Case report on multicystic nephroma

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Abstract
Cystic renal neoplasms of the kidney can be benign or malignant. Multicystic nephroma (MCN) represents a rare benign cystic lesion of the kidney, which usually presents as a unilateral multicystic renal mass without solid elements. According to the World Health Organization (WHO) classification of the renal neoplasms, it is grouped along with mixed epithelial–stromal tumor of the kidney.

Keywords: Multicystic nephroma.

Introduction
Multilocular cystic nephroma (MLCN) is a benign mixed mesenchymal and epithelial neoplasm of the kidney. MLCN is classically described as a benign slow-growing renal tumor, and there are more than 200 cases reported in the literature. Few articles report an exact prevalence of MLCN; however, of 165 primary renal neoplasms, Gallo and Penchansky estimated MLCN prevalence to be 2.4% in their institution. Children younger than 2 years old and adults in middle age (40–69 years old) are more commonly affected. Some case series describe a higher incidence in male children (male-to-female ratio, 3:1) and in postmenopausal women (female-to-male ratio, 9:1).

Case Report
A 62-year old female presented with complaints of heaviness in left flank with occasional pain. There is no history of any undue weight loss, nausea, fever. There is no history of any chronic illness/malignancy in the family or relatives. Systemic examination revealed a vague lump in the right renal region, extending from left renal to left hypochondrium. Lump was non-tender, and minimally mobile on respiration and ballotable s/o renal mass. Hematological and biochemical investigations were within normal limits. Radiologically CECT Abdomen revealed multiloculated, enhancing left renal cyst with few septa occupying more than half of the kidney (Fig. 1). Plan for laparoscopic radical nephrectomy was taken. Intraoperatively few adhesions were found with surrounding structures. Adhesionlolysis were done and kidney was dissected, vessels were clamped and radical nephrectomy was done. Multiple loculated cysts were found containing clear fluid with minimal renal parenchymal (Fig. 2). Wound was closed with a drain in the right hepatorenal pouch which was removed on POD 3 subsequently. Post operative period was uneventful and smooth and the patient was discharged on POD 5 satisfactorily. The postoperative histological report showed columnar to transitional epithelium with focal areas of hobnail appearance of epithelium suggesting multicystic nephroma.

Discussion
MCN has at least 20–25 synonyms, which include multilocular cystic renal tumor, benign multilocular cystic nephroma, polycystic nephroblastoma, and so on.

The pathogenesis of MCN is controversial and its classification is confusing. There are numerous proposed theories indicating the etiology as a developmental defect.[1,2] It has also been postulated that it could be neoplastic in origin, probably arising from the ureteral bud.[12] Various pathological criteria have been proposed in the past to differentiate and classify this entity. First diagnostic criteria were formulated in 1951[2] and later modified in 1956[13] and include the following: (a) lesion must be multilocular, (b) the cyst for the most part lined by epithelium, (c) the cyst must not communicate with renal pelvis, (d) the residual renal tissue should be essentially normal, except for pressure atrophy, and finally (e) no fully developed nephrons. The terminology was modified in 1989, which emphasized neoplastic rather than developmental origin.[9] It was broadly segregated into MCN and cystic partially differentiated nephromablastoma.[9] In MCN, multiple cysts showed septa consisting of mature, well-differentiated tubules among the fibrous tissue. Lesions which were predominately cystic without nodular or solid components and containing blastemal elements were noted as cystic partially differentiated nephroma, especially those occurring in the age group less than 2 years. Further subclassification of cystic partially differentiated nephroma was attempted to predict the aggressiveness of this entity based on the content of septal stromal elements, and presence of more than 50% of mature septal element was classified as grade I and less than 50% as grade II.[9]

Patients usually present with nonspecific symptoms. Abdominal pain, hematuria, and urinary tract infection are common in adults. Hematuria can be seen in all age groups and is thought to be due to extension of tumor into the renal pelvis.[14,15] Presentation can sometimes be with severe colicky abdominal pain due to spontaneous rupture of the cyst,[16] which can lead to a clinical diagnosis of urinary stone disease. Loin pain was the presentation in this case. It usually affects single kidney, although rarely bilateral MCN

has been reported.[3,6,17] Lower pole of kidney is the most favored site and the upper pole is the least favored; however, it can arise from any portion of the renal parenchyma. Interestingly, in this case, upper pole was the site of presentation.

Distinct radiographic features have been described but are not universally present in all cases. Ultrasound is often the first investigation used in evaluating abdominal masses, confirmed by CT scan. The sonographic findings depend on the amount of stromal tissue and size of locules. Cysts usually show up as hypoechoic lesions delineated by hyperechoic septae and this feature can be suggestive of MCN but not diagnostic.

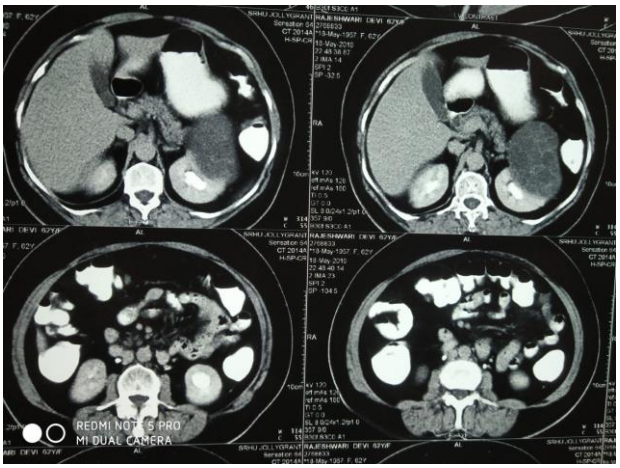

Fig. 1:

The mass is often easily demonstrable at ultrasound, with an average diameter of approximately 10 cm.[18] Our case had a diameter of 10.2 cm. If the cysts are small, the mass may demonstrate internal scattered echoes but without distinct loculations.[3] Calcification has been described as a rare feature of MCN,[6] and curvilinear calcifications may be seen on ultrasound within the septa. Both needle-guided aspiration and color Doppler ultrasound have been proposed to help differentiate between benign and malignant multilocular cystic lesions.[19] CT in this case showed multiloculated cystic lesions with locules of varying sizes, and none of the lesions displayed communication between the cysts or communication between the cysts and the renal pelvis. Lesions contained both thick and thin septa, displaying measurable enhancement on CT. However, none of the lesions contained solid elements on either imaging or on histological analysis.

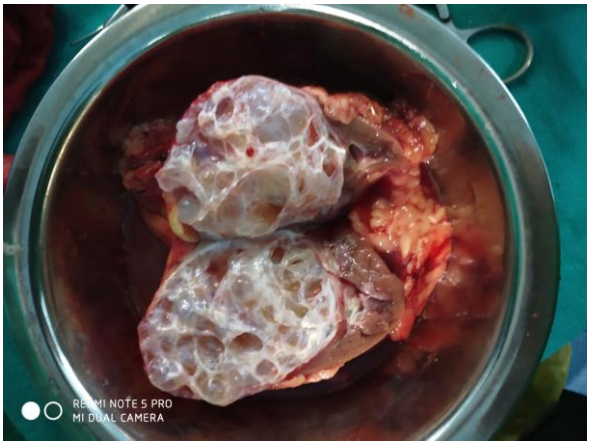

Fig. 2: Cut section of Multicystic nephroma

Although the MCN is not a pre-malignant condition, there are case reports of co-existing foci of renal cell carcinoma in the lining of the cyst wall. MCN is considered to be of benign nature in adults; tumor recurrences have been observed and whether these recurrences are related to the missed foci of malignant area or sarcomatous degeneration is not clear.[19] These recurrences are more often seen after partial nephrectomy. So far, only four cases of local recurrences have been reported, all following partial nephrectomy.[20] However, in a series of 24 patients who underwent partial nephrectomy for MCN, no recurrences were found after a mean follow-up of 39 months.[6] More recently, a case of percutaneous treatment has been described in treating MCN. Percutaneous endoscopic resection of a portion of the cyst protruding into the renal parenchyma was performed, and follow-up CT 4 weeks later revealed complete resolution.[21] However, 3 years later, the authors reported failure with percutaneous approach.[22]

Conclusion

Multilocular cystic nephroma is an uncommon cystic lesion of the kidney and should be considered in the differential diagnosis of malignant cystic renal tumors in both children and adults. Whilst it is important to consider the diagnosis of MCN for any multicystic mass, as our series of six cases has shown, a definitive diagnosis can only be made following surgical treatment with total or partial nephrectomy depending on the size and location of the lesion in the kidney.

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None.

Conflict of interest

None.

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