Cystic Nephroma with Sarcomatous Transformation: a case report

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Cystic nephroma is a rare renal neoplasm which is usually benign histologically. We report a case of a 42-year-old woman with cystic nephroma arising from the lower pole of the right kidney with microscopic foci of sarcomatous stroma. Preoperative sonogram and CT scan were performed. Pathologic study revealed a cystic nephroma with foci of primitive, mesenchymal, undifferentiated, embryonal type sarcoma.

Key words: Kidney; Computed tomography (CT); Kidney neoplasms; Sarcoma; Ultrasound (US)

Cystic nephroma and cystic partially differentiated nephroblastoma (CPDN) are two histologically distinct but grossly indistinguishable lesions encompassed by the term multilocular cystic renal tumor [1, 2]. They are characterized by well-circumscribed, encapsulated masses that contain multiple, noncommunicating, fluid-filled locules [2-4]. The two lesions are only differentiated on histologic grounds by the presence (CPDN) or absence (cystic nephroma) of blastemal elements in the cyst septa [2, 5, 6]. Some of the names that have been applied to the multilocular cystic renal tumor are listed below: multilocular cystic nephroma, multilocular renal cyst, benign cystic nephroma, cystic adenoma, cystic harmatoma, cystic lymphangioma, cystic Wilms tumor, segmental polycystic kidney, segmental multicystic kidney and Perlmann's tumor [2-5, 7]. Although these lesions are usually benign histologically, cases have been reported containing foci of sarcoma or nephroblastoma [3].

CASE REPORT

A 42-year-old woman presented with a palpable mass over the right flank region one week prior to admission. Five days later, she began to suffer from right flank pain and gross hematuria. She had been well without history of abdominal trauma or familial renal disorder. Physical examination revealed a big tender mass over the right flank region. A routine blood examination showed mild anemia with a hemoglobin level of 11.2 mg/dl. The urine examination showed 4-6 red blood cells and 20-25 white blood cells per high power field. Renal function and blood pressure were normal. The sonogram of the abdomen disclosed a multiseptated cystic mass arising from the lower pole and interpolar region of the right kidney (Fig. 1). No concomitant cysts were found in the left kidney, liver and pancreas. Some of the septa were thin and some were thick and irregular. A precontrast CT scan revealed a well-circumscribed, low-attenuating mass measuring about 12 × 8 × 6 cm in size, which is divided by numerous soft-tissue strands but without a solid nodular component.

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Following contrast administration, the septa showed progressive enhancement, but no contrast accumulated within the locules (Fig. 2b). The normal renal parenchyma was splayed, and the right ureter and inferior vena cava were compressed by the tumor. The mass was placed in category III according to the Bosniak classification, as it was a complicated renal cystic tumor with some thick and irregular septa without a solid component on imaging studies. The preoperative impression was cystic renal cell carcinoma, and surgical exploration was performed.

The tumor was resected with right radical nephrectomy, the well-encapsulated tumor being discovered over the lower pole of the right kidney. When bivalved, the specimen revealed multiple cysts of variable size, some of which were filled with blood-tinged fluid. Under microscopy, the tumor showed multiple cystic spaces with cuboid, columnar or hobnail epithelium lining the locules (Fig. 3). Also of note were foci of primitive mesenchymal undifferentiated embryonal type sarcoma within the septal stroma. Between the cysts and underneath the capsule were tiny renal tubules and rare immature glomeruli. The pathologic diagnosis was cystic nephroma on the right kidney with evidence of undifferentiated embryonal spindle cell sarcoma.

Postoperatively, the patient recovered well and was discharged two weeks later. A follow-up CT scan 3 months postoperatively revealed no recurrent tumor.

**DISCUSSION**

Cystic nephroma is generally regarded as a non-familial disease and is not associated with cystic changes in other organs [2]. The affected male patients are usually younger than 4 years of age and female patients typically present with symptoms between 4 and 20 years of age or after 40 years of age [3]. The tumor is seen on either side of the kidney with approximately equal distribution and predominance in the lower pole [3, 7]. The common clinical signs and symptoms include flank pain, palpable mass, hematuria, and urinary tract infection [2, 3, 5, 8, 9].

Grossly, cystic nephroma is a solitary, well-circumscribed lesion containing multiple cysts of variable sizes and is surrounded by a thick, fibrous capsule that often compresses adjacent renal parenchyma and may protrude into the renal pelvis or

**Figure 1.** Sagittal sonogram of the right kidney demonstrates a complex mass (arrows) with multiple cysts and irregular septa. The asterisks '+' and 'x' outline the margin of the right kidney.

**Figure 2.** a. Precontrast axial CT scan shows an encapsulated mass with internal soft-tissue strands. b. Contrast-enhanced CT image in excretory phase shows enhancement of the septa. The crescent-shaped normal parenchyma (white arrows) around the periphery of the tumor confirms its renal origin. Also note the compressed ureter (arrowhead) and inferior vena cava (black arrow).
bulge from the convexity of the renal cortex [1, 3, 9]. The cyst contents are usually clear to yellow, and hemorrhage is uncommon [2, 3, 9]. The size of the cysts may vary from microscopic to several centimeters in diameter [4, 9]. The cysts are lined by flattened, cuboidal or hobnail epithelium and the septa may consist of fibrous tissue that varies from myxoid to collagenous but are devoid of normal renal parenchyma [2-4, 9]. Most cystic nephromas are benign histopathologically [5]. However, a few sarcomas such as undifferentiated embryonal spindle cell sarcoma, low-grade leiomyosarcoma, and pleomorphic high-grade sarcoma arising from cystic nephroma have been reported in the literature [9].

The multiple cystic spaces are best demonstrated using ultrasound. The echogenic septa dividing the cysts are usually thin and smooth with occasional curvilinear calcifications [2, 5]. However, they may be thick or irregular, which may raise the possibility of the presence of malignant stroma in the Bosniak classification [10]. The typical CT findings include a sharply-circumscribed, multiseptated renal mass with the attenuation value slightly exceeding that of water [2]. The size of a cystic nephroma is usually large, averaging about 10 cm in diameter [5]. The septa enhance following administration of contrast material but contrast material does not accumulate within the individual locule [2].

The CT imaging features of cystic nephroma can not be differentiated from other cystic neoplasms such as cystic partially differentiated nephroblastoma, cystic renal cell carcinoma, and cystic Wilms tumor on imaging or clinical findings, and surgery is required for diagnosis [2, 5, 7, 11]. Frozen sections can be obtained during the operation to decide whether nephron-sparing surgery is possible [1, 5, 15]. Cystic nephroma without a malignant element is curative with surgical excision [2, 9]. Distal metastases with poor prognosis have been reported in cases with a sarcomatous component [3, 9].

Treatment of cystic nephroma consists of surgical excision, including nephrectomy or nephron-sparing surgery with tumor-free margins [13, 16]. Long-term follow-up is recommended due to cases of recurrences of cystic nephroma have been described in the literature [16].

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囊腫性腎細胞瘤合併肉瘤性轉變：病例報告

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囊腫性腎細胞瘤是一種少見的腎臟腫瘤，且在組織學上通常呈現良性。我們在這裡報告一個42歲女性診斷為右腎下端的囊腫性腎細胞瘤合併有小部分的肉瘤性質變化。手術前進行超音波，電腦斷層攝影的評估。病理上顯示為一囊腫性腎細胞瘤合併局部呈現原生間質性，未分化的，胚胎性的肉瘤變化。

關鍵詞：腹部；電腦斷層攝影；腎臟腫瘤；肉瘤；超音波