



Oncology

Macronodular adrenal hyperplasia masquerading as an upper pole renal mass

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ABSTRACT

Macronodular hyperplasia (MAH) of the adrenal gland is a rare disease usually presenting with Cushing Syndrome. Although usually readily apparent on imaging, an adrenal tumor in an asymptomatic patient may be mistaken for a renal tumor. We present a patient with combined macro- and micro-nodular adrenal hyperplasia masquerading as an upper pole renal mass. The patient underwent a robotic partial nephrectomy and partial adrenalectomy without complication.

Introduction

With the increasing use of abdominal imaging, there is an increase in incidental findings of renal and adrenal masses⁽¹⁾. Macronodular adrenal hyperplasia is a rare cause of Cushing Syndrome, typically found to be bilateral in origin. We present a case of an inactive, unilateral nodule masquerading as an upper pole renal mass in an asymptomatic patient.

Case presentation

The patient is a 56-year-old African American woman with no significant prior medical history. The patient underwent imaging for an unrelated cause at an outside hospital and was found to have an incidental mass at the upper pole of kidney. There, she was counseled on the need for an open partial nephrectomy. She sought a second opinion at our institution. The patient denied hematuria, dysuria, changes in weight, and lacked stigmata of corticosteroid excess. She had no family history of renal cell carcinoma or chronic kidney disease, with a pre-operative eGFR of 91, and pre-operative creatinine of 0.83 mg/dL. MRI at our institution demonstrated a single enhancing lesion measuring 2.9 × 1.6 cm, interpreted as an upper pole renal mass, likely to be renal cell carcinoma (Fig. 1:a-b). The patient was scheduled for robotic partial nephrectomy.

The procedure began with standard technique for a robotic partial nephrectomy. Upon retracting the liver, the mass was seen, but it had a

golden hue more consistent with adrenal tissue. Gerota's fascia was opened near the mass, revealing that the tumor was contiguous with the right adrenal gland. Intraoperative ultrasound was used to evaluate the depth of invasion into the renal parenchyma, and it was deemed that the tumor only superficially involved the kidney, if at all. No other masses were seen on intraoperative ultrasound. Interestingly, a small, yellow lesion was seen overlying the renal artery that has the typical canary yellow hue of adrenal tissue but was not contiguous with the adrenal gland (Fig. 1: c-d). This was resected and sent for pathologic examination.

Next, an incision was made through the lateral limb of the adrenal gland just at the edge of the tumor and the mass was dissected leaving a rim of normal adrenal tissue for a margin. The renal capsule was scored circumferentially, and a partial nephrectomy was performed removing a rim normal renal parenchyma for margin. Clamping of the kidney was not necessary. The tissues were placed in an Endo Catch™ bag and extracted. A 2-layer renorrhaphy was performed and hemostatic agents were placed in the adrenal bed. Estimated blood loss was 350 mL. In total, greater than 95% of the normal kidney and 90% of the adrenal gland were spared.

Upon pathological review, the specimen was determined to exhibit macro- and micronodular hyperplasia of the adrenal gland, with no evidence of renal neoplasm. Interestingly, while there was a capsule around the adrenal nodule circumferentially, there was no tumor pseudocapsule or normal renal capsule between the adrenal tumor and

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the renal parenchyma (Fig. 2). The perihilar tissue was consistent with ectopic adrenal tissue. The patient was discharged on post-operative day three without complication or need for steroid replacement therapy. The patient's renal function at discharge was unchanged from her pre-operative values with eGFR 91 and creatinine 0.83 mg/dL. Somatic mutation testing revealed no variants of clinical or pathologic significance.

Discussion

We report a case of macro-and-micronodular hyperplasia of the adrenal gland presenting as a renal mass. Interestingly, this patient did not present with any endocrine abnormalities that would be expected in a patient with Cushing syndrome.

Macronodular hyperplasia is an uncommon condition often resulting in adrenocorticotrophic hormone-independent Cushing syndrome (CS). On rare occasions, it presents as an isolated entity. On cross-sectional imaging, macronodular hyperplasia may be mistaken for adrenal cortical adenomas and other adrenal masses.

Micronodular hyperplasia can be seen along with primary pigmented nodular adrenocortical disease which is associated with the Carney complex, an autosomal dominant hereditary condition associated with cardiac and cutaneous myxomas, cutaneous hyperpigmentation and endocrine abnormalities associated with germline mutation in *PRKARIA*.² Adrenal nodular hyperplasia can also be seen in patients affected with Hereditary Leiomyoma and Renal Cell Carcinoma (HLRCC) characterized by germline mutation of the *fumarate hydratase (FH)* gene.³

Several learning points in this case require emphasis. First, even among experienced surgeons and radiologists, adrenal masses and upper pole renal masses may be misinterpreted. While certain adrenal masses, such as pheochromocytoma or lipid-rich adenoma have characteristic imaging features, the mass in this case did not. Given the tumor's morphology, lack of capsule and the presence of ectopic adrenal tissue by the renal hilum, this lesion could also represent nodular hyperplasia of ectopic adrenal tissue, though it appeared contiguous with the rest of the adrenal gland intraoperatively. In cases of diagnostic uncertainty,

biopsy may aid in diagnosis. There are a variety of other retroperitoneal masses that may masquerade as a renal neoplasm. While rare, primary retroperitoneal masses are likely to be malignant (70–80%).⁴ Solid malignant retroperitoneal tumors such as a well-differentiated sarcoma are typically large with features of necrosis or hemorrhage (Table 1). When diagnosing a retroperitoneal mass, there may be significant overlap in imaging findings. It is important to determine its origin (or lack thereof) from another organ and adjacent structures. Schwannomas of the retroperitoneum are typically paravertebral. Features such as cystic components, calcifications and vascular invasion may also direct the diagnosis.

Secondly, when faced with the finding of peripheral adrenal mass, total adrenalectomy is usually not necessary. We have previously reported outcomes of partial adrenalectomy for pheochromocytoma, with low rates of recurrence and need for steroid replacement.⁵ In the case of a macro-and micronodular hyperplasia, a process which can be bilateral, sparing normal adrenal tissue may be advantageous should future biochemically active masses develop and require intervention.

Lastly, pre-surgical work up of an adrenal mass typically includes biochemical work up. While this patient was asymptomatic, we cannot exclude the possibility of sub-clinical Cushing syndrome.

Conclusion

In this report we describe a case of a macro-and-micronodular adrenal hyperplasia masquerading as a solitary renal mass in an asymptomatic patient.

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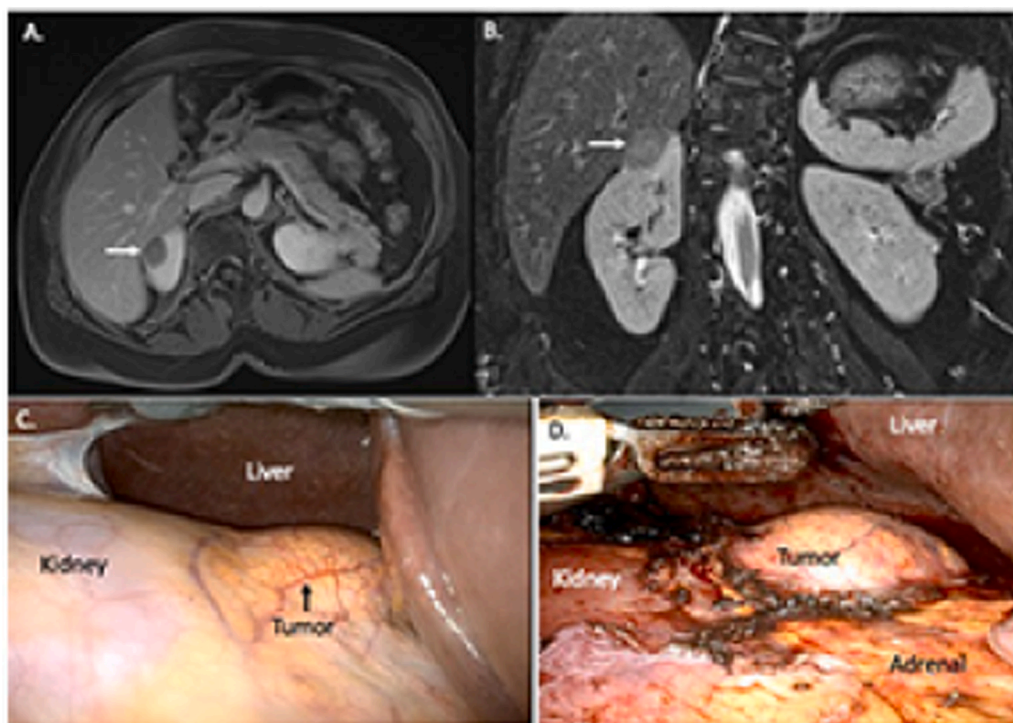


Fig. 1. Radiographic and Intraoperative views of tumor. **A.** Axial T1 contrast-enhanced and **B.** coronal T2 MRI demonstrating lesion at upper of the right kidney indenting the renal parenchyma **C.** Intraoperative image of yellow hued tumor at the upper pole of the right kidney covered by Gerota's fascia and **D.** after incising Gerota's fascia demonstrating continuity right adrenal gland. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

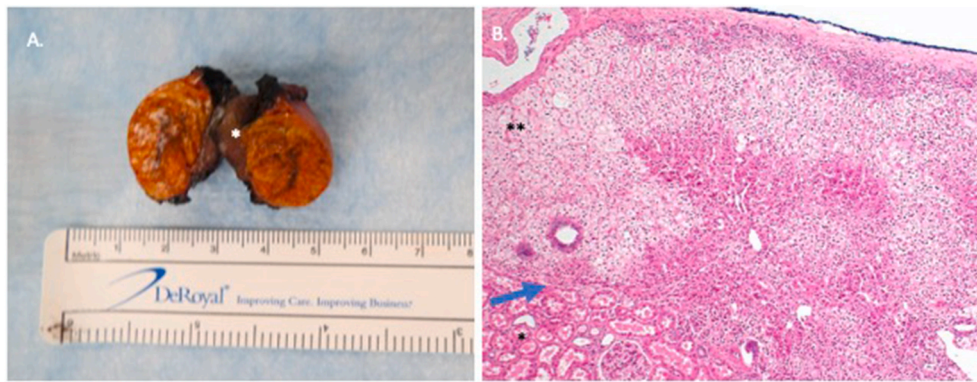


Fig. 2. A) Gross specimen of adrenal nodule with small rim of renal parenchyma denoted by * and B) microscopy demonstrated adrenal hyperplasia denoted by ** with normal renal parenchyma denoted by *. Note the absence of renal capsule between the two areas denoted by blue arrow. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

Table 1
Differential diagnosis of retroperitoneal masses in adults.

Type of Tumor	Characteristic Radiological Features (CT/MRI)	Other Key Points
Solid – Malignant		
Liposarcoma	Typically large tumor with thick nodular septa and fat	Most common primary retroperitoneal sarcoma
Leiomyosarcoma	Necrosis and hemorrhage due to vascular involvement and growth patterns	Typically seen in women ages 50–60s
Lymphoma	Para-aortic or pelvis homogenous mass with lymph node enlargement & + PET/CT	Paraaortic lymph node involvement in both Hodgkin’s and Non-Hodgkin’s variants
Solid – Benign		
Schwannoma	Spherical or ovoid heterogenous mass	Typically seen in women ages 20–60s
Retroperitoneal Fibrosis	Fibrotic mass abutting or infiltrating adjacent structures	Typically seen in men ages 40–60s
Erdheim-Chester Disease	Bilateral masses, periaortic and perirenal	Associated bone lesions common
Cystic		
Teratoma	Fat, calcifications, teeth, hair with cystic features	Investigate testes in male for secondary lesion
Cystic Lymphangioma	Thin-walled, fluid filled, cystic mass	Fluid may be serous, chylous or hemorrhagic

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