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Ectopic hepatocellular carcinoma presenting as a right adrenal mass with IVC thrombus: Case report and review of the literature

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ABSTRACT

Hepatocellular carcinoma (HCC) arises from several different etiologies and carries a poor prognosis. Extrahepatic metastases of HCC are most frequently found in the lungs, lymph nodes, and bones, with adrenal metastases reported in less than 15% of metastatic cases. Herein, we report a case of a 71-year-old man without prior liver disease who presented with a 9-cm right adrenal mass extending into the IVC consistent with HCC on subsequent surgical pathology and immunohistochemical staining. Etiological possibilities for an adrenal tumor as the first presentation of HCC include metastasis from occult primary liver HCC, intra-adrenal hepatic heterotopia, and adrenohepatic fusion.

1. Introduction

Hepatocellular carcinoma (HCC) is the most common primary tumor of the liver. When metastases are present, less than 15% affect the adrenal glands. Herein, we report a case of a 71 year-old man without prior liver disease who presented with a large right adrenal mass extending into the IVC later discovered to be HCC.

2. Case presentation

A 71 year-old man presented to the ED with 2 years of periumbilical and right lower quadrant abdominal pain that acutely worsened. He described no systemic complaints, denied a history of palpitations, headaches, flushing or chest pressure, and physical exam was unremarkable. His medical history was non-contributory. Cross-sectional CT abdomen/pelvis with contrast revealed a 9.1 x 8.2 × 8.6 cm right adrenal mass with a level III thrombus of the inferior vena cava (IVC). A 2 × 2.2 cm hypodense lesion in the liver appeared unchanged compared to previous images. Serum potassium, aldosterone, cortisol, and plasma metanephrines were normal. MRI abdomen revealed a right-sided adrenal mass measuring 7.8 x 8.6 × 10 cm without evidence of fat invasion but showed IVC invasion with near-complete filling of the lumen (Fig. 1).

The patient subsequently underwent open right adrenalectomy. The adrenal tumor was well-encapsulated and separate from the normal-appearing liver but adherent to the renal hilum and lateral aspect of the IVC, requiring radical nephrectomy and IVC reconstruction. His hospital course was uncomplicated, and he was discharged in good condition on post-operative day 8.

Gross examination of the specimen showed a large hemorrhagic mass and tumor invasion through a pseudocapsule (Fig. 2). Pathologic assessment of the mass and IVC thrombus was consistent with HCC (Fig. 3). There was evidence of lympho-vascular invasion as tumor was present at renal artery resection margins. No adrenal tissue was noted.

Post-operatively, the patient was referred to oncology for HCC workup. Laboratory cancer markers were within normal limits (LDH 141 U/L, CEA 1.1 ng/mL, AFP 1.87 ng/mL) and hepatitis workup was negative. PET imaging revealed no evidence of metastasis. It was decided no additional therapy was warranted until there was macroscopic evidence of recurrence. On follow-up MRI at 10 months post-op, he was found to have enhancing soft tissue within the surgical bed concerning for recurrent tumor versus metastatic lymphadenopathy. There was also metastatic portocaval lymphadenopathy and tumor thrombus in the IVC and left renal vein. The inferior right hepatic lobe

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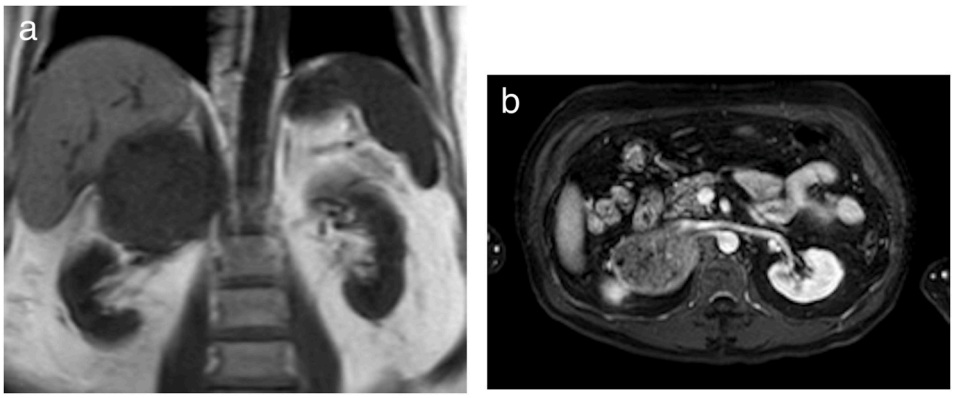

Fig. 1. Pre-operative MRI revealing (a) right adrenal mass (b) IVC thrombus.

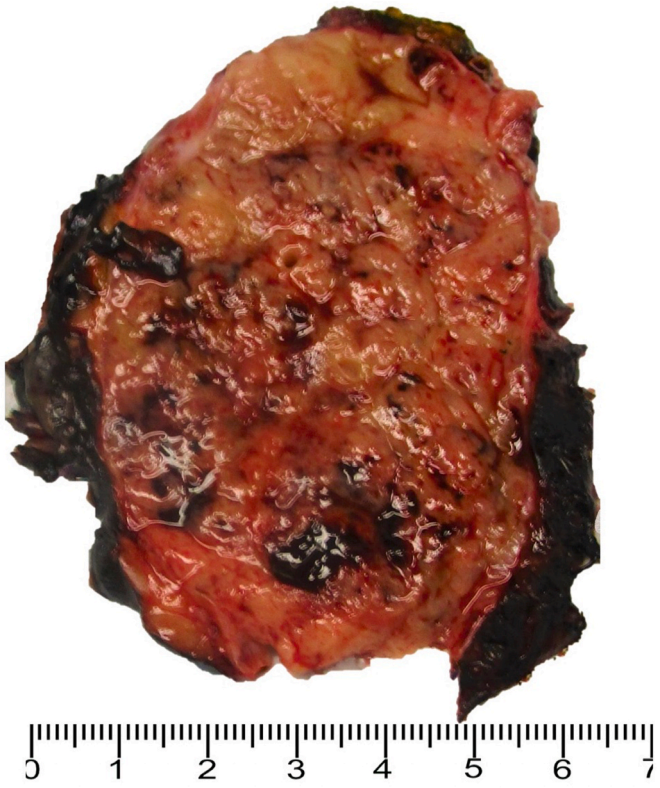

Fig. 2. Section of adrenal mass showing tan yellow to tan pink nodule with areas of hemorrhage. (color photo). (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)

lesion was unchanged from prior imaging. Bone scan was negative for osseous metastasis. AFP, CEA, and CA 19–9 remained non-elevated. He was then started on lenvatinib.

3. Discussion

The differential for an adrenal mass includes adrenocortical carcinoma, adenoma, pheochromocytoma, and metastasis. In patients with known extra-adrenal cancer, the literature describes 30–75% of incidental adrenal masses arise secondary to metastasis, most commonly from the lung and liver.

Herein we report a rare case of a 71 year-old man without liver disease who presented with a 9-cm right adrenal mass extending into the IVC with an adjacent 2.2-cm hypodense liver lesion in the right lower lobe that was non-PET avid and without imaging characteristics suggestive of HCC. Pathological analysis of the adrenal mass showed HCC without adrenal tissue. From our review, cases of adrenal masses as the first presentation of HCC are infrequent. Longmaid et al. reported 7 patients who presented similarly with a primary extrahepatic mass that was found to be HCC. Of these, 7 patients presented with an adrenal mass. Five had liver lesions initially thought to be metastatic while 2 had no notable liver lesions.[1] To our knowledge, there have been only 3 cases of patients presenting with a significant adrenal mass found to be HCC, in the setting of small or occult liver lesions.[2–4] Our patient had the most extensive disease, with extension of the tumor thrombus into the IVC.

While the adrenal mass is most likely a bulky metastasis associated with an occult primary liver tumor, the small liver lesion in our patient had not grown over one year of imaging and was non-PET avid. AFP was not obtained prior to adrenal resection, but post-operative AFP was non-elevated. Other etiologies include intra-adrenal hepatic heterotopia, or adreno-hepatic fusion. Heterotopia is the presence of a tissue type in a non-physiological site.[5] There have been several reports of hepatic nodules developing inside bone, omentum, and the adrenal gland, with a proposed etiology of this phenomenon being an aberrant segment of hepatic diverticulum that wanders into adjacent developing adrenal tissue and is severed upon formation of adrenal and liver capsules. It is possible that our patient had heterotopic liver tissue in the adrenal gland that developed into HCC. Less likely is adrenohepatic fusion, a rare age-related phenomenon where the parenchyma of the right adrenal gland and the liver fuse in the absence of any fibrous capsule dividing the two organs. However, in our case, intraoperatively the liver was found to be separate from the adrenal mass.

Metastatic HCC carries a poor prognosis, with 5-year relative survival rates less than 20% in the US. This also varies based on risk factors, stage at diagnosis, treatment, and etiology of liver disease. Current first-line treatment for metastatic HCC is molecularly targeted therapy with sorafenib or lenvatinib. Systemic chemotherapy has been shown to be less effective for HCC in clinical trials given their side-effect profile and hepatotoxicity. Considering the heterogeneous population of HCC patients, guidance on how to tailor treatment based on the underlying liver etiology continues to be a field of interest. Given our patient's disease progression on 10-month MRI despite lack of suspicious lesions on immediate post-operative PET scan, early systemic therapy in the absence of radiologically visible lesions may be beneficial in delaying progression of metastatic HCC.

4. Conclusion

In this report, we presented a case of HCC of the right adrenal gland with IVC tumor thrombosis in a patient with no known primary tumor. Markers for adrenal medullary and cortical carcinomas were negative.

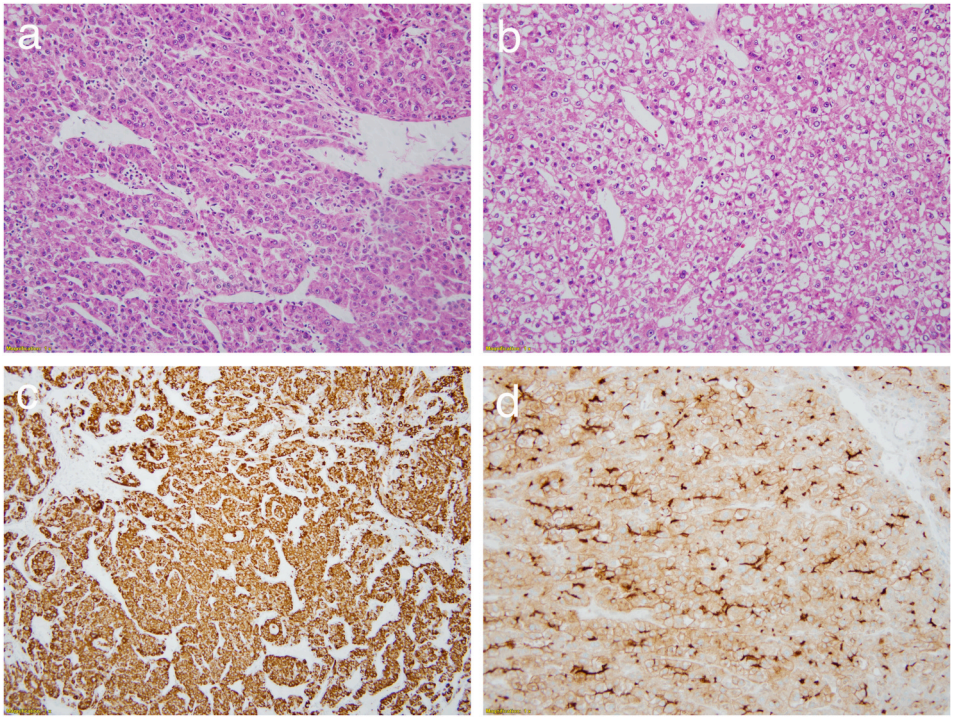

Fig. 3. Histology of tumor. (a) H&E staining demonstrating tumor has a trabecular growth pattern with eosinophilic cytoplasm and prominent nucleoli. (b) Some tumor cells show clear cytoplasm. (c) Cells stained with HepPar-1 show diffuse cytoplasmic positivity in tumor cells. (d) Immunological staining for CD10 demonstrate canalicular pattern positivity. Slides were negative for inhibin, Melan A, chromogranin, synaptophysin, and GATA-3. (color photo). (For interpretation of the references to color in this figure legend, the reader is referred to the Web version of this article.)

The results of the pathology and HCC-specific markers indicated HCC tumor of the adrenal glands from hepatic heterotopic tissue. Further workup of risk factors for HCC should be evaluated.

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