CASE REPORT

Benign multilocular cyst or multilocular cystic nephroma in adults: three case reports

Suresh Bhat · Appu Thomas

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Abstract

Multilocular cystic nephroma is an unusual entity of uncertain etiology that typically arises from the renal parenchyma. We report three such cases in adult females, in their fourth and fifth decades. Since existing imaging modalities cannot definitely exclude the possibility of multilocular cystic renal cell carcinoma these patients should undergo surgery.

Keywords Cystic nephroma · Multilocular cyst · Cystic Wilm's tumour · Cystic Renal cell carcinoma · Renal cyst

Key Messages Multilocular Cystic nephroma in adults is difficult to diagnose preoperatively with CT scan and ultrasound. Most often patient has to undergo radical surgery as possibility of malignancy has to be ruled out. Histopathology examination makes the diagnosis absolutely certain.

Introduction

Multilocular cystic nephroma (MLCN), multilocular cystic renal cell carcinoma (MLCC), cystic renal cell carcinoma form a spectrum, with benign multilocular cyst at one extreme and cystic renal cell carcinoma at the other. Adult patients are eight times more likely to be female and most often present with flank mass, abdominal pain, or hematuria [1].

Case History

Case report 1

A 42-year-old lady presented with right flank pain and hematuria. She was hypertensive and a mass was found out in the right flank. Ultrasonogram revealed a cystic mass with interspersed echogenic areas. CT revealed a multiloculated mass with moderate enhancement of septa well delineated from normal renal parenchyma. Fine needle aspiration cytology (FNAC) showed no evidence of malignancy.

Case report 2

36-year-old female presenting with a right flank mass. A cystic mass was detected while she was pregnant on an antenatal ultrasound 6 years back in 1998. The delivery was uneventful. Postpartum ultrasound evaluation revealed multiple cysts in right kidney. CT abdomen was done at this juncture and a multiloculated mass was found. (Fig. 1a) She was advised surgery but refused. Although she remained asymptomatic for next five years, in year 2004 presented with a mass in the flank of increasing size. On physical examination, she was normotensive and had a non-tender mass in the right lumbar region that was smooth

S. Bhat¹ (✉) · A. Thomas²

¹Department of Urology
Medical College,
Kottayam

²Prof. & Head, Dept. of Urology
AIMS,
Cochin

E-mail: drsureshbhat@yahoo.com

surfaced, mobile and firm. Ultrasound examination revealed a large cystic mass with little echogenic areas in between. CT scan also revealed a huge mass occupying more than half of the abdomen with thin septa and moderate enhancement. (Fig. 1b) CT guided FNAC revealed hemorrhagic fluid and no malignant cells.

Case report 3

A 42-year-old lady presented with a dull ache and a mass in the left flank. She had a cystic mass with multiple loculations and interspersed echogenic areas. CT scan showed

Fig. 1a Contrast enhanced CT picture showing multiloculated mass with septal enhancement with a thin rim of renal parenchyma.

Fig. 1b Contrast enhanced CT of the patient showing increased size of the mass and absence of normal parenchyma, 6 years after initial picture.

Fig. 2 Multiloculated cysts of varying size with moderate septal enhancement separate from renal parenchyma.

multiloculated lesion with septal enhancement well delineated from renal tissue. (Fig. 2) The renal parenchyma was seen separate from the lesion.

In the first and third case radical nephrectomy was done, while in the other case simple nephrectomy was performed. All cases histopathology were reported as multilocular cystic nephroma.

Discussion

Typical multilocular cystic nephroma consist of numerous indistinct cystic areas located within a well-defined encapsulated mass [2]. This lesion is composed of several small cystic spaces that do not communicate with one another or with the renal pelvis. The cystic spaces are lined by cuboid or flattened epithelial cells and separated by fibrous septations, which constitute the only solid component of the lesion. The lesion does not contain mature renal parenchyma. A thick fibrous capsule surrounds the entire lesion [3]. The existence of degenerated multilocular cystic nephroma (DMLCN) remains controversial. It would correspond to a MLCN where the epithelial component would focally degenerate into a clear cell or papillary carcinoma [4]. This lesion was defined as a MLCN with 1 or 2 foci of malignant cells, less than 5 mm in diameter, and located within the wall of the lesion or within a septation. Multilocular cystic renal cell carcinoma (MLCC) has a pseudocapsule and contains several cystic spaces of variable size that are filled with fluid, blood, or necrotic material. Microscopically, malignant cells lining the cystic spaces are typically clear cells. The tissue between the cystic spaces often is hyalinized and also contains clear cells.

Ultrasonography usually shows multiple cystic lesions with intervening echogenic septae. Septations may be thick but regular and moderate contrast enhancement is seen on CT. Ultrasonogram and CT is not sufficiently reliable to distinguish cystic nephroma from Cystic renal cell carcinoma and multilocular cystic renal cell carcinoma. The presence

of thin calcifications along septations and protrusion of the tumor into the renal pelvis have been described as characteristic findings of MLCN. Some authors have reported that MLCN is almost always a well-defined lesion with thin and regular septations whereas MLCC always contains a solid component showing significant enhancement.

Recent reports speculate that magnetic resonance imaging (MRI) of multilocular cystic nephromas will produce imaging features that are highly suggestive, but not always pathognomonic [5]. The characteristic findings include a complex cystic mass with enhancing septae and herniation into the renal collecting system. Curvilinear calcification and herniation into the renal pelvis are highly suggestive of MLCN [5]. In cases with more complex image presentations, fine needle puncture is recommended, preferably in different areas of the mass in order to rule out the presence of malignant cells. Treatment is nephrectomy, but partial nephrectomy can be done in small tumours. Though cystic nephroma exhibits benign behaviour, local recurrence reflects inadequate excision of initial lesion. Since epithelial cells of the cysts resemble renal collecting tubular cells which favours a congenital malformation from segmental maldevelopment of ureteric bud.

Conclusion

The above three cases share some common features like age group, sex, long duration of symptoms, non-specific dull flank pain, slow growth of the mass, good performance status, definite demarcation from normal parenchyma, moderate enhancement of septae and large size of the mass. We found that FNAC was not very useful. We feel that clinical finding along with USG and CT may help in making

a presumptive preoperative diagnosis, but only histopathological examination make the diagnosis absolutely certain. Inability to obtain a preoperative diagnosis prevented us from proceeding with a nephron sparing surgery in two of our cases. In our opinion intra-operative frozen section study may not be useful in such large tumours because of the risk of harbouring RCC inside the specimen. We also would refrain from advising observation because of the risk of rapid growth, rupture and a rare chance of development of malignancy. Further experience in MRI may help us to distinguish between a benign and a malignant multilocular cyst.

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