Case report

Multicystic nephroma--report of two rare cases in a span of seven years

P Paudyal[1], M Agarwal[1], A Pradhan[1], AK Sinha[1], AK Karak, R Agarwal[2]
[1] Department of Pathology and [2]Department of Surgery
B P Koirala Institute of Health Sciences, Dharan, Nepal

Abstract

Background: Multicystic nephroma (MCN) is an uncommon but distinctive benign pediatric renal neoplasm but may present clinically at any age. It is scantily reported in the literature. To the best of our knowledge it has not yet been reported from Nepal. It is characterized by a well circumscribed mass with nodular outer surface that contain multiple fluid filled locals. Microscopy show cystically dilated spaces lined by flattened to columnar epithelium and is separated by fibroblastic stroma. A hobnail pattern is common. **Case description:** Here we report two cases of multicystic nephroma in a span of seven years. The first case is of a one year old female child who was diagnosed of having huge left sided renal mass clinically and the second case is of a 35-year-old male revealing hydatid cyst in USG, CT scan and IVP. Both of them underwent nephrectomy and their microscopic examination revealed typical characteristics of a multicystic nephroma. **Conclusion:** Since MCN has a benign behaviour it must be differentiated from focal cystic neoplastic lesions, including cystic partially differentiated nephroblastoma (CPDN), which has a low but distinct capability for local recurrence; from Wilm's tumour with cystic change; and cystic renal cell carcinoma.

Keywords: benign renal neoplasm, hobnailing, multicystic nephroma

Introduction

Multicystic nephroma (MCN) is an uncommon but distinctive benign renal neoplasm with excellent prognosis.[1] It represents 2-3% of all primary renal tumors in pediatric age group but may present clinically at any age. It needs to be differentiated from cystic partially differentiated nephroblastoma (CPDN) and from other renal neoplasms showing extensive cystic changes.[2] MCNs are commonly found incidentally on radiographic studies, but may present as an abdominal mass found on routine physical examination. MCN and CPDN/solid nephroblastoma (Wilms' tumor) represent benign and malignant ends of a spectrum, respectively; however, the exact relationship between these entities is not known and is rather controversial. Differentiation between these entities has a prognostic and diagnostic significance and may be impossible by clinical and radiological examination.[3] Since this tumour is often confused clinically and radiologically with other renal lesions, most patients undergo nephrectomy in absence of an accurate preoperative diagnosis. MCN can easily be diagnosed histologically and therefore biopsy should be considered to prevent an unnecessary nephrectomy.

Address for correspondence
Punam Paudyal
Associate Professor
Department of Pathology
B.P.Koirala Institute of Health Sciences, Dharan
Email: drpunampaudyal@gmail.com

Case Reports

Case 1

First case is of a one-year-old female child who was diagnosed of having asymptomatic left sided huge renal mass. It was clinically

diagnosed as Wilms tumor. The gross appearance of the nephrectomy specimen showed a large cyst containing clear fluid with multiple small cystic spaces and solid areas having gelatinous, myxoid, whitish appearance. Cystic spaces ranged from 1 to 6 cm in the maximum diameter. Normal appearing kidney was seen at one pole.

Case 2

Second case is of a 35-year-old male who presented to the outpatient department with the complaint of persistent right flank pain of two months duration. On physical examination, there was a mass at right postvertebral angle. Investigations including USG, CT scan and IVP revealed features of hydatid cyst. On gross examination the nephrectomy specimen showed multiple cystic spaces, filled with clear fluid like a honeycomb. Normal appearing kidney was not seen in the specimen. The specimen was completely replaced by the tumor (Figure 1)

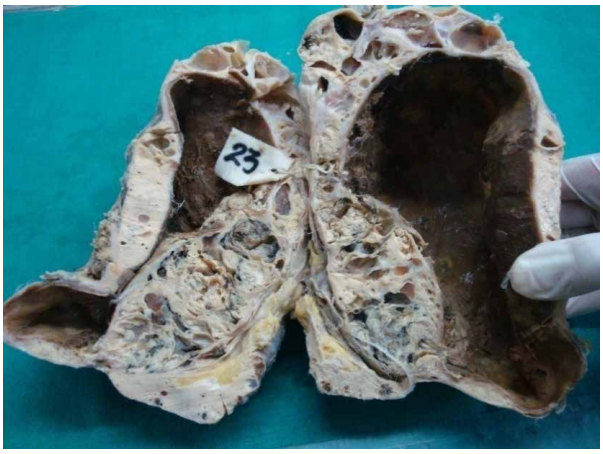

Figure 1: Gross photograph of nephrectomy specimen revealing multiple cystic spaces of various sizes replacing the kidney almost completely.

Histological study (Figure 2) in both the cases showed cysts lined by a single layer of flattened to cuboidal cells having hobnail appearance. Among cysts, a moderately cellular stroma is seen, consisting of fusiform cells with regular oval nuclei.

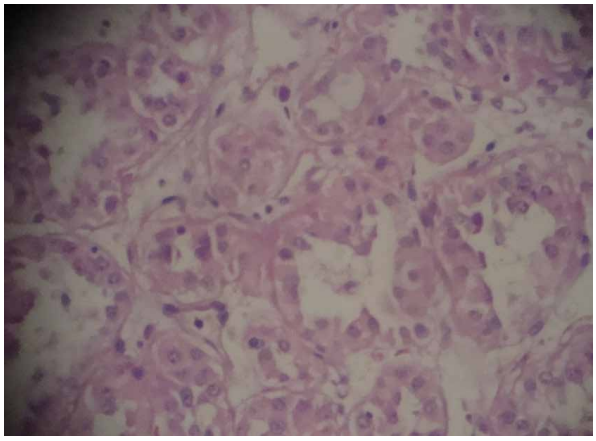

Figure 2: Cysts lined by a single layer of flattened to cuboidal cells with hobnailing. (40x H&E)

Our both the patient had an uneventful recovery after undergoing nephrectomy.

Discussion

MCN is a rare, non-heritable, unilateral benign renal neoplasm of uncertain cause. The tumor was originally described by Edmunds in 1892 as a "cyst adenoma" and since then, the spectrum of histologic findings and multiple theories of the pathogenesis of cystic nephroma have given rise to many synonymous terms, including benign multilocular cystic nephroma and cystic nephroblastoma.[5,6] Less than 200 cases have been reported till date in the international literature.[4] Studies have confirmed a biphasic age and sex distribution: two-thirds of multi-locular cystic renal tumors occur in a predominantly male pediatric population between three months and two years of age; approximately one-third occurs in the female population, with a peak in the fifth and sixth decades of life.[7] The common clinical features in adults are flank pain, hematuria, hypertension and urinary tract infection while painless abdominal mass is common in children.[8] In the case reported here, adult case presented with flank pain and there was huge mass in left flank in case of the child. Precise pre-operative diagnosis of MCN by clinical and radiological means may be impossible and the final diagnosis is made by surgical excision and histopathological examination.[9] Here in our report also the radiological findings were not

consistent with the gold standard histological findings. To our knowledge, however, there have been no reports of cystic nephroma demonstrating local aggressive behavior or malignant transformation. MCN is diagnosed by set of eight criteria as suggested by Powell et al in 1951. These are (i) unilateral involvement (ii) solitary lesion (iii) multilocular nature (iv) non communication of the cysts with one another (v) non communication with the renal pelvis (vi) loculi lined by epithelium (vii) interlobular septa devoid of renal parenchyma (viii) normal residual renal tissue if present.[10]

In two pediatric cases of MCN reported from India computed tomography in both the cases revealed a unilateral cystic lesion in the lower pole of kidney. Keeping in mind the age, clinical presentation and radiologic appearance, a possibility of Wilms' tumor with cystic change couldn't be ruled out preoperatively and both the patients underwent nephrectomy. Histopathologic examination showed it to be a multicystic nephroma.[11]

Similarly, in a case reported in a 7-month-old female with a left sided abdominal lump diagnosis of Wilms' tumor was made based on computed tomography. Fine needle aspiration cytology the child reported as rhabdomyosarcoma. However, histopathologic examination showed it to be a multicystic nephroma.[2] This points out gross disparity in radiological as well as clinical diagnosis, from the histopathological diagnosis in such cases.

Conclusion

MCN of kidney carries an excellent prognosis. If the pathologist can make a pre-operative diagnosis with certainty, then the surgeon can offer the patient conservative surgery to preserve as much kidney as possible. However, in most of the cases, surgery precedes the diagnosis, and confirms diagnosis can only be made with certainty by histological examination of the respected specimen.

This article reports MCN to bring awareness about benign nature which carries an excellent prognosis.

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