

Multicystic nephroma in an elderly man. Case report

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SUMMARY: Multicystic nephroma in an elderly man. Case report.

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Multicystic nephroma is a relatively rare tumor of the kidney presenting unclear histological origin. Abdominal mass is a common onset sign in children while abdominal flank pain or abdominal discomfort, hematuria and recurrent urinary tract infections usually affect adults. Preoperative diagnosis is impossible especially in the adult variant of the tumor where clear cells carcinoma with cystic degeneration must always be suspected.

We herein report a case of a 77 year old man complaining of flank abdominal pain and recurrent episodes of urinary tract infection due to a right-sided multicystic nephroma successfully treated with nephrectomy.

RIASSUNTO: Nefroma multicistico in un paziente anziano. Caso clinico.

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Il nefroma multicistico è un tumore raro del rene di origine istologica non chiara. Si presenta di solito come massa addominale nei bambini, mentre dolore addominale localizzato o diffuso, ematuria e infezioni urinarie ricorrenti sono la presentazione più comune negli adulti. La diagnosi preoperatoria è impossibile, particolarmente negli adulti nei quali il carcinoma a cellule chiare deve comunque essere sospettato.

Riportiamo il caso di un anziano di 77 anni con dolore al fianco e ricorrenti infezioni urinarie da nefroma multicistico del rene destro trattato con successo con nefrectomia.

KEY WORDS: Multicystic nephroma - Adult - Surgery.
Nefroma multicistico - Adulto - Chirurgia.

Introduction

Multicystic nephroma (MCN) is a rare, benign lesion of the kidney. This tumor has been described in the past with a variety of names such as multilocular cystic tumor, renal multilocular cyst, multilocular cystic nephroma, renal cystadenoma and partial polycystic kidney (1,2). Recently, the World Health Organization (WHO) classified MCN as a special entity in the context of mixed epithelial and stromal tumors (MESTK) (3). The disease usually occurs in childhood, although a second peak of incidence concerns middle aged patients (3). Abdominal mass is the main sign of presentation in children while

abdominal fullness or pain, recurrent urinary tract infections or hematuria is usually the main symptoms in adults (4).

We herein report a case of a 77 year old man presented with flank abdominal pain and recurrent episodes of urinary tract infection attributed, after an extensive investigation, to a right-sided multicystic nephroma successfully treated with nephrectomy.

Case report

A 77-year old man came to the outpatient facilities complaining of mild persistent right flank pain. Patient's medical history included hypertension, atrial fibrillation (under treatment with digoxin, acenocumarol and ramipril) and appendectomy 35 years ago. The patient also refers two recent episodes of urinary tract infection due to *Escherichia coli* successfully treated with ciprofloxacin.

Physical examination was unremarkable. Laboratory findings were within normal limits, excepting for microscopic haematuria, urinary white blood cells and nitroses resulted from urine analysis. He underwent abdominal ultrasound (US) in order to investigate possible cause of recurrence of the urinary infection such as nephrolithiasis

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or prostatic lesions. The results of the US surprised us, demonstrating a multilocular cystic mass of the right kidney with thick, avascular septa. No dilatation of the homolateral pelvicalyceal system was observed. Computed tomography (CT) of the abdomen showed a multilocular cystic lesion of 6,5 cm in diameter, located at the lower pole of the right kidney (Fig. 1). The intravenous administration of the contrast demonstrated only an intermediate enhancement. There were not additional cystic lesions in the remaining renal parenchyma. Angiography, urography and fine-needle aspiration biopsy were not performed, according to the current literature. The magnetic resonance imaging (MRI) described a multicystic mass of the middle and lower pole of the right kidney with a peripheral enhancement after the administration of intravenous paramagnetic contrast and suggested a clear cell type mass (Figs. 2 and 3).

Coagulation parameters were adjusted and a radical nephrectomy was performed, without intra- and post operative complications. The patient was discharged 7 days after the admission to the hospital. The pathology report described multilocular cystic nephroma. Gross examination revealed a 7×5×4,5cm multilocular cystic lesion of the lower pole of the right kidney. Cysts were filled with serum or gelatinous-like content. Microscopically the cysts were lined by a single layer flattened, cuboidal or cylindric epithelium and were separated by a fibroblastic stroma of variable cellularity (Figs. 4, 5, 6 and 7). Signs of pelvicalyceal inflammation were also present. The immunohistochemical examination revealed a positive stain for CK7, CK19, Vimentin, 34BE12 and a negative stain for CK20, CEA, and RCC. Positivity of CK19 and 34BE12 suggested an aberrant renal-tubular differentiation. The remaining renal parenchyma was normal.

During 24-months of follow up, no signs of recurrence were observed.

Discussion

Cystic nephroma is a benign renal neoplasm. It was first described in 1892 by Edmunds as a cystic adenoma of the kidney (5). The neoplasm may be congenital as well as acquired (6). Rarity and lack of registration do not permit a valid estimation of incidence age and sex distribution although, Madewell et al. in a retrospective analysis reported considerable incidence in patients older than 30 years old (40%) (7). The adult variant of MCN usually affects women, with a male to female ratio of 1:9, while the same ratio of MCN in the childhood is 3:6 (2, 8).

Many theories have been proposed in order to investigate the pathogenesis of this tumor and the possible correlation with the Wilms' tumor. Powel et al. (9) established the criteria for the diagnosis of MCN in 1951 and considered the tumor as a developmental malformation. Steele et al. (10) observed similarities of the tumors' stroma with the ovarian stroma, attributing the tumor's origin to a possible entrapment of Müllerian type tissue into the kidney. Some authors described ciliar and microvillar appearance of the epithelium in electron microscopy and linked this fact with similar aspects of the collecting tubules (11). Joshi and Beckwith proposed a connection between MCN, nephroblastoma and cystic partially differentiated nephroblastoma; however, they

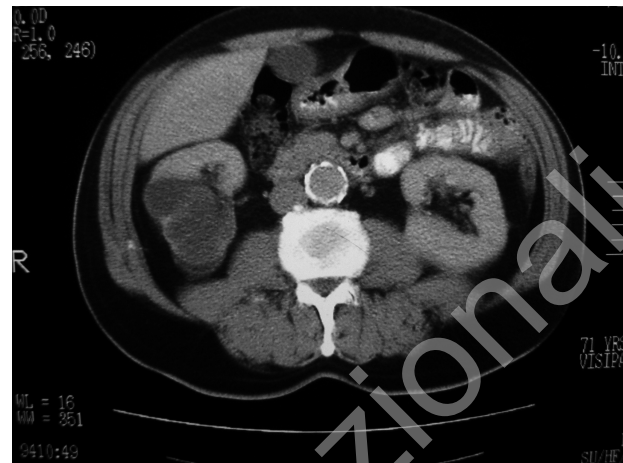


Fig. 1 - Axial abdominal CT image without iv contrast administration, demonstrating a multiloculated cystic mass, with internal septa, originating from the lower pole of the right kidney.

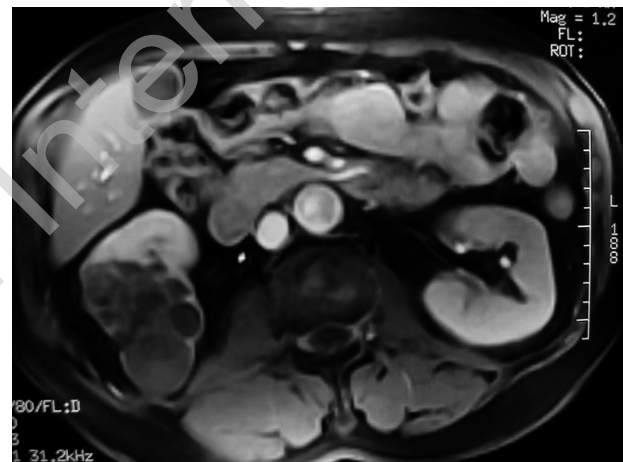


Fig. 2 - MRI Axial T1-weighted post-gad image with fat suppression demonstrating enhancement of septa.

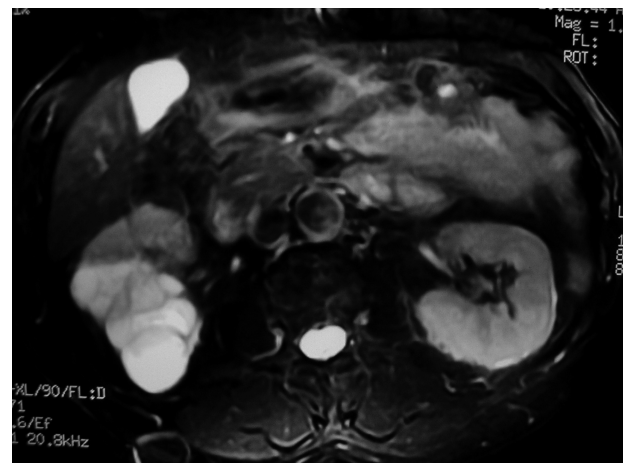


Fig. 3 - MRI Axial T2-weighted post-gad image with fat suppression demonstrating a high signal intensity (cystic) multiloculated mass with capsule and septa.



Fig. 4 - Histology. Low-power microscopic appearance of the tumor showing multiple cysts lined by single-layered epithelium (H&E; x10).

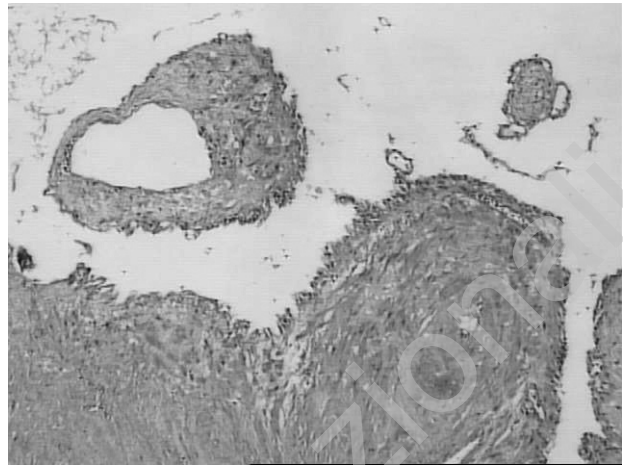


Fig. 6 - See figure 7 legend.

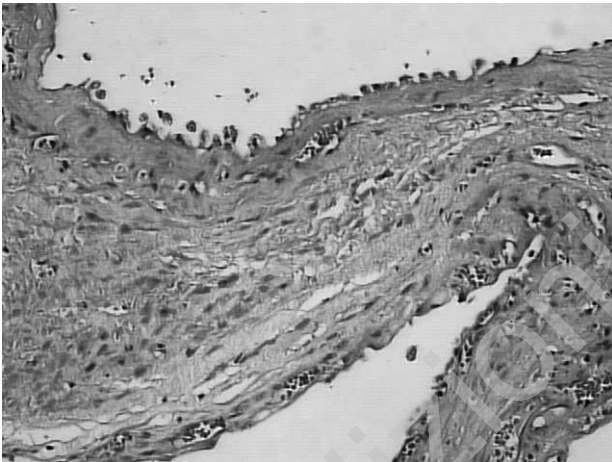
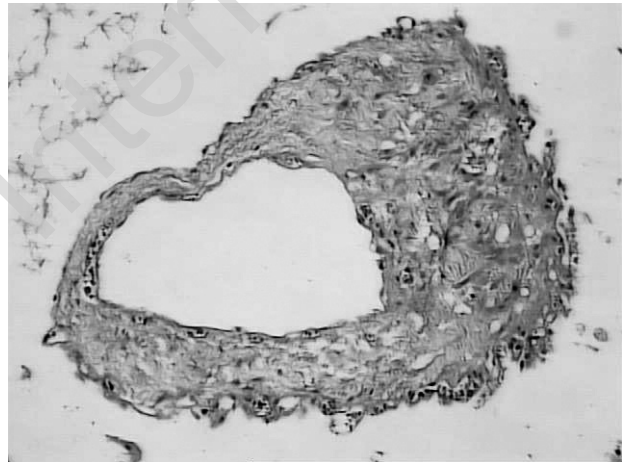


Fig. 5 - Histology. Cystic spaces lined by hobnail epithelial cells (H&E, x 20).



Figs. 6 and 7 - Histology. The cystic lesions were lined by flattened epithelium (H&E, x10 and x20, respectively).

emphasized the absence of immature tissue in the histological appearance of the tumor (12). A hormonal hypothesis has been proposed in order to justify the fact that MCN affects almost exclusively women. This hypothesis is also enforced by the common positivity in estrogens and progesterone receptors in the stroma of these tumors. Although, in many cases stroma results negative for estrogen and progesterone receptor expression indicating an unclear hormonal mechanism of origin (13).

According to Eble and Bonshib, two distinct variants of MCN exist, the children and the adult variants. The children variant should be considered a Wilm's-related tumor, like the partially differentiated nephroblastoma, and should not be connected with the presence or the absence of Wilm's tumor elements. On the other hand, the adult variant should not be related with

the Wilm's tumor or nephrogenic remnants and should be considered as a separate entity. In addition, the Authors modified the histological criteria proposed by Powel (and later modified by Joshi and Beckwith) and included in the histological description the following: 1) an expansive mass with fibrous capsule; 2) interior composed by cysts and diaphragms; 3) absence of nodularity; 4) cystic wall lined by flattened and cuboidal epithelium; 5) septa containing epithelial structures, similar to mature renal tubules (2). The WHO classifies MCN as or similar to mixed epithelial stromal tumors (MESTK), especially when ovarian type stroma or stroma with white or fibrous elements is present (3). Recently, some authors proposed the unifying term of 'renal epithelial and stromal tumors' (REST) in order to encompass the spectrum of findings observed in these tumors (13).

MCN is usually unilateral. In children, a painless abdominal mass is the most frequent onset sign. In adults, abdominal discomfort or pain, hematuria and urinary tract infection are the main symptoms (4). Routine investigations for other reasons often reveal the tumor, whose the distinction from malignant cystic neoplasms is difficult. Imaging studies such as US and CT usually demonstrate the multilocular character of the neoplasm; however, distinction between class II and III cyst, following the Bosniak classification, is often difficult (4,7). Echocolor-Doppler seems to be helpful, demonstrating MCN as an hypovascular or avascular cystic renal lesion. Renal angiography is also not specific (4). Fine-needle aspiration biopsy (FNAB) combined with image study seems to be useful in children, however, its value is limited in adults (15).

Surgical treatment with a radical nephrectomy seems to be both diagnostic and curative in order to exclude a possible malignant degeneration (16,17) Partial-sparing nephrectomy may be an alternative for a rare bilateral renal involvement (18,19).

Conclusions

Multicystic nephroma is a rare benign renal tumor, whose preoperative differential diagnosis is impossible. Surgical intervention is diagnostic and curative. The limited number of cases described, the uncertain oncologic behavior and the lack of clear recommendations for surveillance make a close post-operative follow-up mandatory.

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