Clinical History:

A 54 year-old asymptomatic woman, was referred to ultrasound examination due to the presence of microscopic hematuria discovered in a routine appointment. There were no other relevant clinical aspects.

Imaging Findings:

Ultrasound showed a large cystic mass with various septa in the left kidney, with predominant anechoic areas, measuring 66 mm in maximum diameter (fig.1a). There wasn't evident solid areas, or vascularization at Doppler interrogation (fig.1b).

CT confirmed the presence of a multiloculated cystic mass, in the middle third of the left kidney, some cysts presented hyperdense material suggesting hemorrhagic content (fig.2). After contrast administration the mass didn't show significant enhancement of the septa or the presence of solid areas (fig.3). Furthermore the lesion appears to herniate to the renal pelvis (fig.4).

This complex cystic lesion was referred as a Bosniak type 3 lesion, so the patient performed radical nephrectomy.

Pathological analysis revealed cystic multiloculated tumor, with several cysts of variable dimension. The interior of the cysts showed smooth inner liming, the majority contained serous material, while some contained hemorrhagic content. Histological findings were consistent with multilocular cystic nephroma (MCN).

Discussion:

The pathogenesis of MCN is controversial and the classification is unclear. Some proposed etiology as a developmental defect, while some postulated it could be neoplastic in origin [1].

First described as cystic adenoma in 1892[2], the name was later changed to MCN in 1956 [3]. In 1989 authors [4] proposed a scheme in order to differentiate MCN from cystic partially differentiated nephroblastoma (CPDN): while they both are anatomically and radiologically indistinguishable, the later present embryonic elements in septa.

The Bosniak classification [5] is used for CT evaluation of renal cystic lesions, including 5 categories: 1 and 2 are benign, without follow-up; 2F are minimally complex lesions requiring follow-up; Type 3 lesions are indeterminate
lesions possessing a malignant potential with surgical indication; Type 4 lesions are clearly malignant lesions requiring surgical treatment.

MCN presents a bimodal age distribution: two-thirds occur under the age of two, mostly in boys; the remaining occur mainly in females between the 5th-6th decades [6].

Symptoms are nonspecific; they consist of abdominal pain and hematuria, sometimes with urinary tract infection depending on the obstruction of the calyx due to the cystic volume [7]. The hematuria can result from the infection but also from cysts herniation into the renal pelvis [8].

MCN isn’t considered pre-malignant, however there are reports of co-existing foci of renal cell carcinoma in the lining of the cysts. In adults MCN is considered benign, but recurrences have been reported, and it is unclear if they are related to missed malignant foci or sarcomatous degeneration [1].

Sonographically MCN appears as multiple anechoic spaces separated by thin ecogenic septa, with no solid elements. The presence of multiple cysts can difficult the exam due to several acoustic interfaces. Color-Doppler imaging is suggested as a useful tool for the differential of malignant versus benign lesion [9].

At CT they present as multilocular cystic lesion. When the cysts are small they can be confusing for solid elements due to septa proximity. After contrast they don’t enhance significantly. The cysts don’t communicate with the excretory system and they characteristically herniate to the renal pelvis [1].

MRI is rarely needed; its features include hypo-intense signal of septa in all sequences, and variable cyst signal accordingly to its content.

MCN should be considered in the differential of malignant cystic renal tumors in children and adults. Some features can suggest MCN, however definitive diagnosis should only be made after surgical treatment. **Differential Diagnosis List:** Adult multilocular cystic nephroma, cystic partially differentiated nephroblastoma, cystic wilms tumour

**Final Diagnosis:** Adult multilocular cystic nephroma

**References:**


Figure 1

a

**Description:** Longitudinal ultrasound of the lesion in the left kidney, demonstrating a cystic multiseptated mass, with posterior acoustic enhancement, thin septa and no evident solid areas. **Origin:** Serviço de Imagiology - CHTV Viseu

b

**Description:** Color Doppler interrogation demonstrating no evident vascularization in the lesion. **Origin:** Serviço de imagiology - CHTV Viseu
Figure 2

Description: Multiloculated cystic formation in the middle third of the left kidney, with some discrete hyperdense focus. Origin: Serviço de Imagiology - CHTV Viseu
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Figure 3

Description: Corticomedullary phase, demonstrating the lesion without significant contrast enhancement. Origin: Serviço de Imagiology - CHTV Viseu
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Description: Corticomedullary phase, demonstrating the lesion without significant contrast enhancement. Origin: Serviço de Imagiologia - CHTV Viseu
Description: Nephrographic phase. Origin: Serviço de Imagiology - CHTV Viseu
Description: Nephrographic phase, demonstrating the herniation of the lesion into the renal pelvis.
Origin: Serviço de Imagiology - CHTV Viseu
Description: Delayed phase. Origin: Serviço de Imagiology - CHTV Viseu
Description: Coronal reformat demonstrating the multilocular cystic lesion. Origin: Serviço de Imagiology - CHTV Viseu
**Description:** Coronal reformatted image in the nephrographic phase. **Origin:** Serviço de Imagiology - CHTV Viseu
Description: Coronal reformatted image in the nephrographic phase. Origin: Serviço de Imagiology - CHTV Viseu
**Description:** Coronal reformatted image illustrating the herniation of the lesion into the renal pelvis.

**Origin:** Serviço de Imagiology - CHTV Viseu
Figure 5

Description: Glass slides of the removed lesion Origin: Anatomopathology department, Centro Hospitalar de Coimbra (Image courtesy Dra. Raquel Pina)
**Description:** Microscopic appearance of the specimen: cysts with flattened epithelial lining, with septums of dense stroma cells without clear cells, thus allowing its distinction from the cystic clear cell renal carcinoma. **Origin:** Anatomopathology department, Centro Hospitalar de Coimbra (Image courtesy Dra. Raquel Pina)
Description: Microscopic appearance of the specimen: cysts with flattened epithelial lining, with septums of dense stroma cells without clear cells, thus allowing its distinction from the cystic clear cell renal carcinoma Origin: Anatomopathology department, Centro Hospitalar de Coimbra (Image courtesy Dra. Raquel Pina)
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