Case Report

Adult Cystic Nephroma: A Rare Incidental Tumor of the Kidney

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Abstract

Mixed epithelial and stromal tumors/adult cystic nephroma are rare, biphasic, incidentally detected tumors of the kidney. They require surgical intervention. Rare cases show malignant morphology.

Keywords:

kidney, malignant, mixed epithelial and stromal tumor

Introduction

Mixed epithelial and stromal tumor (MEST)/adult cystic nephroma is a biphasic renal tumor comprising spindle cell stroma and epithelium lining the tubules/cystic spaces. They are mostly benign; however, rare cases with malignant morphology have been reported. We present a case of large MEST of the kidney, which was incidentally detected.

Case Report

A 48-year-old female complained of loose motions for two weeks. On examination, the treating physician noted a lump in the left lumbar region. Her CRP was 2.8, creatinine 0.8, BUN 3.9, Sodium 141.6, Potassium 4.52, and Chloride 105.1.

CT Abdomen & Pelvis (Figure 1) showed a large hypodense lesion arising from the left kidney, involving the upper as well as mid-pole region, measuring approximately 14.4 × 12.5 × 14.8 cm (AP × ML × CC). The lesion showed multiple thin enhancing septae causing variable-sized locules. The renal mass extended to involve the renal medulla and pelvis, causing calyceal distortion. It was distending the perinephric space and causing displacement of the bowel loops to the right; the stomach and pancreas were displaced cranially. There was compression and anterior and lateral displacement of the distal transverse and descending colon. There was a single renal artery on both sides. Both renal veins were well opacified. The kidneys showed normal and symmetrical enhancement. The right kidney, urinary bladder, uterus, liver, gallbladder, pancreas, spleen, and adrenal glands were normal.

Impression: Large left renal multilocular cystic lesion with multiple thin enhancing septae, no obvious calcification or enhancing mural nodule, causing calyceal distortion and mass effect as described. Findings were suspicious for neoplastic etiology.

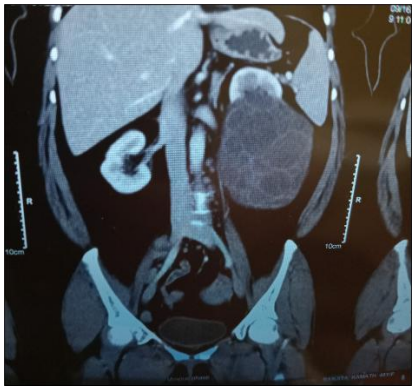

Figure 1: CT Abdomen & Pelvis: Large hypodense multilocular cystic lesion arising from the left kidney.

Patient underwent left nephrectomy. We received an already cut open left nephrectomy specimen measuring 25 × 18 × 15 cm (1685 gms). The left kidney measured 8 × 6.5 × 4.5 cm. The ureter measures 7 cm in length. Renal artery and vein measured 1 cm in length each. The external surface along the mid-pole of the kidney towards the hilar side shows a grey-brown cystic mass measuring 18 × 16 × 10 cm attached to the renal sinus. On cutting open, clear fluid oozed out. The cut surface of the mass was grey-white, multilocular cystic, with locules ranging from 0.3 cm to 6 cm, with variably thickened interspersed grey-white septae (Figure 2). Cyst wall thickness ranged from 0.3 cm to 1 cm. The kidney was already cut open with the capsule stripped off. The cut surface was unremarkable. The renal sinus was congested and irregular. Part of the adrenal gland measures 2 × 1 × 0.8 cm. No lymph nodes were dissected on gross examination from the hilum.

Microscopic examination from the cystic mass represented a mixed epithelial and stromal tumor (Figure 3a), revealing epithelial elements in the form of cystic glands, many of which are dilated and lined by flattened denuded to cuboidal epithelium with clear (Figure 3b) to eosinophilic cytoplasm, surrounded by fibrocollagenous stroma with smooth muscle fibres, ovarian type (Figure 3c) of stroma (positive for ER by immunohistochemistry – Figure 3d), foci of hyalinisation (resembling corpus albicans), and edema. Glands/tubules showed eosinophilic secretions in the lumen at places. The stroma showed mild lymphocytes, histiocytes, plasma cells, and multinucleated histiocytes at places. There was no evidence of atypia, mitosis, or necrosis. Renal parenchyma close to the cystic mass along the renal sinus/medulla showed features of chronic pyelonephritis. The rest of the renal parenchyma was unremarkable. Ureteric, renal artery and renal vein cut margins, perinephric fat, Gerota's fascia, and random ureter were unremarkable. A single tiny unremarkable lymph node was noted along the hilar fat. The impression given was adult cystic nephroma/mixed epithelial and stromal tumor of the kidney.

Figure 2: Gross: Renal mass with grey white multilocular cystic cut surface showing variably thickened interspersed grey white septae.

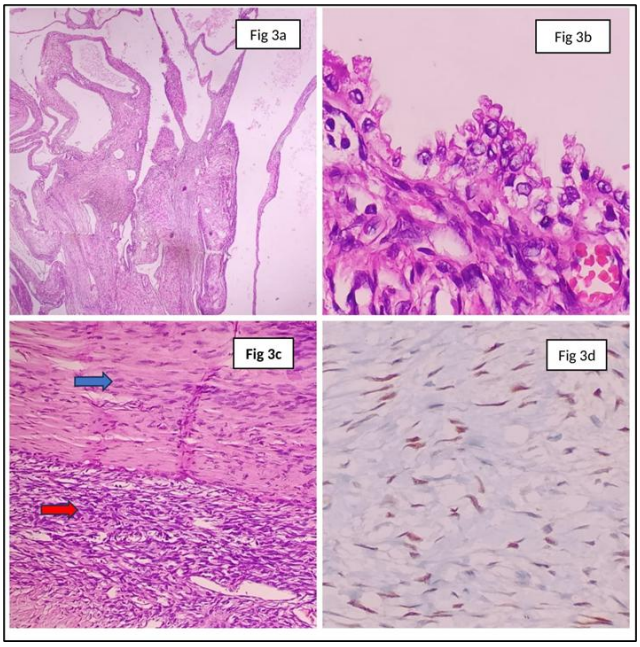

Figure 3: Microscopy: a) cystic mass showing biphasic epithelial and stromal components (HE x 100) b) Epithelium lined by cuboidal epithelium with clear cytoplasm (HE x400), c) fibrocollagenous (blue arrow) and ovarian type of stroma (red arrow) (HE x 100), d) Stromal ER positivity by immunohistochemistry (HE x 400).

Discussion

Mixed epithelial and stromal tumor and adult cystic nephroma of the kidney are rare biphasic renal tumors composed of epithelial cysts lined by benign epithelium (usually of renal cell type) and a proliferation of cytologically benign-appearing spindle-shaped stromal cells. They frequently occur in middle-aged women (M:F ratio 1:7). For the rare tumors that occur in men, there is often a history of hormone therapy. These tumors are mainly asymptomatic. If symptomatic, they mostly present with haematuria and abdominal pain [1].

The mean tumor size is about 6 cm in diameter [2].

Mixed epithelial and stromal tumor (solid cystic with firm white solid area) and adult cystic nephroma (entirely cystic) are well-

demarcated and unencapsulated tumors.

On microscopy, cysts of cystic nephroma are usually lined by flat, cuboidal, hobnail-shaped cells with clear cytoplasm, also sometimes with mucinous goblet, endometrioid type of cells. Cysts may resemble thyroid follicles, complex branching tubules, phyllodes-type architecture, and papillary structures. Septal stroma consists of hypocellular to hypercellular fibrous and smooth muscle stroma, sometimes resembling ovarian stroma. Stromal luteinization and nodules resembling ovarian corpora albicantia can occur. Fat, myxoid change, and edema can occur in the stroma. Scattered mitoses are seldom noted, but there won't be atypia or necrosis. Malignant transformation can rarely occur, presenting as sarcoma arising from the stroma.

By immunohistochemistry, the stromal component expresses actin, desmin, CD10, ER, and PR. The epithelial component expresses Pax2/8 [1].

Surgical intervention (partial or radical nephrectomy) is the preferred modality of treatment [3]. The majority of MEST cases in literature are benign tumors with or without local invasion and without recurrence. Prognosis after undergoing surgical resection is excellent. Rare cases of malignant transformation with aggressively enlarging tumors have been reported, which may display rhabdoid, rhabdomyosarcomatous, and chondrosarcomatous elements [4].

There are no established guidelines for the treatment of malignant MEST. Radical nephrectomy with extended lymphadenectomy had to be performed in suspected malignant cases. MESTs are seen to respond to doxorubicin and ifosfamide chemotherapy [5].

Conclusion

Mixed epithelial and stromal tumour is a rare benign renal neoplasm and presents with non-specific symptoms. Diagnosis is based on histopathology. Outcomes are favourable with surgery alone. However, long-term follow-up is essential considering its rare chances of local recurrence and malignant transformation.

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