International Journal of Research in Medical Sciences
Venkatesh K et al. Int J Res Med Sci. 2018 Sep;6(9):3175-3178
www.msjonline.org
pISSN 2320-6071 | eISSN 2320-6012

DOI: http://dx.doi.org/10.18203/2320-6012.ijrms20183665

Case Report

Multicystic nephroma: a rare benign renal tumor with diagnostic predicament

Kusuma Venkatesh[1]*, Savithri Ravindra[1], Ankita Agrawal[1], Ankit Malhotra[2]

[1]Department of Pathology, Kempegowda Institute of Medical sciences, Bangalore, Karnataka, India
[2]Department of Haematology, All India Institute of Medical Sciences, Delhi, India

Received: 23 June 2018
Accepted: 26 July 2018

***Correspondence:**
Dr. Kusuma Venkatesh,
E-mail: kusumav28@gmail.com

ABSTRACT

Multi cystic Nephroma is a rare benign tumor of kidney occurring in adults which has clinical, radiological and morphological features causing diagnostic dilemma as it mimicks other cystic renal lesions. Distinguishing it from a cystic renal carcinoma is very important. Multicystic nephroma is usually unilateral, more common in females, presenting as a well capsulated mass lesion with multiple non communicating cysts lined by hobnailing epithelium. A similar lesion occurring in children represents a well differentiated nephroblastoma. The case presented here was a female patient complaining of pain in the left flank and had a mass lesion which was diagnosed as cystic renal cell carcinoma radiologically. The nephrectomy specimen showed a multilocular cystic mass well delineated from adjacent renal parenchyma. Histopathologically the cysts were lined by hobnail type of epithelium and separated by fibrocollagenous stroma. The stroma had hyalinised areas, chronic inflammatory cell infiltration and foci of mature adipose tissue. No atypia or mitoses were seen in the epithelium or stromal cells. Based on the histological criteria a diagnosis of multicystic nephroma was made. It is important to make a diagnosis of multicystic nephroma based on histomorpholgical criteria as it relieves the patient from the burden of a malignant lesion.

Keywords: Cystic renal masses, Multicystic nephroma, Renal tumors

INTRODUCTION

Multilocular cystic nephroma (MCN) is an unilateral benign neoplasm of kidney. The first case was an unilateral described as cystic adenoma by Edmund et al, in 1892.[1,2]

It has a bimodal age distribution, common between 2-4 years with male predominance (73%) with a male to female ratio of 3:1. The 2nd peak occurs in adults in the age group of 40 to 60 years with female predominance with M:F=1:8.[1,3] MCN has a very good prognosis and over 200 cases have been reported in literature.[4]

Its histology and etiopathogenesis is controversial but according to WHO it is categorised under Mixed Epithelial Stromal Tumors (MEST), although Renal Epithelial Stromal Tumors (REST) can be used to encompass both MCN and MEST.[1]

The diagnosis of MCN clinically, radiologically using ultrasonogram (USG), Computed Tomography (CT), Magnetic Resonance Imaging techniques is difficult. Even on gross morphology it is indistinguishable from a cystic renal cell carcinoma. Only on microscopy it can be diagnosed after applying the histopathological criteria and hence it is necessary to be aware of the possibility of MCN as a differential as it has a benign course. The

MCN case presented here had all the classical histomorphological features.

CASE REPORT

A 39-year-old female presented to the department of urology with flank pain of two months duration. There was no history of hypertension or diabetes mellitus. On examination, there was a non tender organomegaly in the left lumbar region. Laboratory findings were normal, except for mild anemia with Haemoglobin of 11.6gm/dl. Results of urine examination were normal and urine sediment cytology was negative for malignant cells. Patient underwent abdominal ultrasound to investigate the causes of organomegaly. USG revealed a well defined round to oval mass lesion measuring 7.8×6.3×7.0cm involving mid portion and lower pole of left kidney with multiple thick internal septation and no internal vascularity or calcification. CT showed multiseptate cystic lesion in middle portion of the left kidney measuring 8.0×7.0cm with specks of calcification and peripheral enhancement on contrasts. However, no extension to renal pelvis and medulla was noted. The right kidney was normal and there was no other mass or enlarged lymphnodes. Based on clinical and radiological evaluation, a diagnosis of cystic renal cell carcinoma was made and left radical nephrectomy was performed.

Grossly the left nephrectomy specimen with perinephric fat measured 11.5×10.0×8.0cm. The kidney, measured 10.5×8.5×7.0cm with intact capsule and bosselations in the middle portion. Lower pole and posterior part of the kidney appeared normal. Cut surface showed a well circumscribed cystic lesion (Figure 1), measuring 7.4×6.5×5.0cm in the middle portion extending on to the upper and lower poles pushing the renal cortex. There were multiple cysts, exuding straw coloured fluid with solid grey white areas and yellow foci. The cystic structures were not connected with the pelvicalyceal system. The renal vessels, 2cm long ureteric stump and perinephric fat were unremarkable. No lymph nodes were found.

Figure 1: Gross specimen of kidney showing multiple cysts with interwening stroma.

Microscopically sections showed a neoplasm with thick fibrocollagenous capsule. Variably sized cysts were seen lined by flattened, cuboidal to columnar epithelium (Figure 2A) and showing hobnailing of the nuclei (Figure 2C, 3A and B).

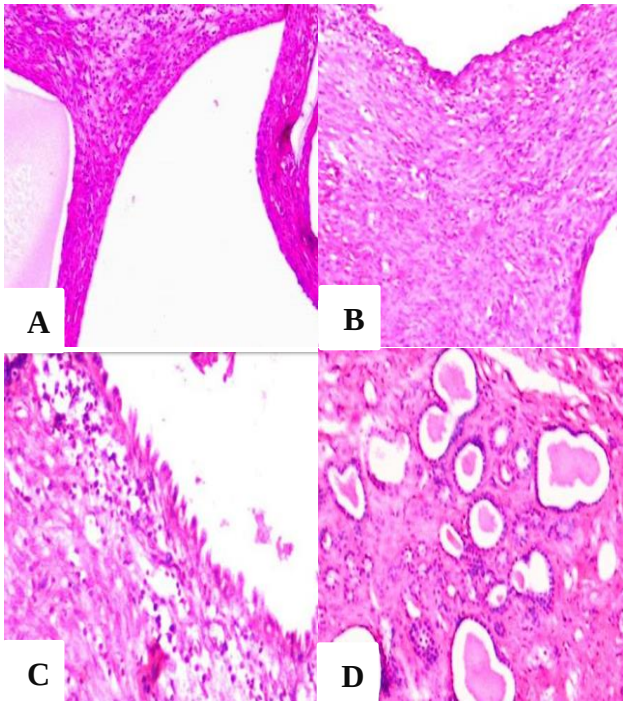

Figure 2: Microphotographs. A) Multiple cysts, with B) Abundant spindle celled stroma, C) Cyst wall lined by hobnail epithelium and D) Small cysts and tubules containing eosiophillic material. Haematoxylin and eosin stain. 40x.

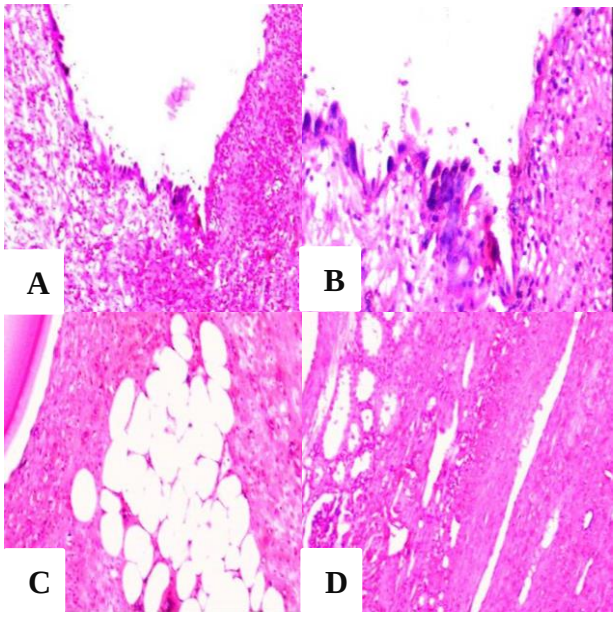

Figure 3: Microscopy A) Cyst wall, H and E 40x, B) Same cyst lined by hobnail epithelium. H and E 100x, C) Fat seen as a stromal component and D) Adjacent normal renal tissue. Haematoxylin & eosin stain. 40x.

Many cysts and small tubular structures seen were filled with eosinophilic secretions (Figure 2D). The stroma was fibrocollagenous having plump spindle cells with focal areas of hyalinization (Figure 2B), edema, haemorrhage, calcification and haemosiderin laden macrophages. The stroma had moderate chronic inflammatory cell infiltrate with variably sized blood vessels and some foci of mature adipose tissue (Figure 3C). There was no atypia or mitotic activity in the epithelium or stromal cells. Multiple bits were studied to look for blastemal cells and to rule out carcinoma. Renal parenchyma adjacent to the tumor (Figure 3D) and ureter were unremarkable. The above morphological features fulfilled the Joshi and Beckwith criteria based on which the final diagnosis of multicystic nephroma was made.[2] Patient is doing well on follow up of two and a half years post surgery.

DISCUSSION

MCN is a rare, benign, non heritable, unilateral tumor comprising 1-2 % of all renal tumors.[4] It has a bimodal age distribution, in males of less than 4 years age and in postmenopausal females. However, adult onset MCN and pediatric MCN are known to have different histomorphology.[5] MCN has many synonyms such as multilocular cystic renal tumor, cystic nephroma, multilocular benign cystic neoplasm, multilocular cyst and polycystic nephroblastoma.[3,6]

Etiology of MCN is controversial and multiple theories proposed include Mullerian origin because of ovarian like stroma and thought to be a developmental lesion having malignant potential.[1,2,4] Scanning electron microscopy shows epithelial cells with short microvilli and long cilia resembling collecting duct epithelium, suggesting the origin from collecting duct.[7] Origin from the ureteric bud has also been suggested.[4] Powel et al, in 1951 divided MCN into two entities namely, cystic nephroma based on the absence of blastemal elements and cystic partially differentiated nephroma (CPDN) which has blastemal or embryonal elements.[1,5] The symptoms are non specific like abdominal swelling or flank pain, presenting as urinary tract infection, rarely with hematuria and hypertension.[4] The tumor is also detected as an "incidentalloma" in an asymptomatic individual during routine clinical and imaging studies. Most common site of involvement is the lower pole but can involve any portion of the renal parenchyma.[1]

Radiographic features are non specific for MCN, both USG and CT show well circumscribed multicystic architecture with variable echogenecity and rarely calcification is noted as in this case and can be confused with malignancy. Guided needle aspiration assisted with colour Doppler can help in differentiating between benign and malignant lesion.[1,2]

The first pathological diagnostic criteria for MCN were given by Boggs and Kimmelstein and then modified by Joshi and Beckwith in 1989.[2] These criteria are: 1) entire lesion is composed of varying sized cysts divided by septae, 2) cystic mass is well demarcated from renal parenchyma, 3) only solid component is the fibrous septa, 4) cysts are lined by flattened, cuboidal or hobnail epithelium and 5) septae are made of fibrous tissue and may have well differentiated tubules. The case presented here had all these features.

Powel et al, gave 8 diagnostic criteria: unilateral involvement of the kidney, solitary lesion, multilocular lesion, cysts should be non-communicating with renal pelvis, non communicating with each other, cysts should be lined by epithelium, absence of normal renal parenchyma in the septae and residual renal tissue present adjacent to the lesion is normal. [8,9]

Differential diagnoses of MCN in pediatric age group are CPDN, cystic Wilm's tumor and in adults are partial polycystic kidney, multilocular cystic renal cell carcinoma (MCRCC), clear cell papillary RCC, tubulocystic carcinoma of kidney. CPDN and Wilm's tumor have blastemal elements which are not seen in MCN. In comparison to MCN, MEST has more complex epithelial and stromal elements with thicker septae. MCRCC will show CK, EMA, CD10, vimentin positivity and TCCK shows CD10, BHK, vimentin, AMACR positivity.[2]

There are no definite serological or immunohistochemical markers that can differentiate between MCN, MEST or renal carcinomas. Neoplastic epithelial cells of MCN show weak positivity for CK19 and stroma is ovarian like with ER and PR positivity.[6] Some genetic studies hypothesise that DICER1 mutation and hotspot missense mutation to be associated with MCN.[2,10] Although surgery remains the best treatment option, due to its benign nature partial nephrectomy or nephron sparing surgery can be done depending on the location and size of the MCN.[4]

CONCLUSION

MCN is a rare benign, non genetic neoplasm of kidney which should be differentiated from malignant tumours. Non specific clinical and radiological findings make it a diagnostic dilemma. Detailed histopathological study and using the morphologic criteria are essential to establish the diagnosis.

ACKNOWLEDGEMENTS

Authors would like to thank Dr. H. R. Madhusudan, Professor, Department of Urology, Kempegowda Institute of medical sciences, Bangalore, for providing the clinical details of the case.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Wilkinson C, Palit V, Bardapure M, Thomas J, Bowring AJ, Gill K, et al. Adult multilocular cystic nephroma: Report of six cases with clinical, radiolopathological correlation and review of literature. Urol Ann. 2013 Jan-March;5(1):13-7.
2. Ozturk H, Karaaslan S. Case report urethral invagination of multilocular cystic nephroma; a case report of a new pathologic variant. Int J Clin Exp Pathol. 2014;7(8):5271-79.
3. Deger AN, Capar E, Ucar BI, Deger H, Tayfur M. Adult multi cysticnephroma: Case report and review of the literature. J Clin Diag Res. 2015;9(8):22-3.
4. Atti LD. An unsual presentation of cystic nephroma in an adult man. Rare tumors. 2015;7:81-3.
5. Joshi VV, Beckwith JB. Multilocular cyst of the kidney (cystic nephroma) and cystic, partially differentiated nephroblastoma. Cancer. 1989;64:466-79.
6. Ivanova V, Dikov T, Alexandrov A. A case of multilocular cystic nephroma: clinical, histological and immunohistochemical aspects. Science and technologies. 2013;3(1):55-9.
7. Szymañska JD, Kaczmarek J, Papierz W. Cystic nephroma in adults. A report of two cases and review of the literature. Pol J Pathol. 2005;56(2):93-6.
8. Singh S, Chowdhury V, Dixit R, Manchanda A. Multilocular cystic nephroma of the kidney: A case report. Indian J Radiol Imaging. 2006;16:901-4.
9. Han KR, Janzen NK, McWhorter VC, Kim HL, Pantuck AJ, Zisman A, et al. Cystic renal cell carcinoma: biology and clinical behavior. InUrologic Oncology: Seminars and Original Investigations. Urol Oncol 2004;22(5):410-4.
10. Chang CP, Li JR, Yang CS, Ou YC, Cheng CL. Multilocular cystic nephroma: A case report and review of the literature. Urological Science. 2014 Dec 1;25(4):109-11.

Cite this article as: Venkatesh K, Ravindra S, Agrawal A, Malhotra A. Multicystic nephroma: a rare benign renal tumor with diagnostic predicament. Int J Res Med Sci 2018;6:3175-8.