CASE REPORT

Malignant cystic nephroma

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Aim: *To describe a malignant cystic nephroma in an asymptomatic man.*
Methods: *Case report and review of the literature.*
Results: *A 60 year old white male presented with an incidentally discovered right perirenal mass. An MRI demonstrated a large perinephric encapsulated mass with diffuse heterogeneity. Patient underwent a radical nephrectomy and retroperitoneal node dissection. Histopathological analyses of the resected specimen revealed malignant cystic nephroma.*
Conclusion: *This represents the first published report of this rare tumor in an adult patient.*

Key Words: malignant cystic nephroma, renal mass

Introduction

Cystic nephromas in adults typically are considered a benign entity consisting of stroma and epithelium of uncertain histological origin. In children, similarly identified cystic nephromas are often highly cystic Wilms' tumors of nephroblastic origin with little or no malignant invasive potential.[1] In general, cystic tumors are more likely benign if surrounded by a fibrous capsule, the cysts are fluid-filled bereft of solid nodules, and cystic fluid is clear.[2] Malignant cystic nephroma in adults is a rare entity of uncertain etiology.

Case report

The patient is a 60-year-old Caucasian man with a past medical history significant for hypertension, type 2 diabetes and coronary artery disease, who presented for further evaluation of an incidentally discovered right peri-renal mass on an abdominal CT scan, Figure 1a. No significant abnormalities were noted on his physical exam or laboratory parameters. He underwent a core biopsy of his perinephric mass; however the pathology was inconclusive. MRI demonstrated a right perinephric encapsulated-appearing mass measuring 5.6 cm x 5.9 cm x 7.9 cm, with diffuse heterogeneity, and T2 hyperintensity with some mixed T1 hyperintensity, Figure 1b. The mass compresses but does not appear to invade the upper renal pole. Post contrast images demonstrate moderate peripheral heterogeneous enhancement. There appears to be no involvement of the right adrenal gland, renal vein and renal artery. An enlarged (1 cm) aortocaval lymph node at the level of the lower renal pole was noted. Whole body FDG tumor localization PET Scan was negative for metastasis or other malignancies.

Due to these imaging findings, the patient elected for operative management of his renal mass and underwent a right radical nephrectomy and retroperitoneal node dissection. Post-operatively he did well. Follow up CT at 3 years in this asymptomatic patient shows no evidence for local recurrence or metastatic disease.

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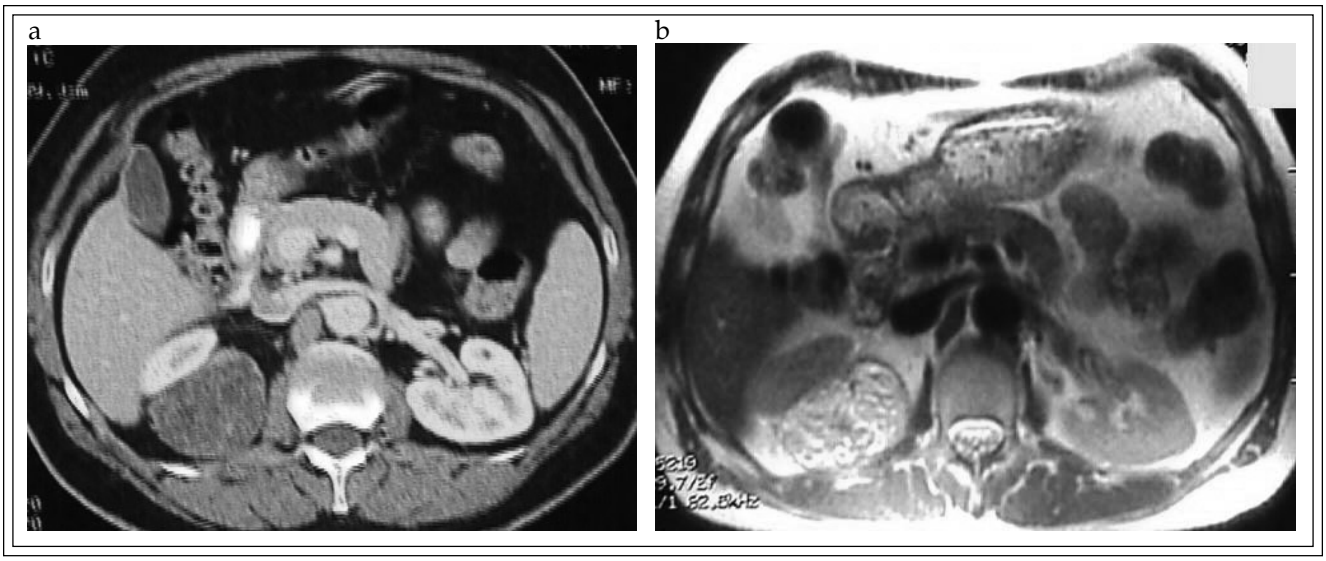

Figure 1a,b. CT demonstrating right perirenal mass (1a). MRI with gadolinium enhancement of the right kidney with mass in transverse T2-weighted section (1b).

Histopathological analyses revealed malignant cystic nephroma with adenomatous elements, Figure 2a. This case has many unique features: a tubulopapillary lesion within its stroma that is very similar to the common cortical adenoma, the solid quality of the gross specimen, the thick inter-cystic septa and the pleiomorphism and mitoses of the stromal cells, Figure 2b distinguish this tumor from the benign variants. Presumably, this is yet another example of the potentiality of the nephroblastic precursor cell as illustrated best in Wilms tumor. The pathological features were reviewed and independently confirmed by a senior pathologist at the Armed Forces Institute of Pathology, USA.

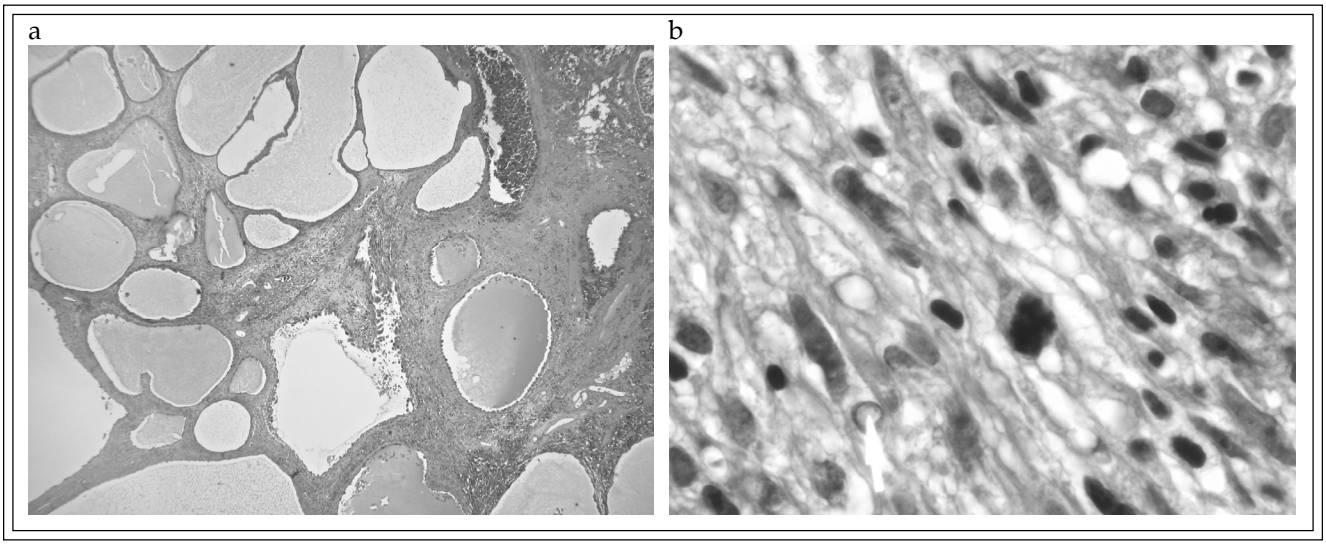

Figure 2a, b. Histopathological examination of resected peri-renal mass (2a): The majority of the tumor consisted of cystic spaces lined with cuboidal epithelium, with eosinophilic proteinaceous material, set within a variably cellular spindled stroma (H&E, 4x objective) (2b): In an area of tumor dominated by stroma, atypical spindle cells with enlarged, elongated, blunt-ended nuclei, nuclear envelope irregularities, prominent nucleoli and occasional nuclear folding are present in a loosely fascicular arrangement (H&E, 100x objective).

Discussion

Cystic renal neoplasms are a source of diagnostic confusion and controversy especially preoperatively because of their similarities in gross appearance, in pathological features, and on imaging studies. Characteristic MR findings are helpful but not definitive when trying to diagnose cystic nephromas. A complex cystic renal lesion with enhancing septa and herniation into the collecting system are common findings of benign cystic nephroma. Due to the difficulty of distinguishing malignant cystic nephroma from other cystic renal diseases from any imaging study alone, surgical intervention is the only effective method for proper differentiation.[3]

Upon review of the literature, our case of malignant cystic nephroma is the only current report of this highly unusual neoplasm. Histopathological assistance in the diagnosis cannot be overstated. Various histopathological features distinguish this tumor from the benign variants including the thickened septations between cysts, and the multiple pleiomorphic mitoses. Early diagnosis of such a rare tumor is difficult even with proper imaging studies, yet early surgical intervention by the aware urologist is vital. □

References

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