



Case Report

Mixed epithelial and stromal tumour with extension to vesicoureteric junction

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Abstract Mixed epithelial and stromal tumour (MEST) is an uncommon renal tumour with a tendency to protrude into the collecting system. We present a 50-year-old woman with a renal tumour extending up to the vesicoureteric junction (VUJ) who was suspected to have an upper tract transitional cell carcinoma for which a nephroureterectomy was performed. Histopathologic examination revealed a MEST arising from the kidney and extending up to the VUJ. To the best of our knowledge, this is the first report of a renal MEST with extension to the VUJ.

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1. Introduction

Mixed epithelial and stromal tumour (MEST) is an uncommon renal tumour occurring predominantly in perimenopausal women. These tumours have been reported to have tendency to protrude into the collecting system. Extension into the pelvis and the upper ureter have been previously reported [1]. We present a case of MEST arising from the left kidney with extension into the collecting system and the ureter up to the vesicoureteric junction (VUJ).

2. Case report

A 50-year-old post-menopausal woman presented to us with left flank pain of 3 months duration. On examination she was found to have a 15 cm × 10 cm bimanually palpable left flank mass. Routine blood investigations were within normal limitations. Urine microscopy revealed microscopic haematuria. Urine cytology showed dysplastic cells. On contrast enhanced computed tomography (CECT) (Fig. 1) a complex multicystic left renal mass was seen with extension to the pelvi-calyceal system and the ureter up to the left VUJ. No retroperitoneal lymphadenopathy was evident. A cystoscopy examination did not reveal any abnormality. A left retrograde ureterogram was attempted, however the ureteric catheter could not be negotiated beyond the VUJ.

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Figure 1 Contrast enhanced computed tomography abdomen. (A) Complex cystic mass of left kidney. The mass is seen extending to the upper ureter; (B) Mass seen extending to mid ureter; (C) Mass seen in lower ureter.

A metastatic evaluation including a chest CT and liver function tests were performed with negative results.

With a provisional diagnosis of a left upper tract transitional cell carcinoma, we proceeded with left nephroureterectomy. Intraoperatively a large renal mass was found. The entire left ureter up to the VUJ was grossly dilated and tortuous with the mass palpable within. Left nephroureterectomy with excision of the VUJ and adjacent bladder cuff by an extravesical technique maintaining a closed system was accomplished.

On gross pathologic examination (Fig. 2) the left kidney was grossly enlarged with a bosselated surface. On bivalving the kidney a pale white polypoidal mass was seen arising from the central portion of the kidney extending to the pelvi-calyceal system and the entire ureter till the VUJ. The cut surface of the tumour had a gelatinous consistency.

Microscopy (Fig. 3) revealed large cysts, microcysts and tubules lined by columnar to cuboidal epithelium admixed with stroma containing spindle cells with plump nuclei and abundant cytoplasm. This suggested a diagnosis of MEST. The patient was counselled regarding the diagnosis and advised to follow up.

The patient was followed up with a CECT at 3 months and subsequently abdominal ultrasonography every 6 months. At the time of this report, she remains well with no evidence of disease recurrence at 48 months post-surgery.

3. Discussion

MEST is a relatively uncommon renal tumour first described by Michal and Syrucek in 1998 [2] and was subsequently included in the WHO classification of tumours in 2002 [3]. These tumours have been described by diverse nomenclature in the past such as adult mesoblastic nephroma, solid and cystic biphasic tumour of the kidney and cystic hamartoma of the renal pelvis. These tumours occur predominantly in middle-aged or older women. Their occurrence in women on hormone replacement therapy and in men receiving androgen deprivation or oestrogen therapy for prostate cancer has suggested an association with oestrogen exposure [4].

In most series these tumours have been symptomatic with flank pain, haematuria and recurrent urinary infections as the common presenting feature. Up to 25% cases are incidentally detected on imaging studies [5].

On imaging, MEST appears as well-circumscribed multi-septate cystic mass with thin or thick septae and enhancing solid components, and is usually characterised as Bosniak III or IV cysts. A pure solid pattern has also been reported [6].

MEST has been noted to have a peculiar propensity to extend into the collecting system. Protrusion into the renal pelvis and up to the upper ureter has been reported [1]. To the best of our knowledge, this is the first reported instance

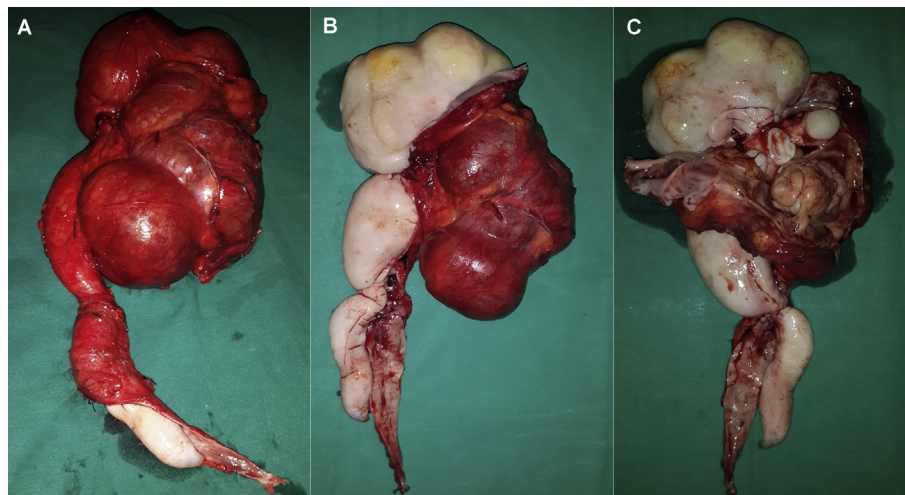


Figure 2 Gross specimen. (A) Left renal mass with bosselated surface and the lower ureter that has been bivalved showing the mass extending up to the vesicoureteric junction; (B) Specimen with the pelvis and ureter bivalved; (C) Bivalved kidney showing the full extent of the mass.

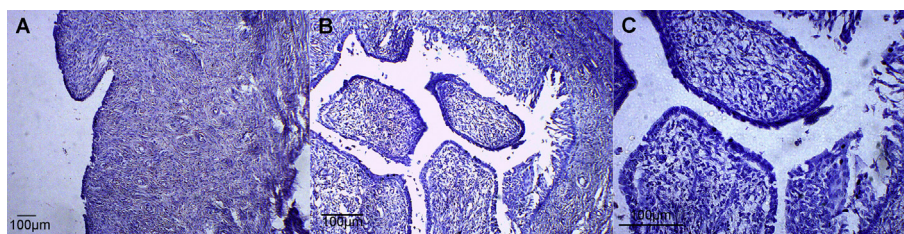


Figure 3 Photomicrograph. (A) Parts of cyst wall containing spindle cells and numerous congested blood vessels, lined by flattened to cuboidal epithelium; (B) and (C) Sections of tumour showing microcystic spaces lined by cuboidal epithelium.

of extension to the VUJ. MEST has been hypothesised to arise from the remnants of the metanephric blastema, subsequently undergoing both mesenchymal and epithelial differentiation. An alternative origin of MEST from a hormone sensitive periductal foetal mesenchyme has also been postulated [7]. We hypothesize that the tendency to protrude into the collecting system may be attributed to the origin of the tumour from the central portion of the kidney from cells close to the collecting system. The vast majority of these tumours are benign and lack the ability to invade tissue, hence they grow along the path of least resistance, i.e., into the collecting system and the ureter.

On microscopy a biphasic pattern of epithelial elements comprising cuboidal or columnar cells and stromal component comprising spindle cells were seen.

The differential diagnosis includes adult cystic nephroma (ACN), complex renal cyst, angiomyolipoma with epithelial cysts, and multilocular cystic renal cell carcinoma (RCC). The differentiation of ACN from MEST remains a matter of debate with some authorities suggesting that these entities represent a spectrum of the same entity, with a variable ratio of stromal to epithelial components [8].

A preoperative diagnosis of MEST cannot be reliably made based on imaging studies. As such these patients are subjected to radical or partial nephrectomy with a diagnosis of MEST made postoperatively on pathologic evaluation.

All attempts should be made to achieve negative surgical margins as positive margins have been associated with local tumour recurrence. Recently malignant transformation has also been described which again emphasises the need for complete resection [9]. Tumour spillage should also be avoided as it has been associated with peritoneal seedling [10].

The vast majority of MEST behave in a benign manner and have a good prognosis. Malignant transformation of the epithelial, stromal or both elements has been reported and portends a poor prognosis [11].

4. Conclusion

To summarise, MEST must be suspected in a patient presenting with a complex cystic renal mass particularly if there appears to be an extension into the collecting system, without evidence of local or distant tumour spread. Although these tumours are predominantly benign, malignant transformation, local recurrence and tumour seedling have been reported. Tumour excision with clear margins should be the standard of care.

Author contributions

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Final approval of manuscript: Arun Ramdas Menon, Nivedita Suresh, Suraj Hegde, T.P. Rajeev.

Conflict of interest

The authors declare no conflict of interest.

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