Adult cystic nephroma: A case report and a review of the literature

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Abstract

A 23-year old women who underwent radical nephrectomy due to right renal mass is presented. The histopathological examination is reported as adult cystic nephroma, a rare benign lesion of the kidney. The epidemiology, differential diagnosis, histopathological features, and treatment alternatives are discussed and the literature is reviewed. © 2009 Elsevier Inc. All rights reserved.

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1. Introduction

Cystic nephroma (CN) is an uncommon, benign lesion of the kidney with uncertain etiology. Cystadenoma, cystic renal hamartoma, polycystic nephroma, multilocular cystic nephroma, and papillary cystadenoma are the other designations for this entity [1]. Recent advances in diagnostic imaging have resulted in an increased awareness of this type of renal tumor. In this article, we present our case of adult CN and a review of the literature.

2. Case reports

A 23-year old woman was incidentally diagnosed with right renal mass while investigating urinary infection. The patient had a history of macroscopic hematuria twice in the last 1 year. Total blood count and biochemical features were at normal range. Computerized tomography (CT) reported a \(73 \times 60\) mm in circumference, smoothly capsulated renal mass consisting cystic elements and contrast holding septations, covering the upper pole and the center of the kidney (Fig. 1). The mass was compressing the calyceal system and vascular structures at hilus, but no peripheral invasion or distant metastases were detected. No skeletal metastases were reported at the total body bone scintigraphy. Open right radical nephrectomy was performed. The postoperative period was uneventful and the patient was discharged 4 days after the operation. The weight of the resected specimen was 378 g. Macroscopic examination revealed a large multilocular cystic tumor originated from the middle and upper parts of the kidney. After dissection of the specimen from the posterior pole, a cystic tumoral lesion grayish-white in color, smoothly marginated with thin fibrous capsule was detected. A cut section showed a multilocular cystic lesion composed of noncommunicating fluid-filled cysts of various sizes separated by thin or relatively thicker fibrous septae. The mass was clearly demarcated from the surrounding normal renal tissue. Microscopically, the cysts were lined with flattened to cuboidal or hobnail epithelium (Fig. 2). The cyst epithelial cells were strongly positive for keratin and epithelial membrane antigen but negative for F VIII, CD34, and vimentin. The septae were mainly composed of mature, predominantly collagenous fibrous tissue, and contained dilated vessels. No blastemal or poorly differentiated tissue was observed. The final histopathological diagnosis was adult cystic nephroma. The vascular, ureteral, renal pelvis, and capsular surgical margines were intact. The postoperative period follow-up showed no recurrence in 36 months.

3. Discussion

Approximately 200 cases of CN have been described in the literature. The first case was reported in 1892 by Ed-
munds as “cystadenoma of the kidney” [2]. The largest series were reported by Madewell et al. in 58 patients [3]. Although most cases occur in adulthood, 2 peaks of incidence were reported, 1 in childhood, exceptional after the age 2 years, and 1 in middle age, exceptional before the age 30 years. There is a marked female preponderance of 8:1 in adult CN, whereas similar tumors in children occur with a 1:1 gender ratio or a slight predominance of males [4]. Only one case of familial CN has been reported in the English literature [1]. Most cases are asymptomatic and discovered incidentally during routine examination or radiological investigation such as CT, magnetic resonance imaging and ultrasonography. The main complaints of cystic nephroma were abdominal mass, flank pain, and hematuria [5]. Our patient was a 23-year old female adult appropriating the description in the literature, diagnosed incidentally while investigating the etiology of urinary infection, and had a history of hematuria.

Pathologically, these benign tumors are well circumscribed, encapsulated masses that contain multiple, non-communicating fluid-filled cysts. The cysts are lined with a layer of flattened or hobnail cuboidal epithelium and separated by distinct stroma, and a clear line distinguishes neoplastic tissue from normal renal parenchyma [6]. The diagnostic criteria of CN proposed by Powell have been widely accepted [7]: (1) unilateral, (2) solitary, (3) multilocular, (4) no communication between the renal pelvis and the cyst, (5) no communication between loculi, (6) a definite epithelial lining to the loculi, (7) no renal elements within the main cyst, and (8) normal residual kidney tissue, if present. Later, Boggs and Kimmelstiel modified the criteria that there is no normal nephron in septae of the cyst [8]. These criteria are similar in our case.

The differential diagnosis for these cystic tumors includes nonneoplastic cystic renal diseases, multicystic renal cell carcinoma, sarcomatoid renal cell carcinoma, and nephroblastoma. Non-neoplastic renal cystic diseases include hereditary and acquired conditions with many clinical settings and renal manifestations. In contrast to the neoplastic cystic renal lesions, the surrounding kidney parenchyma shows abnormal architecture, and remnants of nephrons are often present in the cyst walls. In multicystic renal cell carcinoma, aggregates of clear cells are present in the cyst wall. Clear cell nests should not be present in CN. Adult nephroblastomas are rare and contain blastema, which is never present in CN [9]. Also, adult CN and mixed epithelial and stromal tumor (MEST) of the kidney are benign lesions of the kidney with similar clinical, morphological, and immunohistochemical features. In addition, bear in mind that hormonal imbalance has been implied in the pathogenesis of MEST [9]. Jevremovic et al. favored that CN and MEST of the kidney likely represent a single entity, with a variable stroma-cyst ratio. The difference between CN and MEST of the kidney is based primarily on the degree of solid growth and the cellularity of the stroma. CNs are described as being purely cystic with thin-walled cysts. In contrast, epithelial and stromal tumors are partially cystic with thicker-walled cysts and contain areas of more solid growth [9].

Bal et al. reported two siblings with CN, one of them with pleuropulmonary blastoma (PPB), which is a rare entity. They described that the synchronous occurrence of both tumor types in the siblings is a strong evidence that oncogenetic factors play a role in the development of these neoplasms; they suggested that genetic analysis, such as the presence of trisomy 8, WT1 gene abnormality, and the development of other tumors, should be investigated in patients with CN and/or PPB and in their relatives [1].

The etiology of CN is uncertain, however, oncogenetic factors, genetic abnormalities, hormonal imbalance, and familiar factors might be responsible in pathogenesis and should be further investigated.

Three cases of recurrence of cystic nephroma have been described in the literature. Also, a case of local recurrence described in a unilateral cystic nephroma after complete

Fig. 1. The CT image of CN.

Fig. 2. Histopathologically, cysts lined with flattened cuboidal or hobnail epithelium (H&E ×200). (Color version of figure is available online.)
cyst enucleation in an adult was reported. Enucleation of the cyst was probably not sufficient and may have led to local recurrence [10]. Although no cases of metastatic disease have been described, long-term follow-up is recommended to rule out local recurrence [10]. In one of the largest series, 29 patients were followed from 3 months to 8 years (mean 39 months), and no evidence of local recurrence or metastatic disease was found [5]. These results in this series have been similar to our case; no recurrence or metastasis has developed in our 36-month follow-up.

Radical nephrectomy is the standard treatment modality in renal masses due to the suspicion of renal cell carcinoma, such as in our case. But nephron-sparing surgery should be kept in mind if the mass is solitary, localized, unilateral, smaller than 4 cm, and the diagnosis of cystic nephroma is considered preoperatively and verified intraoperatively by frozen biopsy. If the renal mass shows cystic formation in radiologic imaging that does not correspond with Bosniak criteria, the physician should be aware of cystic nephroma for differential diagnosis. When cystic nephroma is suspected, the patient should be prepared for nephron-sparing surgery, and intraoperative frozen biopsy should be necessary. If CN is verified by frozen biopsy, nephron-sparing surgery should be available. Nephron-sparing surgery becomes more important for the patients who have solitary kidney or contralateral renal pathology, diabetes mellitus, hypertension. Since CN is a benign lesion of the kidney, follow-up without any operation might be an alternative, if diagnosed preoperatively, for the patient who has high risk for an operation.

Further investigations, especially radiological, should be helpful for preoperative diagnosis and management of cystic nephroma.

References


