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Case study

Malignant mixed epithelial and stromal tumor of the kidney with rhabdoid features: report of a case including immunohistochemical, molecular genetic studies and comparison to morphologically similar renal tumors

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Summary Mixed epithelial stromal tumor of the kidney (MEST)/adult cystic nephroma (CN) is a lesion characterized by epithelial lined tubular or cystic structures interspersed within a variably prominent, distinctive spindle-cell stroma. Although typically benign, cases with malignant features have been reported. Herein, we report a MEST/CN with malignant stromal features and rhabdoid differentiation arising in the left kidney of an 84-year-old woman. Histologically, the tumor displayed multiple tubules and variably sized cystic structures lined by benign epithelium with an intervening malignant-appearing spindle-cell stroma. The malignant stroma displayed condensation in the regions surrounding the epithelial component consistent with the ovarian-like stroma typically observed in MEST/CN. In addition, the stromal cells displayed extensive rhabdoid differentiation. Immunohistochemical analysis revealed strong expression of cytokeratin 7, CAM 5.2, AE1/AE3, wide-spectrum keratin, and epithelial membrane antigen by the epithelial component. The stromal component displayed strong immunohistochemical expression of WT-1, CD-99, CD-56, INI1, and estrogen receptor; focal actin positivity; and was negative for desmin, myogenin, and progesterone receptor. Analysis by reverse transcriptase polymerase chain reaction failed to identify the SYT-SSX1 or SYT-SSX2 fusion transcripts characteristic of synovial sarcoma. To our knowledge, this represents the first report in the literature of malignant MEST with rhabdoid features and suggests that this entity should be considered in the diagnosis of renal stromal malignancies with prominent rhabdoid features.

1. Introduction

Mixed epithelial and stromal tumor of the kidney (MEST)/cystic nephroma (CN) is a recently described neoplasm that predominantly affects perimenopausal women [1-3]. The tumor is typically solid and cystic,

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composed of admixed spindle-cell stroma and epithelial elements [1-4]. The epithelial component may display a tubular, microcystic, macrocystic, or complex branching architecture with a typical hobnail epithelial lining [1-4]. The stromal component consists of a bland spindle-cell proliferation of variable cellularity with occasional areas of myoid change, fascicular growth, and fibrosis. Although characterized by a benign histology and clinical course, malignant features of MEST and CN have recently been identified in a few cases [1,3,5-8]. We describe the histologic, immunohistochemical, and molecular genetic features of a case of malignant MEST with extensive rhabdoid differentiation.

2. Report of a case

An 84-year-old woman was incidentally discovered to have a partially cystic tumor within the left kidney during evaluation for cholecystitis. She demonstrated no clinical abnormalities associated with her kidney lesion, and extensive further clinical evaluation showed the patient to have no other tumors. She subsequently underwent left radical nephrectomy. Follow-up studies of the patient showed no indication of recurrence or presence of other malignancies 17 months after nephrectomy.

3. Pathologic findings

The left nephrectomy specimen contained a tan, partially hemorrhagic, partially cystic 10.5 × 7.0 × 6.5-cm mass. The lesion involved the renal medulla and extended into the renal calyceal system as well as into the renal cortex. The tumor contained multiple, grossly obvious, loculated cysts, with the largest cyst measuring 2.5 cm in diameter.

Histologically, the tumor was composed of epithelial lined cystic and tubular structures with an intervening malignant spindle-cell stroma (Fig. 1). The density of the epithelial component was variable with areas characterized by back to back cysts separated by only a thin fibrous capsule

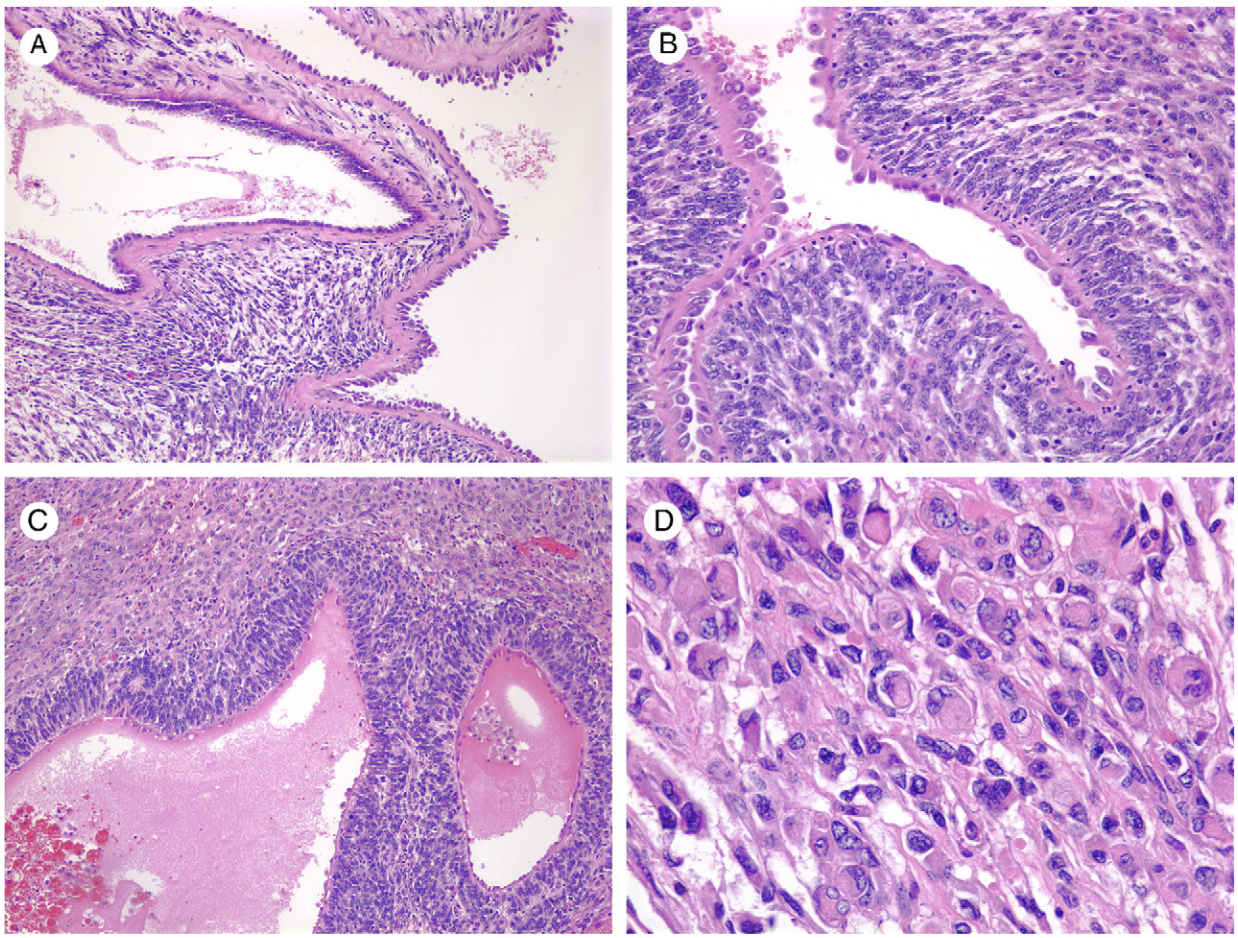

Fig. 1 Hematoxylin and eosin–stained sections of malignant mixed epithelial stromal tumor of the kidney showing characteristic cystically dilated spaces with associated stroma (A). The cysts are lined by a single-cell layer of cuboidal to columnar eosinophilic cells with variable hobnail appearance (B). The malignant stromal condensation forms periepithelial cuffs. The stromal cells show marked atypia and brisk mitotic activity (C). The neoplastic stromal cells demonstrate focal rhabdoid morphology (D).