiderin laden macrophages [Figure 3]. On immunohistochemistry (IHC), the tumor cells showed diffuse and strong membranous positivity for CD10, weak positivity for CK and were negative for chromogranin, epithelial membrane antigen (EMA) and inhibin [Figure 4]. The pathological diagnosis was extra-renal clear cell RCC.

The postoperative course of the patient was uneventful and no recurrence was noted after 6 months of follow-up.

DISCUSSION

Renal cell carcinomas have been sporadically described in the literature to occur in ectopic,[3] and supernumerary kidneys,[4] however only a single published report describes this entity in the presence of normal bilateral kidneys.[2] Extra-renal RCC refers to the occurrence of RCC in locations other than the normal native kidneys, likely arising from mesonephric remnants.[2] We report this first case of RCC masquerading as an adrenal mass, in the presence of normal bilateral kidneys.

Radiologically, the large mass was found to be inseparable from left adrenal gland and showed heterogeneous

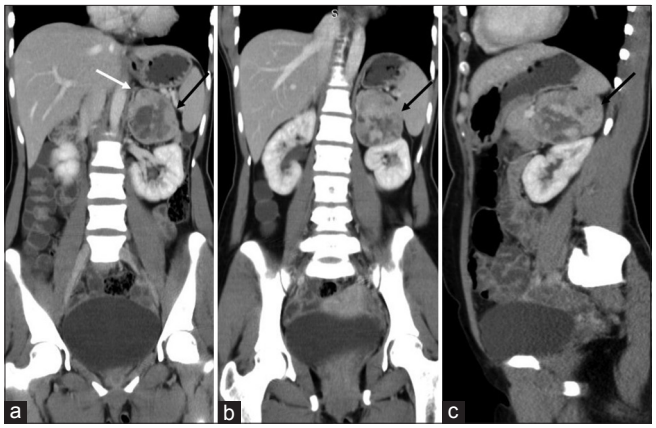

Figure 1: Contrast-enhanced computed tomography abdomen: Coronal (a and b) and sagittal (c) images reveal a large heterogeneously enhancing, left suprarenal mass showing central necrosis (black arrow), closely abutting the adrenal gland (white arrow), with maintained intervening fat planes with left kidney

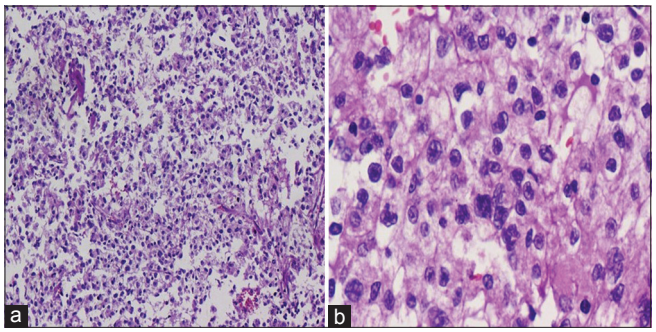

Figure 3: Microscopy demonstrates cytomorphologic features consistent with conventional clear cell renal cell carcinoma (a) H and E, ×100 (b) H and E, ×400

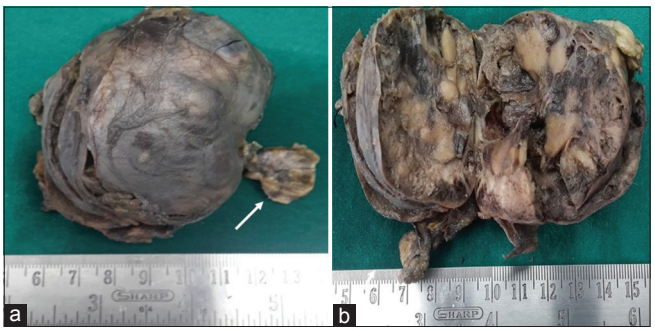

Figure 2: (a) A large nodular mass lesion with peripherally compressed preserved normal adrenal gland (arrow), (b) Cut section shows grey brown, cystic, hemorrhagic and necrotic areas

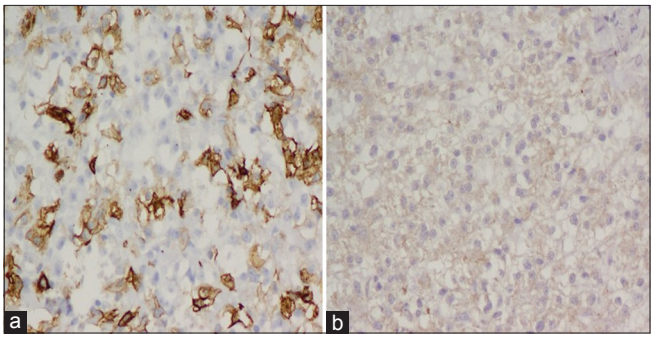

Figure 4: The tumor showed immunoreactivity for (a) CD10, ×200 and (b) CK, ×200

enhancement with slow contrast washout, imaging features which are associated with malignant adrenal lesions like adreno-cortical carcinoma and metastasis.[5] The radiological differentials were an adrenocortical carcinoma, metastasis, and pheochromocytoma. Pheochromocytoma was ruled out in the absence of hypertension with normal levels of 24 h urinary VMA, catecholamines, and metanephrines. Since there was no other detectable primary seen, metastasis was considered less likely. As the urinary cortisol levels were normal, primary differential suggested was nonfunctioning adreno-cortical carcinoma.

Per-operatively the mass was found attached to the adrenal gland, distinctly separate from the left kidney. No separate ureter or arterial supply was found for the mass, thus effectively ruling out the possibility of the supernumerary kidney.

Pathologically, the differentials in our case included pheochromocytoma, adrenal cortical carcinoma and RCC as renal metastasis was ruled out in lieu of radiologic and per-operative demonstration of uninvolved kidneys. In most of the cases, morphology and IHC lend a diagnosis even with equivocal clinico-radiological findings.

On IHC, adrenal cortical carcinomas are immunoreactive for inhibin, A103 and Melan A whereas RCCs are negative for these markers. Pheochromocytomas show chromogranin, synaptophysin and neuron specific enolase positivity, whereas CD10, EMA, and cytokeratin AE1/AE3 are positive in the majority of RCCs.[6]

In our case, the mass was seen arising in the vicinity of the adrenal gland and compressing it. However the whole adrenal gland was easily resected away from the tumor and was found to be disease free. On IHC, inhibin and chromogranin negativity effectively ruled out adrenal cortical carcinoma and pheochromocytoma respectively, whereas CD10 and CK positivity confirmed the diagnosis of extra-renal RCC.

To conclude, we are documenting this very rare case of clear cell RCC masquerading as an adrenal mass, with normal bilateral native kidneys. Since there was no evidence of any communication with the kidneys, or any demonstrable renal tissue seen within the mass, the origin of this tumor can only be attributed to delayed neoplastic transformation in mesonephric embryonal rests. Thus it is imperative upon the clinicians, radiologists and pathologists to be aware of this rare entity, extra-renal RCC, and include it in the differentials for an adrenal mass.

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