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Prikaz slučaja /
Case report

ADULT CYSTIC NEPHROMA OF KIDNEY – *Case report*

ADULTNI CISTIČNI NEFROM BUBREGA – *Prikaz slučaja*

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Ključne reči
cistični nefrom; cista; bubreg.

Abstract

Introduction: Adult cystic nephroma (CN) is a part of mixed epithelial and stromal tumor (MEST) family, constituting about 2.4% of all primary renal lesions. It typically occures in females, especially in postmenopausal women. **Case report:** We report a case of a 44-year old female who was admitted to Urology Clinic of Clinical Centre of Vojvodina after incidental finding of a mass in her left kidney. The Computed tomography scan showed a 7x6x7 cm, unilocular, well-circumscribed cyst, localized in the middle part and upper pole of the left kidney. Macroscopic examination revealed a well-circumscribed multicystic tumor mass, measuring 7cm. It contained tan-white septa and smooth walls and was filled with clear yellowish fluid. Microscopic examination showed variably sized cysts lined mostly by cuboidal epithelium, with a focal hobnail appearance without atypia, focally flattened. The septa consisted of fibrous stroma with hypercellular areas and areas of cellular condensation ovarian stroma like. Immunohistochemical analyses showed that epithelial component was positive for Pax 8. The stroma was positive for estrogen receptor, progesterone receptor, actin, desmin, calretinin, CD 10 and partially positive for inhibin α. **Conclusion**: CN is a rare neoplasm that must be considered in the presence of multicystic changes in the kidney, and which in the absence of clear cells is often misdiagnosed as simple cortical cyst of the kidney. Although there has not been the evidence of local recurrence or metastatic in the literature, the long-term follow-up is recommended.

INTRODUCTION

Adult cystic nephroma (CN) is a part of mixed epithelial and stromal tumor (MEST) family, first described in 1892 as cystic adenoma of the kidney. [1,2] In the past different designations were used for this tumor, such as multilocular cystic tumor, renal multilocular cyst, multilocular cystic nephroma and partial polycystic kidney. [3]

CN is a rare benign neoplasm, constituting about 2.4% of all primary renal lesions. [4,5,6] It typically occurs in females, especially in postmenopausal women. A male-to-female ratio is 1:8. [4,5]

The etiology and histogenesis are controversial, and it has been considered to be a developmental lesion with malignant potential. [7]

Presenting signs and symptoms are nonspecific and include abdominal pain, **hypertension, urinary tract infection, microscopic or gross hematuria. Incidental finding is usual.** [5, 8]

It is always unilateral and large, with a mean size of 9 cm. [6, 9] Ultrasound is often the first investigation used in evaluating abdominal masses, confirmed by Computed tomography-CT scan. [7]

In this study, we report a case of adult cystic nephroma of the kidney, which was incidentally diagnosed·

CASE REPORT

We report a case of a 44-year-old female who was admitted to Urology Clinic of Clinical Centre of Vojvodina after an incidental finding of a mass in her left kidney. She had no signs or symptoms. Also, she was a non-smoker and had no family history of renal carcinoma.

The physical examination was unremarkable, without pain in the left kidney area.

Firstly, the patient underwent abdominal ultrasound (US), which revealed a mass in the left kidney. Then she went to CT which showed a 7×6x7 cm, unilocular, well-circumscribed cyst, localized in the middle part and upper pole of the left kidney (Figure 1).

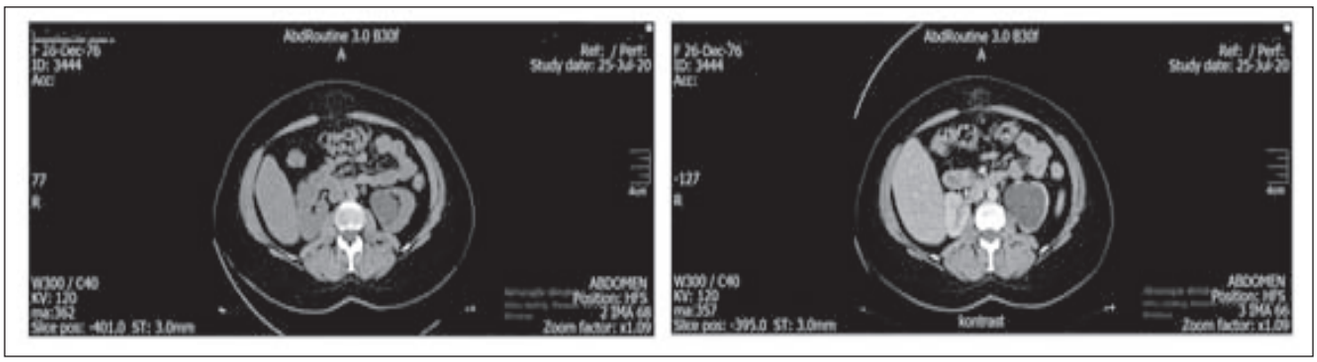

***Figure 1**: Computed tomography scan of the tumor in left kidney.*

The Council of Urology Clinic indicated radical nephrectomy of the left kidney which was performed in general anesthesia. **The operation was performed without any complication. The patient had an uneventful postoperative recovery and was discharged after the fifth day of the operation.**

Grossly, dimensions of the kidney were 14x6x1,5 cm with ureter 7,5 cm in length. The macroscopic examination revealed a well-circumscribed multicystic tumor mass, measuring 7 cm. It contained tan-white septa and smooth walls and was filled with clear yellowish fluid. Healthy parenchyma and renal pelvis were reduced, compressed.

Microscopic examination showed variably sized cysts lined mostly by cuboidal epithelium, with a focal hobnail appearance without atypia and focally flattened. The septa consisted of a fibrous stroma with hypercellular areas and areas of cellular condensation, ovarian stroma-like. Clusters of cuboidal epithelium formed tubules within the septa of the cysts.

The tumor was clearly limited, with no surrounding kidney parenchyma or pelvis invasion. Lymphovascular, perineural spaces as well as mitosis, hemorrhage and necrosis were not observed.

Immunohistochemical analyses showed that epithelial component was positive for Pax 8. The stroma was positive for estrogen receptor, progesterone receptor, actin, desmin, calretinin, CD 10 and partially positive for inhibin α (Figure 2).

Surrounding kidney parenchyma showed chronic tubulointerstitial disease.

According to the histological features and immunohistochemical characteristics the diagnosis of adult cystic nephroma, pT1b was made.

DISCUSSION

Cystic nephroma (CN) is an uncommon, non-familial, benign tumor of the kidney with nonspecific clinical presentation. Presenting symptoms include flack pain, hematuria and urinary tract infection. However, it is usually discovered incidentally, as in our case. [3]

In 1956, Boggs and Kimmelstiel proposed the criteria for the diagnosis of a multilocular cyst: 1. a multilocular mass; 2. no communication between cysts; 3. cysts lined by epithelium; 4. no communication between cysts and pelvis; 5. remaining kidney essentially normal; and 6. no normal nephrons in the septa of cysts.[11]

In 1989, Joshi and Beckwith made a modification of the criteria which specified that: 1. the tumor is composed entirely of cysts and their septa; 2. cystic nephroma is a discrete well-demarcated mass; 3. septa are the sole solid component and conform to the outlines of the cyst without

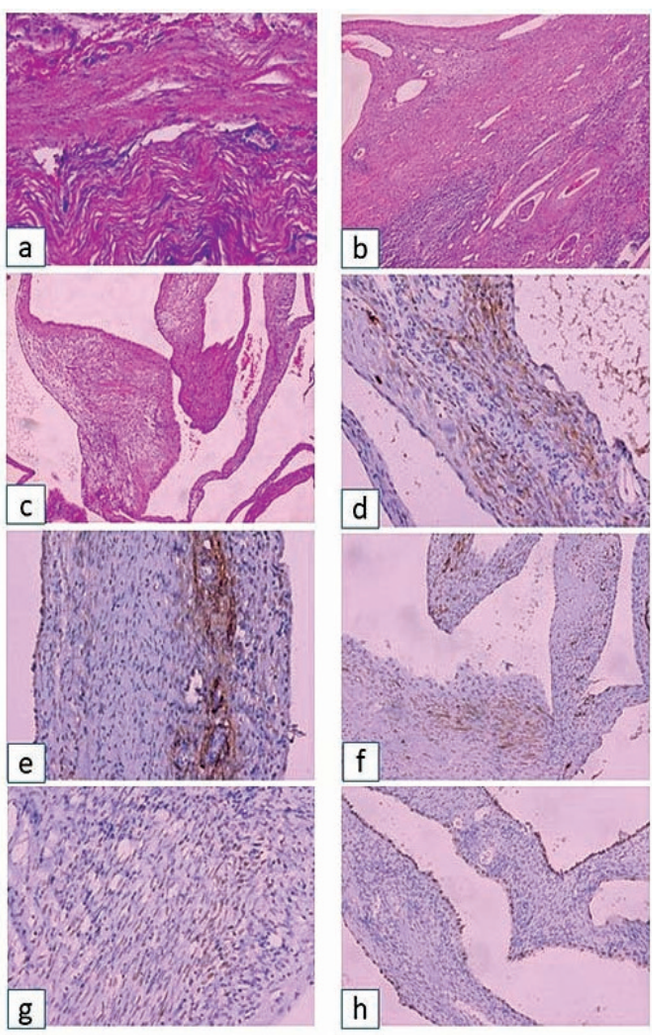

***Figure 2:** Adult cystic nephroma; a) HE, 200x; b) HE, 50x; c) HE, 50x; d) Inhibin, 200x; e) CD 10, 200x; f) Calretinin, 100x; g) Estrogen, 200x; h) Pax8, 100x.*

expansive nodules; 4. cysts are lined by flattened, cuboidal, or hobnail epithelium; and 5. septa contain fibrous tissue, in which well-differentiated tubules may be present.[2]

Ultrasound is often the first radiologic examination used in evaluating abdominal masses, which can confirm the cystic nature of the lesion. The more precise information is given by CT scan or magnetic resonance imaging (MRI).[7]

However, preoperative imaging and gross examination can't reliably distinguish CN from a malignant cystic lesion of the kidney, surgical intervention is required for both diagnosis and treatment.[11] This includes excision of the entire lesion, nephron sparing surgery and radical nephrectomy.[4] In our case, radical nephrectomy was performed and the tumor was totally resected.

Most CN are unilateral. Approximately 3.5% of patients may present with bilateral renal involvement. [9] Left and right kidney are equally affected, and lower poles are more frequently involved than the upper ones. [6] In our case, the tumor was unifocal, which is in accordance with the literature. It was located in the middle part and upper pole of the left kidney.

Imaging of cystic nephroma shows lobulate, well-defined, encapsulated multicystic mass with enhancing septa and usually without solid areas. Herniation of the mass into the renal pelvis is a characteristic, but not specific imaging finding and may cause hydronephrosis and hemorrhage. [9,12]

In the literature, grossly, cystic nephromas are large, multilocular, encapsulated tumors. On cross-section, they consist of multiple cysts, subdivided by fibrous trabeculae and filled with clear fluid. The diameter of the cysts varies from a few millimeters to several centimeters. Hemorrhage and necrosis are uncommon. Calcification has been described as a rare feature of CN.[6,7]

Mostly, those tumors have benign biological potential, but rare examples are associated with aggressive behavior, particularly those with malignant alteration. Sarcomatous degeneration to undifferentiated sarcoma, leiomyosarcoma, pleomorphic sarcoma, and malignant mesenchymoma have been described in the literature. [13]

Cystic nephroma/mixed epithelial and stromal tumors need to be differentiated from other cystic renal neoplasms that share a similar clinic presentations and imaging characteristics. [14]

Our case emphasizes clinical and radiological similarity between CN and other cystic renal neoplasms which necessitate histopathological examination of the excised tumor for definitive diagnosis.CN is a rare neoplasm that must be considered in the presence of multicystic changes in the kidney, and which in the absence of clear cells is often misdiagnosed as simple cortical cyst of the kidney.

Although the literature does not provide evidence of local recurrence or metastasis, the long-term follow-up is recommended.

Sažetak

Uvod: Adultni cistični nefrom (CN) spada u porodicu mešanih epitelnih i stromalnih tumora (MEST) i čini oko 2,4% svih primarnih bubrežnih lezija. Tipično se javlja kod žena, posebno u postmenopauzi. **Prikaz bolesnika:** Prikazujemo slučaj 44-godišnje žene koja je primljena na Kliniku za urologiju Kliničkog centra Vojvodine nakon slučajnog pronalaska lezije u levom bubregu. Na kompjuterizovanoj tomografiji uočena je unilokularna, jasno ograničena cista dimenzija 7x6x7 cm, lokalizovana u središnjem delu i gornjem polu levog bubrega. Makroskopskim pregledom opisana je jasno ograničena multicistična tumorska masa, veličine 7 cm, sa žućkasto-beličastim pregradama i glatkim zidovima, ispunjena bistrom žućkastom tečnošću. Mikroskopski, tumor je bio izgrađen od cistarazličite veličine, obloženih uglavnom kubičnim epitelom, sa fokalnim hobnail izgledom bez atipije, mestimičnoaplatiranim. Septe su bile izgrađene od fibrozne strome sa hipercelularnim područjima i oblastima nalik stromi jajnika. Imunohistohemijskom analizom, epitelna komponenta je pokazala pozitivnost na Pax 8. Stroma je bila pozitivna na estrogenski receptor, progesteronski receptor, aktin, desmin, kalretinin, CD 10 i delimično pozitivna na inhibin α. **Zaključak:** CN je redak tumor na koji se mora misliti u slučaju multicističnih promena u bubregu i koji se u nedostatku svetlih ćelija često pogrešno dijagnostikuje kao jednostavna kortikalna cista bubrega. Iako u literaturi nema dokaza o lokalnom recidivu ili metastazama, preporučuje se dugoročno praćenje.

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