CASE REPORT

Multilocular cystic renal cell carcinoma: a rare entity

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SUMMARY

Multilocular cystic renal cell carcinoma (MCRCC) is an uncommon subtype of clear cell renal cell carcinoma that appears to have a favourable prognosis. Literature reports a very low incidence of 1–2% of MCRCC among the renal neoplasms. The first such reported case was in 1957 by Robinson. In 1998, Eble *et al* suggested following diagnostic criterion for MCRCC: (1) an expansile mass surrounded by a fibrous capsule; (2) interior of tumour entirely composed of cysts and septa with no expansile nodule or solid component confined to more than 10% of entire tumour and (3) septa containing aggregates of clear epithelial cells. We report a case of MCRCC in a 60-year-old male patient who presented with abdominal lump and after suspicion of renal malignancy underwent nephrectomy. The patient was perfectly well till last follow-up of 3 months postoperatively.

BACKGROUND

Multilocular cystic renal cell carcinoma (MCRCC) is a rare entity of renal neoplasms and has been recently added to the WHO classification of renal neoplasms.^{1–4} Because of its close resemblance to other cystic renal lesions, it tends to get missed. This manuscript has been written to provide clear-cut distinction between the cystic renal neoplasms, by citing this case, as otherwise MCRCC has an excellent prognosis and often does not show recurrence or distant metastases unlike its few other differentials.

CASE PRESENTATION

A 60-year-old male patient presented to the urology out patient department of our institution with chief complaints of gradual distension of right side of abdomen since 3 months and difficulty in micturition since 15 days. There was no history of any associated pain, hematuria, loss of appetite or weakness. On palpation, a non-tender balotable mass was felt over right lumbar region.

INVESTIGATIONS

Urinanalysis showed mild proteinuria with non-significant proteinuria and a few atypical epithelial cells in background. CT scan of the lower abdomen revealed a well-defined and margined round hypodense lesion measuring 12×10×5 cm involving almost whole of kidney with evident internal septations and few small necrotic areas (figure 1A). Nephrectomy was planned and dissection of the right kidney was carried out via right subcostal incision. We received the formalin-fixed gross nephrectomy specimen which revealed a well-encapsulated, multiseptate tumour mass with

variegated appearance involving upper and middle lobes of the kidney. Cut section of the tumour showed multiple non-communicating cysts, ranging from 0.2 to 0.6 cm in diameter and filled with serous fluid. No solid nodules or papillary projections were seen (figure 1B). No lymph nodes or adrenal gland were identified or received with the specimen. Microscopic sections revealed a multilocular, multicystic tumour mass with cysts separated by fibrocollagenous connective tissue lined by a mixture of cells including flattened to cuboidal and clear cells. The clear cells had small hyperchromatic nuclei with mild anisonucleosis and inconspicuous nucleoli with Fuhrman's nuclear grading 1/2 (figure 2). Tumour cells were limited to renal capsule. Ureter showed changes of chronic non-specific inflammation. There was no evidence of tumour extension to major vessels and perinephric fat. Adjacent renal parenchyma showed focal thyroidisation of tubules with interstitium infiltrated by chronic inflammatory cells. Minor vessels showed mild intimal thickening but were otherwise unremarkable. Based on the above findings, a final diagnosis of multilocular cystic variant of renal clear cell carcinoma; Fuhrman's nuclear grade 1/2 with tumour size, node status, metastasis (TNM) staging T_{1b}N₀M₀ was made.

DIFFERENTIAL DIAGNOSIS

The chief differential diagnosis of MCRCC lies with other cystic renal neoplasms and it is often very difficult to distinguish them based on clinical, radiological and gross features alone and microscopy aided with immune histochemistry is essential. MCRCC is believed to be a part of the spectrum of cystic renal neoplasms with cystic nephroma at the benign end and tubulocystic carcinoma with cystic variant of renal cell carcinoma at the malignant end. Cystic nephroma chiefly involves women and

Figure 1 (A) CT abdomen: Well-defined and margined round hypodense lesion involving whole of kidney with evident internal septations and few small necrotic areas. (B) Gross: Well-encapsulated, multiseptate tumour mass with variegated appearance involving upper and middle lobes of the kidney.

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Figure 2 Multilocular multicystic renal cell carcinoma with thin fibrous septa lined with clear cells with grade 1/2 nuclei (inset).

there is no evidence of clear cells in stroma, albeit it is ovarian type. In tubulocystic carcinoma, the cyst lining cells have abundant eosinophilic cytoplasm with higher grade nuclear features. Cystic variant of renal cell carcinoma can be secluded on the basis of presence of cystic as well as solid areas, the communicating cysts and higher grade nuclear features.⁴

TREATMENT

Nephrectomy was planned and dissection of the right kidney was carried out via right subcostal incision.

OUTCOME AND FOLLOW-UP

Postoperative recovery was uneventful and the patient was discharged after 1 week with the advice for regular follow-up. Till the last follow-up, approximately 3 months after surgery, the patient was absolutely fine.

DISCUSSION

MCRCC, also known as multilocular clear cell carcinoma and multilocular cystic clear cell renal cell carcinoma, is an uncommon subtype of renal cell carcinoma. Men are more commonly affected than women.⁵ Genetic studies have shown its association with 3p deletions similar to clear cell renal cell carcinoma.⁶ The mandatory histological criterion to classify a renal tumour as MCRCC comprises of presence of a benign, circumscribed, expansile lesion composed entirely of non-communicating cysts and septa lined by a single layer of low-grade clear cells.⁷

As MCRCC presents with lower nuclear grade (Fuhrman's grade I–II) and contains lesser number of malignant cells as compared with solid renal cell carcinoma, there is very low

potential for recurrence and metastasis, and hence the prognosis is extremely good. Suzigan *et al*¹ have also reported in their series of 45 cases of MCRCC, a 100% disease-free survival rate at a mean of about 66.1 months after surgery. Nephron sparing conservative surgery is usually the treatment of choice as the tumour cells are confined to small aggregates within the septa only.^{8–10}

Learning points

- Multilocular cystic renal cell carcinoma (MCRCC) is an entity that one should keep in mind while evaluating the cystic renal masses. As it has a low malignant potential and an excellent prognosis, a correct diagnosis of MCRCC serves in planning the postoperative follow-up schedule accordingly and also alleviates the patient as well as clinician from fear of recurrence or any other complication.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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