Case report

Multilocular cystic nephroma: A case report and review of the literature

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Abstract

Multilocular cystic nephroma is a rare cystic renal tumor, which is benign and has an excellent prognosis. However, preoperative diagnosis is challenging and is made exclusively by pathological findings. We reported a 41-year-old woman with chronic flank pain, and abdominal computed tomography revealed a multiloculated renal cystic tumor. The possibility of a cystic variant of renal cell carcinoma could not be excluded. Laparoscopic radical nephrectomy was performed, and diagnosis of multilocular cystic nephroma was made. In this report, we describe the clinical presentation, radiological finding, and histopathology of this case.

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1. Introduction

Multilocular cystic nephroma is a rare entity of benign multicystic renal tumor. It was first described in 1892, and only approximately 200 cases have so far been reported in the related literature. The diagnosis is made based exclusively on the pathological finding. Histological features include a flat, cuboidal epithelium and an ovarian-like stroma. We present a case of multilocular cystic nephroma, and describe the clinical presentation as well as the radiological, histopathological, and immunohistochemical findings; a review of the literature is also described.

2. Case report

A 41-year-old woman presented with left flank pain for 3 months. She denied fever, gross hematuria, or body weight loss. Ultrasonography revealed a multicystic cystic mass with multiple thick septa (Fig. 1A). Abdominal computed tomography showed a multiloculated renal cystic mass with contrast-enhancement septa, which was consistent with Category III of the Bosniak classification (Fig. 1B). Multicystic renal cell carcinoma was suspected and laparoscopic left radical nephrectomy was performed. The resected left kidney weighed 300 g, measuring 13 cm × 7.5 cm × 7.5 cm without a solid component. A well-capsulated multicystic cyst mass was noted in the upper pole of the left kidney. This cystic mass was noncommunicating and composed of a clear fluid. Microscopically, the cystic wall had a flat, hobnail, cuboidal epithelium lining. The septum consisted of ovarian-like fibroblastic stroma (Fig. 2). Immunohistochemistry stains were positive for cytokeratin (CK) 7, CK 19, CK AE1/AE3, vimentin, estrogen receptor (ER), and progesterone receptor (PR) (Fig. 3). The final diagnosis of multilocular cystic nephroma was made. The patient underwent an uneventful recovery.

3. Discussion

Multilocular cystic nephroma is a rare and benign renal neoplasm with an excellent prognosis. It has a bimodal distribution in children within 2 years of age with a male predominance and in adults over 30 years of age with a female predominance. Most cases are asymptomatic and found incidentally during imaging investigation. Clinical presentations include flank pain, gross hematuria, abdominal mass, and urinary tract infection. The etiology and pathogenesis remain unknown. Associations between genetic abnormalities, hormones, and oncogenetic factors require further investigation. Bahubeshi et al reported that mutation of germline DICER1 is associated with familial cystic nephroma.

On the image study, a multilocular, cystic nephroma appears as a multilocular cyst with multiple septa, using contrast enhancement, and watery fluid inside the cyst. Sometimes, extension into the renal pelvis may be seen, which results in hydronephrosis and hemorrhage. Cystic renal neoplasms comprise both benign and malignant phenotypes. The differential diagnosis should include multilocular cystic nephroma, multicystic renal cell carcinoma, multicystic dysplastic kidney, medullary sponge kidney, tubulocystic carcinoma,
and mixed epithelial and stromal tumor. The Bosniak classification for computed tomography is helpful in determining the risk of malignancy. However, multilocular cystic nephroma is usually assigned to Category III and above, and the malignant potential is greater than 54%. Shannon et al. reported that among 235 biopsies for less than 5 cm incidental renal masses, 184 (78%) were diagnostic and 51 (22%) were nondiagnostic due to insufficient material or only normal, inflammatory, fibrotic, or necrotic tissue contents. Diagnostic biopsies

Fig. 1. (A) Renal ultrasonography reveals a multilocular cystic lesion with septa in the left kidney. (B) Computed tomography of the kidney reveals a well-defined multilocular cystic mass measuring 6.5 cm × 5 cm in size with contrast-enhanced septa in the left kidney.

Fig. 2. (A) Gross appearance of the excised kidney, which measures 13 cm × 7.5 cm × 7.5 cm and has a honeycomb-like cyst. (B) Renal cyst lined by a single layer of flattened cuboidal epithelium (hematoxylin and eosin (H&E) stain, original magnification 10×). (C) Cuboidal, hobnail epithelium of the cystic wall, and fibrous stroma resembling ovarian stroma (H&E stain, original magnification 100×).

Fig. 3. Immunohistochemical stains (original magnification 40×). Positive expression for (A) cytokeratin 7, (B) vimentin, (C) estrogen receptor, and (D) progesterone receptor.
revealed 138 malignant (75%) and 46 benign (25%) lesions. For a small renal mass (<4 cm), preoperative renal biopsy can be considered and has the potential to avoid a significant number of major surgical procedures. In this case, the renal cyst measured 7.5 cm and was multilocular and noncommunicating; these properties of the cystic mass make it difficult to obtain adequate tissue and pose a danger of malignant cells seeding. Therefore, we did not perform preoperative renal biopsy.

Histopathological examination from a resected specimen seems to be the only feasible method of making the correct diagnosis. The diagnostic criteria of cystic nephroma established by Joshi and Beckwith et al. are used widely and include the following: multilocular, solitary, unilateral, noncommunication between the renal pelvis and the cystic lesion, a definite lining of epithelium on the loculi, no nephron in the interlobular septa, and normal residual renal tissue. Our case meets these criteria. Although the use of immunohistochemistry in diagnosis has not been studied well, it may provide some useful information. In our case, it showed positive expressions for CK 7, CK 19, CK AE1/AE3, vimentin, ER, and PR. A positive expression of CK suggests aberrant renal tubular differentiation. The ovarian-like stroma expresses ER/PR.8

Because preoperative diagnosis is difficult to achieve and multicystic renal cell carcinoma is suspected, radical nephrectomy is the standard treatment historically. However, nephron-sparing surgery should be borne in mind if the mass is smaller than 4 cm, unilateral, solitary, and localized, or when multicystic renal cell carcinoma is suspected, radical nephrectomy should be borne in mind if the mass is smaller than 4 cm, is the standard treatment historically. However, nephron-sparing surgery is considered and has the potential to avoid a significant number of major surgical procedures. In this case, the renal cyst measured 7.5 cm and was multilocular and noncommunicating; these properties of the cystic mass make it difficult to obtain adequate tissue and pose a danger of malignant cells seeding. Therefore, we did not perform preoperative renal biopsy.

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Conflicts of interest

The authors declare that they have no financial or non-financial conflicts of interest related to the subject matter or materials discussed in the manuscript.

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References