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Case Report

Multilocular Cystic Nephroma of the Kidney in a Young Adult: Case Report

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ABSTRACT

We report the case of a 20-year-old female patient who presented with a left abdominal mass of 4 years duration. Ultrasonography showed a multicystic left renal mass, and intravenous urography revealed a non-functioning kidney. Based on these findings a multicystic renal cell carcinoma was suspected and radical nephrectomy was performed. Histological examination of the resected specimen confirmed the diagnosis of multilocular cystic nephroma. This is a very rare renal tumor with only few cases described in the literature. Based on our case and a review of the literature the epidemiology, pathogenesis and diagnostic features are discussed.

Key Words: Cystic nephroma, renal cell carcinoma, kidney

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INTRODUCTION

Multilocular cystic nephroma is a very rare renal tumor of unknown etiology. It is characterized by a well-circumscribed, encapsulated mass that contains multiple non-communicating fluid-filled cysts[1]. Since its first description in 1982 by Edmunds, there have been few reports in the literature[1,2]. A variety of names have been given to cystic nephroma, including cystadenoma, cystic renal hamartoma, partial polycystic kidney and cystic partially differentiated nephroblastoma. It is usually asymptomatic, but common presenting features include abdominal or flank pain and mass[2,3]. The differential diagnosis includes cystic nephroblastoma, cystic renal cell carcinoma and other cystic developmental and neoplastic conditions. Although pre-operative diagnosis can be suggested by imaging studies, definitive diagnosis is usually made histologically. The purpose of this case report is to describe this rare tumor in an Ethiopian patient and to discuss the clinical, pathologic and diagnostic features.

CASE REPORT

A 20-year-old female patient from Gondar City, North West Ethiopia, presented with pain and a slowly growing mass in the left abdomen, which had been present for 4 years. The pain was mild and vague, but the mass progressively increased and was noted by the patient. There was no history of hematuria, no symptoms of renal failure or similar illnesses in the family.

On examination, the vital signs were normal, while abdominal examination revealed a huge bimanually palpable mass extending from the left flank to the mid-abdomen. The mass was firm in consistency and moved with respiration. No mass was detected on the right side. No other abnormality was noted.

Ultrasonography showed a huge multicystic, multi-septated mass, extending from the hypochondrium to the pelvic brim. The

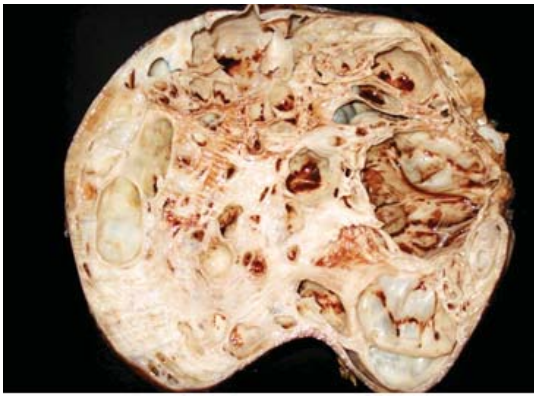

Fig. 1: Cut surface of a cystic nephroma, showing multiple variably sized cysts.

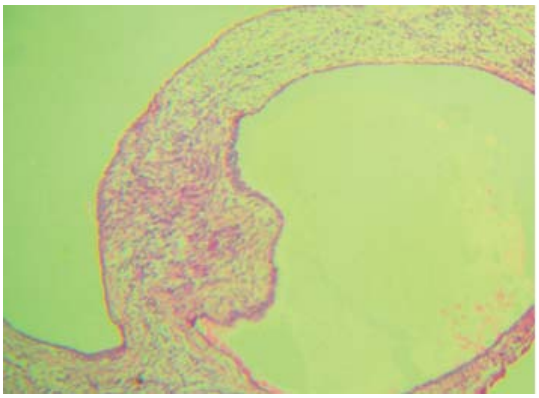

Fig. 2: Lower magnification view of cystic nephroma showing a fibrous septum lined by epithelial cells (HE x 200).

renal parenchyma was not visualized. No free fluid, peri-aortic or renal lymph-node enlargement was noted. The right kidney was in its normal position and showed no features of cystic disease. Intravenous urography (IVU) showed normal excretion on the right side but no excretion in the left kidney. Other investigations including renal function tests and hematocrit evaluation were normal.

With a clinical suspicion of cystic renal cell carcinoma the patient was operated under general anesthesia. A huge, partly cystic mass arising from the left kidney was identified and mobilized. No renal parenchymal tissue was evident on inspection intra-operatively. After ligating the renal vessels and the ureter, the mass was removed.

The post-operative course was uneventful. The patient was discharged 7 days after the operation. Clinical and radiologic follow-up at 9 months showed no evidence of recurrence.

The resected specimen weighed 1800 gm and measured 18 x 15 x 13 cm. It was a white-tan, well-encapsulated mass with a largely smooth surface. The tumor extended to the renal capsule without breaching it. The renal vessels and ureter were normal. The cut surface showed a multiloculated tumor composed of variably sized cysts measuring up to 5 cm (Fig 1). The cysts were smooth-walled and non-communicating, containing clear watery fluid. The tumor was largely cystic, but there were intervening solid areas, some of them with tiny cysts. There was no necrosis, hemorrhage or calcification.

Histologically there were multiple cysts lined by simple and regular flat or cuboidal epithelium. The intervening solid tissue was largely composed of fibrous connective tissue, with scattered mature tubular structures and dilated vessels (Fig. 2 and 3). No blastemal, mesenchymal or poorly differentiated tissue was observed.

DISCUSSION

Renal cystic diseases are heterogeneous entities which include hereditary, non-hereditary and acquired conditions. Among the rare acquired causes of cystic renal diseases are neoplastic lesions. Neoplastic lesions refer to isolated cystic masses not accompanied by cystic changes of the remaining renal parenchyma. There are 4 major entities in this category: cystic nephroma, mixed epithelial and stromal tumor, cystic partially differentiated nephroblastoma, and multilocular cystic renal cell carcinoma.

The first description of cystic nephroma was given by Walter Edmunds in 1892 under the term “cystic adenoma of the kidney”[1].

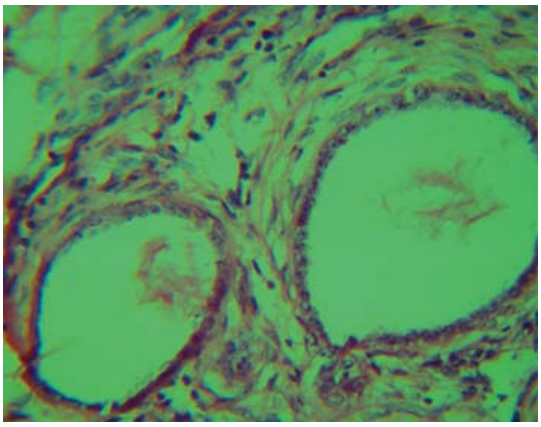

Fig. 3: Higher magnification view showing cysts lined with regular cuboidal epithelium (HE x 400)

Since its original description there has been much confusion regarding the true nature of the lesion, its origin and relationship with other predominantly cystic lesions of the kidney, such as cystic partially differentiated nephroblastoma and the cystic variant of Wilms tumor[2,3]. The different names given to the lesion also made classification more challenging. Cystic nephroma has also been referred to as multilocular cystic tumor of the kidney, cystic partially differentiated nephroblastoma, renal cystadenoma and partial polycystic kidney[4].

In 1989, Joshi and Beckwith modified the histologic criteria originally set in 1956 by Boggs and Kimmelstiel, which include the following criteria[5]:

- Cystic nephroma is entirely composed of cysts and septa.
- The lesion forms a discrete mass.
- The septa are the only solid component.
- The cysts are lined by flattened, cuboidal or hobnailed epithelium.
- The septa are composed of fibrous tissue in which mature tubules may or may not be present.
- The cysts should not communicate with the renal pelvis.
- The residual renal tissue should be essentially normal. In addition, poorly differentiated tissues and blastemal cells should not be present; lesions containing these components are now regarded as cystic partially differentiated nephroblastoma.

All of the above features were present in our case.

Cystic nephroma is a very rare tumor, with only about 200 case reports in the literature[6]. Two peaks of incidence are reported for cystic nephroma: one in the childhood, usually less than 2 years of age, and the other in adulthood. In childhood it is more common in boys, while in adults it is more common in women with a ratio of 8:1. It usually affects one kidney, but bilateral cases have been reported[7]. Most adults and pediatric patients with cystic nephroma are asymptomatic at presentation. Abdominal mass is a common presenting feature and less frequently patients come to attention with hematuria, hypertension or urinary tract infection.

The etiology and pathogenesis of cystic nephroma is not well established, and there have been several proposed explanations[1-5]. Previously cystic nephroma was thought to be a developmental defect. Currently the most accepted view proposes that cystic nephroma is a well-differentiated neoplasm within the spectrum of a renal blastemal tumor and is related to Wilms tumor[5]. Some of the arguments for the neoplastic nature of cystic nephroma include sharp circumscription of the lesion from the normal tissue and lack of association with other developmental defects.

Pre-operative diagnosis is generally suggested by computerized tomography and magnetic resonance imaging which can detect the character of the cysts. Imaging helps in differentiating cystic nephroma from other cystic renal masses such as Wilms tumor with cyst formation, clear cell sarcoma, cystic variants of mesoblastic nephroma, and

cystic renal cell carcinoma[8]. In our case the exact diagnosis could not be suggested, and this could be a result of the extensive cystic nature of the mass with very little normal residual renal parenchyma.

Cystic nephroma generally behaves in a benign fashion with no reports of metastasis. However, recurrences have been reported in the literature[9]. Long-term follow-up is generally recommended to rule out local recurrence. Castillo et al. followed 29 patients from 3 months to 8 years but found no recurrence or local metastasis[10].

Simple nephrectomy has been the preferred modality of treatment for cystic nephroma. The type of resection varies from complete nephrectomy to partial nephron-sparing procedures. Kidney-sparing or nephron-sparing procedures have recently gained larger acceptance as treatment of choice for small renal masses. In cases of solitary, localized, unilateral lesions less than 4 cm with frozen section proven diagnosis, nephron-sparing surgery is advocated[2]. Radical nephrectomy is associated with increased cardiovascular morbidity and mortality, chronic kidney disease and decreased overall survival. Partial nephrectomy preserves renal functions and decreases the risk of chronic kidney disease which is an independent risk factor for cardiovascular disease. Consequently, many authors suggest that routine radical nephrectomy for small renal tumors is unjustified and nephron-sparing procedures should be done whenever possible[11].

In our patient a diagnosis of cystic renal cell carcinoma was considered preoperatively, and complete nephrectomy was performed. Radical nephrectomy is also the preferred mode of treatment for such large tumors, including cystic nephromas. Nine months after treatment the patient was asymptomatic and there was no radiologic evidence of recurrence.

In conclusion, though rare, cystic nephroma should be considered in the differential diagnosis when clinical and imaging studies suggest a multilocular renal mass, and histopathologic examination is advised to confirm the diagnosis.

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