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Case Report:
Multilocular Cystic Renal Cell Carcinoma: An Unusual Gross Appearance

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Abstract:
Multilocular Cystic Renal Cell Carcinoma (MCRCC) represents a rare variant of clear cell (conventional) renal cell carcinomas. Attributable to its distinct characteristics in prognosis and its natural history, MCRCC was recognised as a separate subtype of renal cell carcinoma in the 2004 WHO classification of adult renal tumors. We report this case of MCRCC from antemortem surgical specimen, due to its unusual gross appearance and a rare clinical entity.
Key Words: Multilocular Cystic renal Cell Carcinoma; Renal Cell Carcinoma

Introduction:
Multilocular Cystic Renal Cell Carcinoma (MCRCC) is a distinct subtype of clear cell renal cell carcinomas and appears to have a favourable biology. These tumours are a rare clinicopathological entity, comprising 1 to 2 % of all renal tumors.[1] MCRCC is an uncommon tumour of the kidney composed of multiple cysts with clear cells in the septae indistinguishable from grade 1 RCC.[2] Computerised tomography (CT) is the most valuable investigation and the diagnosis is established by histopathological findings.

Case Report:

A 70 years old male presented with left flank pain since two months. The pain was of insidious onset, dull aching and non radiating. Ultrasound revealed a mass in upper pole of left kidney. The mass was mildly vascular on Doppler and showed small cystic regions. CT showed on exophytic mass in left kidney, replacing the upper pole parenchyma with loss of cortico medullary differentiation, infiltrative in nature, involving the upper pole. Renal function tests were within normal limits. A left nephrectomy was carried out and the specimen was sent for histological study in department of pathology. Post operative period was uneventful. Histopathological specimen grossly measured 11×7×7 centimetres, which included the tumour measuring 7x6 centimetres in diameter. Tumour was well circumscribed and seen reaching up to the capsule. Cut section of the tumour was multiloculated cystic. The cysts measured 0.2 to 2.5 centimetres in diameter. Cysts contained haemorrhagic fluid and blood clots (Figure 1). Microscopically the sections showed multicystic tumour, cysts separated by thin fibrocollagenous septae lined by aggregated clear cells. The clear cells had mild hyperchromatic nuclei with mild anisonucleosis and inconspicuous nucleoli (Figures 2 and 3). Pelvis, ureter, adrenals and renal vessels were not involved by the tumour. The histomorphology was compatible with diagnosis of MCRCC Fuhrman nuclear grade 1. The patient is well with no recurrence.

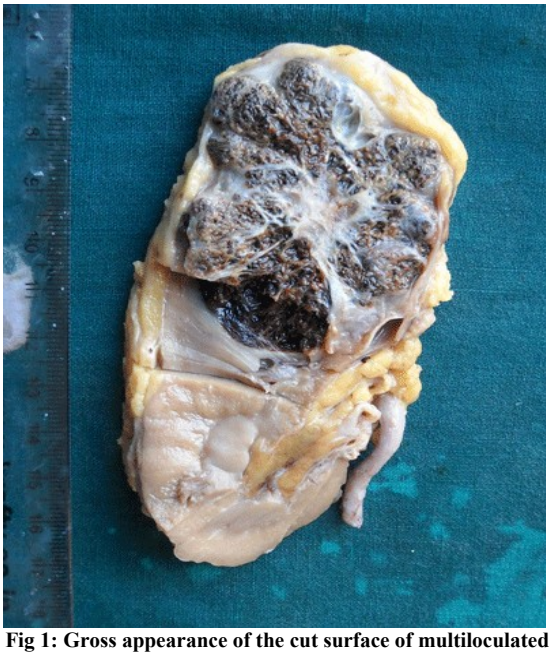

Fig 1: Gross appearance of the cut surface of multiloculated cystic kidney

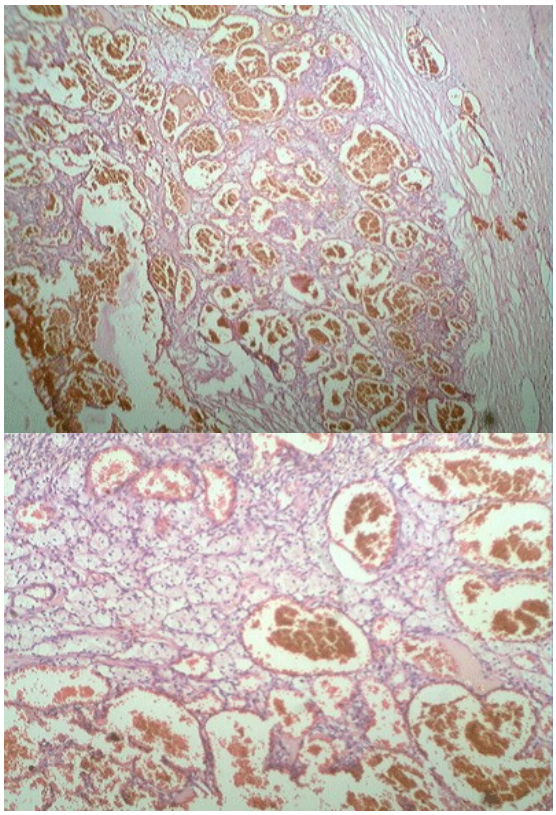

Figure 2 and 3: Photomicrograph showing renal tumour composed of multiple cysts of varying sizes separated by thin connective tissue septae lined by aggregated clear cells, cysts containing haemorrhagic fluid. (H&E X100, X400)

Discussion:

Renal cell carcinoma (RCC) accounts for 80 to 85 % of primary malignant neoplasms of kidney.[3] It usually presents as a solid mass, however in 10 to 22 % of cases RCC appear as unilocular or multilocular cystic mass on imaging studies.[3] The 2004 W.H.O classification of kidney tumours recognises MCRCC as a rare distinct variant of clear cell RCC with good prognosis.[4] The term MCRCC should be exclusively used to identify cystic RCC with 15 to 25 % of neoplastic clear cells in the cyst wall.[5] When the imaging studies demonstrate a cystic renal mass, MCRCC should be included in the differential diagnosis.[6] Other possibilities are cystic nephroma and mixed epithelial and stromal tumour of kidney.[7] Multilocular cystic nephroma is an uncommon benign entity, grouped among the cystic non genetic diseases. It is characterised by multilocular cysts lined by a single layer of flattened, low cuboidal epithelium with no nuclear atypia. The cytoplasm is eosinophilic and fibrous septae within cyst wall resemble ovarian stroma and may contain mature tubules. Clear cells must not be found in the wall or stroma of such cases.[7] Since, MCRCC has good prognosis, early diagnosed, and is of low nuclear grade, renaming it as multilocular cystic RCC of low malignant potential may help the urologist approach the patient conservatively, namely nephron sparing surgeries.

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