Multilocular Cystic Nephroma – A Surgical and Radiological Dilemma

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ABSTRACT

Multilocular cystic nephroma is a slow growing benign renal tumor. It has been identified as exclusive adult lesion, more common in females. It commonly occurs as an asymptomatic mass, occasionally with hematuria. Clinical presentation & radiological features puts urologist under dilemma. Only a few studies have correlated the Bosniak renal cyst classification with pathological findings; none of them has managed to recruit many patients, and all have case selection bias. Although new imaging techniques are available, surgical excision and histological analysis of the tumor are the only effective methods to distinguish benign from malignant cystic lesions of the kidney. Here in we present a case to highlight the need for proper diagnosis and designation of these tumors. These renal tumors are benign and carry an excellent prognosis.

1. Introduction

Multilocular cystic nephroma is a unique and characteristic renal lesion with benign biological behavior. Multilocular cystic renal tumor is characterized as a solitary, well-circumscribed, multiseptated mass of noncommunicating fluid-filled loculi that is surrounded by a thick fibrous capsule and compressed renal parenchyma. In the past, multilocular cystic renal tumors have been considered to be lesions of developmental origin, hamartoma, or hamartomas with malignant potential. In 1956, Boggs and Kimmelstiel first proposed the true neoplastic nature in a case report, suggesting the term benign multilocular cystic nephroma for this condition [1]. Most adults present with variety of non specific signs and symptoms, including abdominal and flank pain, urinary tract infection, and hypertension. Hematuria, either microscopic or gross, can occur. The differential diagnosis includes a cystic Wilm's tumor in children and a cystic renal cell carcinoma in adults. Imaging modalities, like ultrasounds (US), Computed Tomographic (CT) scans, and Magnetic Resonance Images (MRI) are helpful in revealing multilocular cystic nature of the lesion and its content, but cannot distinguish multilocular cystic nephroma from other entities with similar presentation. The Bosniak system has been widely accepted because it addresses a difficult clinical problem, is easy to apply, and is well liked by radiologists and urologists [2]. The diagnostic performance of the Bosniak system is broadly sound, but it can be difficult to accurately classify category II and III cysts. There is lack of evidence supporting the classification's ability to distinguish between these surgical and non-surgical cases. In addition, interobserver variation is greatest in these cases [3]. Here in we present a case of multiloculated cystic nephroma diagnosed after histopathological examination. Radical nephrectomy was performed because of clinical and radiological suspicion of renal cell carcinoma.
2. Case Report

A 53-year-old woman was admitted with history of mild fever and gross hematuria. She had undergone mitral valve replacement 10 years ago. She complained of repeated burning micturation and flank pain, since several months. Her routine blood count was normal. ESR was in upper normal range. Routine urine examination showed plenty of red blood cells. No pus cells noted. Urine culture was negative. On ultrasound examination left kidney showed a complex renal cyst (Fig. 1a). On further image analysis, CT scan confirmed complex, renal cyst involving the mid and lower zone (Fig.1b). A second opinion was obtained from a senior radiologist, with divided opinion between Bosniak Class III/ IV. Urologist rendered treatment options available, to the patient for both class of renal cyst (III/IV). Considering risk of hindering a malignancy, patient opted for complete nephrectomy.

We received a nephrectomy specimen measuring 10x 6x 4.5 cm. Weighed 896 grams. Outer surface was unremarkable. Cut surface showed relatively well-circumscribed lesion composed of multiple, small non-communicating cysts. Cysts measured 0.1 to 2 cm, many were filled with serous fluid. The large cavity was filled with blood clot. The pelvi-calyceal system was pushed aside by relatively demarcated lesion. On microscopic examination many cysts of varying sizes were noted (Fig.1c). Cysts were lined by flattened to “hobnail” cells (Fig.1d). The intervening stroma between the cysts was composed of fibroblasts, dilated tubules and chronic inflammatory cells. No blastemal or heterologous elements were noted. The lesion was relatively demarcated from adjacent normal kidney. Based on these findings a diagnosis of Multilocular cystic (mesoblastic) nephroma was rendered.

3. Discussion

Multilocular cystic nephroma is a distinct renal tumor of uncertain origin. Joshi and Beckwith proposed a modification to the existing terminology [4]. Their modification emphasized a neoplastic rather than a developmental or hamartomatous origin. First, they recommended that the cystic nephroma be used to describe a multicystic tumor lacking blastemal or other embryonal elements. Second, they suggested that the term cystic partially differentiated nephroblastoma (CPDN) be used to denote a predominantly cystic lesion without nodular solid regions and in which the septa contain blastemal or other embryonal elements. Furthermore, they proposed that both terms be used as subsets of the category term multilocular cystic renal tumor. Our case fits into this terminology, more accurately.

Multilocular cystic renal tumor has bimodal age and sex distribution and tends to occur in children (mostly boys with CPND) between 3 months and 4 years of age and in adults (mostly women with cystic nephroma) between 40 and 60 years of age. Multilocular cystic renal tumor is usually solitary, but bilateral tumors have been described. Recent evidence suggests that in the adult population, tumors that were initially diagnosed as cystic nephroma actually represent a genetically and histologically
distinct entity called mixed epithelial and stromal tumor of the kidney [5]. Some authors have proposed that cystic nephroma and mixed epithelial and stromal tumor of the kidney represent a spectrum of the same entity [6]; however, further studies are needed to challenge this hypothesis. In our case the stroma was composed of dilated tubules & fibroblasts.

At CT, cystic nephroma typically appears as a well circumscribed, encapsulated multicystic mass with variably enhancing septa and no excretion of contrast agent into the loculi. The contents of the cyst may have similar or slightly higher attenuation than that of water, and if the cystic spaces are very small, the densely packed septa can mimic a solid mass [7]. On Magnetic resonance imaging the contents of the cyst are hyperintense on T2-weighted images, possibly because of the different concentrations of old hemorrhage or protein. The septa have also been shown to enhance on MR images following administration of gadolinium [8] Although renal scintigraphy has been described in the work-up of MCRT, it presently has no diagnostic role. There is lack of evidence supporting Bosniak system classification’s ability to distinguish between surgical and nonsurgical cases. It can be difficult to accurately classify category II and III cysts. Such diagnostic delima was encountered by our radiologists. To our knowledge, no imaging features have been described in the literature to date that can be used to distinguish these tumors.

Since carcinomatous degeneration may occur within the wall of such tumors, especially Von Hippel Lindau disease, surgery remains treatment of choice for multilocular cystic nephroma [9]. Moreover cystic Wilm’s tumour and cystic renal cell carcinoma are difficult to distinguish from multilocular cystic nephroma on the basis of imaging findings alone. Our patient was well informed about the same. Although the Bosniak classification system is useful to evaluate renal cystic lesions and communicate findings, it is not definitive at the boundary between category II and III cysts. Ensuring that CT is technically adequate and conducted by experienced radiologists with all relevant clinical information available can minimize errors. Any renal cyst needs to be viewed with suspicion by both radiologist and urologist; until there are results from larger prospective follow-up studies to help stratify the risk in such cysts.

8. References


