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Clear cell variant, urothelial carcinoma of ureter: A rare entity

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ABSTRACT

Clear cell variants of transitional cell carcinoma of upper urinary tract is extremely rare. Although clear cell of urinary bladder has been reported, its occurrence in the upper tract has not been reported readily in literature.

We present a case of 77 year old female who was suspected with TCC of the ureter, but was found to have clear cell variant on histology post radical nephroureterectomy.

This was the first such case in our institution.

1. Introduction

Urothelial carcinoma is the most common cancer of the urinary tract. Urothelial carcinoma may show many histological variants that mau include squamous differentiation variant, glandular differentiation variant, nested variant, microcystic variant, micropapillary variant, lymphoepithelioma-like variant, lymphoma-like variant, plasmacytoid variant, sarcomatoid variant[1] and so on.

Clear cell adenocarcinoma (CCA) is a rare histologic subtype of adenocarcinoma in the urinary tract. The tumour has histomorphological features resembling CCA of the female genital tract (or Müllerian origin).[2]

CCA occurs almost exclusively in the urethra and urinary. CCA of ureter is extremely rare. Here we describe a case of 77 year old female who was diagnosed with clear cell carcinoma of the upper ureter post surgery.

2. Case presentation

A 77 -year-old female with no co-existing comorbidities presented to urology OPD in our hospital with a history of left flank pain and haematuria. A CT scan revealed a 6 mm filling defect in the left renal pelvis and lower pole calyx. There were no enlarged lymph nodes or any other co existing lesions in the genito-urinary tract (Fig. 1). A uretereroscopic brush cytology was taken which was suggestive of malignant cytology.

The patient underwent left radical nephroureterectectomy. Pathologic examination of the specimen revealed a $2 \times 1.5 \times 0.6$ cm tumour in the lower calyx of the renal pelvis extending into the ureter. Renal sinus was free of tumour, vascular and ureteral margins were negative with no lymphovascular invasion. To our surprise histological subtype was invasive papillary urothelial carcinoma with clear cell change. Tumour cells showed clear cytoplasm, round nuclei with evenly distributed chromatin and inconspicuous nucleoli (Fig. 2, Fig. 3).

Drain was removed on post op day 3, Catheter was removed on post op day 4 and the patient was discharged on post op day 7.

On follow up 1 month after surgery, the patient was well with no complaints.

3. Discussion

CCA of the female genital tract has a characteristic histologic appearance with papillary and/or tubulocystic pattern and the presence of clear cells and hobnail cells. The occurrence of the same in the upper urinary system however is very unusual.

It has been proposed that CCA in the upper tract may originate from a remnant or derivative of mesonephric duct or Müllerian duct, nephrogenic adenoma, or urothelium[3]

Histologically, CCA of the renal pelvis is difficult to differentiate from clear cell or papillary RCC and the other types of carcinoma of urothelial origin showing clear cell features, which include urothelial carcinoma and adenocarcinoma of non-CCA type.[4] Especially in females patients, metastasis from CCA of gynecologic origin should be ruled out before embarking on a surgical therapy.

The distinction of CCA from other types of urothelial carcinoma with clear cell features is difficult due to the rarity of CCA in the renal pelvis. Non CCA types of urothelial carcinoma can be excluded by positive immunoreaction for HNF and PAX8 (or PAX2)[5] Nephrogenic adenoma, may be a differential diagnosis of CCA but nuclear pleomorphism,

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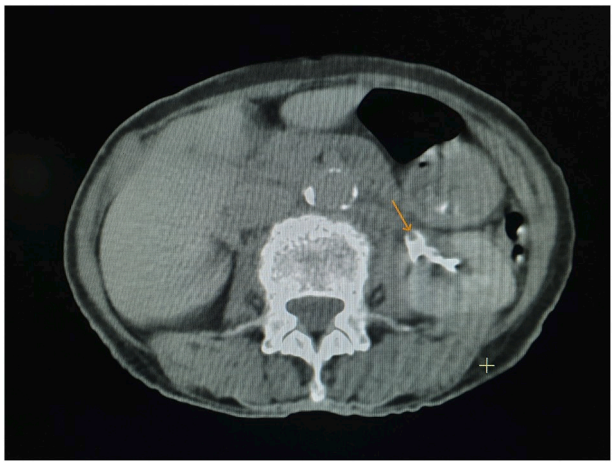

Fig. 1. CECT showing filling defect in left renal pelvis.

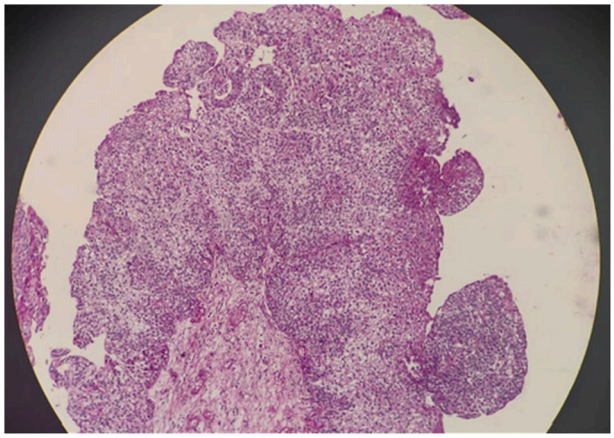

Fig. 2. Histology showing clear cell variation in tumour cells.

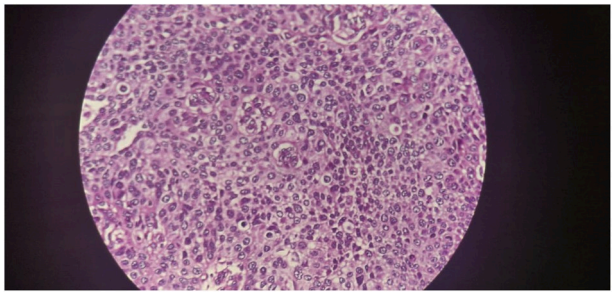

Fig. 3. High power magnification of tumour cells.

diffuse nuclear hyperchromasia, and high mitotic rate favour the latter.

CCA of the lower urinary tract is aggressive with majority (76%) of the patients having recurrent or metastatic disease, whereas only 24% of patients survived with no evidence of disease after treatment. Due to the rarity of CCA in the urinary tract, an effective treatment regimen remains to be determined.

4. Conclusion

Upper tract carcinoma is mostly transitional type. Clear cell carcinoma is very rare which can be diagnosed on histopathology. Further studies are needed for defining the prognosis of upper tract clear cell carcinoma.

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