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Three Dimensional (3D) Laparoscopic Nephron-sparing Treatment of a Huge Cystic Nephroma: A Case Report and Literature Review

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Abstract. *Background/Aim: Cystic nephroma (CN) is a very rare, benign, renal cystic lesion, which is characterized by a usually unilateral, multicystic kidney mass. In adults it is seen more frequently in females (1:8 male-to-female ratio). The peak incidence of CN is between 50 and 60 years of age. Median age at diagnosis is 55 years for females and 44 years for men and it is a rare entity in adults under 30 years of age. Case Report: We report the case of a 52-year-old female patient with chronic right-flank pain, who was treated at our hospital. A multiloculated 10×8.6 cm Bosniak IV renal cyst tumor was depicted on retroperitoneal computed tomography. After a three-dimensional laparoscopic partial nephrectomy, the histopathological specimen examination revealed: a multilocular cystic nephroma. Conclusion: CNs are rare benign tumors that should be included in the differential diagnosis when treating large multiloculated complex renal cysts.*

Cystic nephroma (CN) is an infrequent, benign lesion of the renal parenchyma. Its etiopathogenesis, formation and differentiation are controversial. In the past, CN has been reported as a renal cystadenoma, renal multilocular cyst, multilocular cystic tumor of kidney, multilocular cystic nephroma, and partial polycystic kidney (1, 2). Approximately 200 cases of CN have been reported in the literature, with the first report in 1892 by Edmunds (3, 4). Nowadays, due to the progression in diagnostic radiology, we have an improved identification of this type of kidney lesion (5).

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Key Words: Cystic nephroma, multiloculated renal cyst, laparoscopic partial nephrectomy.

Case Report

We report an uncommon case of a 52-year-old woman, who was diagnosed with CN during an investigation of chronic flank pain and underwent three-dimensional (3D) laparoscopic nephron-sparing surgery. A 52-year-old woman visited our Department of Urology with a history of sporadic right flank pain. The patient's medical history included hypertension and cholecystectomy 20 years before. The patient reported intermittent right flank pain during the last 3 months. Physical examination revealed mild pain during palpation of the right flank area. Laboratory findings were within normal limits. The sonographic examination revealed a cystic mass on the right kidney (about 9 cm in max. diameter). Computed tomography (CT) showed a 10 cm × 8.6 cm, multilocular, well-circumscribed cyst containing numerous calcifications on the walls with no solid components (Figure 1). A retroperitoneal MRI scan classified it as a Bosniak IV cystic lesion (Figure 2). No local invasion or distant metastases were detected. Based on the clinical and radiological findings, 3D laparoscopic nephron-sparing resection was performed to remove the renal cystic lesion. The vascular pedicle was identified and skeletonized. The mass was completely circumscribed and a clampless enucleation was followed with superselective ligation of the feeding artery of the tumor (Figure 3 and Figure 4). The parenchyma of the kidney was sutured with a continuous 3-0 V-loc suture. The entire procedure was performed in 95 minutes. Histological examination showed that the mass was a cystic nephroma. The cyst epithelial cells were strongly positive for PAX-8 (6). Stromal cells were strongly positive for estrogen receptor (ER) and progesterone receptor (PR) (Figure 5). The surgical margins were negative. The

postoperative course was uneventful, and the patient was discharged on the 5th postoperative day.

Discussion

Cystic nephromas usually present with abdominal flank pain, hematuria, urinary tract symptoms, or an abdominal lump often found during a routine examination or discovered incidentally during a radiological investigation performed for other reasons. They account for approximately 1-2% of all renal tumors (7). Two-thirds of CN presenting in children between three months and two years, more frequently in males. The adults, mainly females, older than 30 years old are affected (1:8 male-to-female ratio). The peak incidence of CN is between ages 50 and 60 years. Median age at diagnosis in women is 55 and in men is 44 years. It is rare in adults under 30 years of age. Only 5% of this rare tumor is seen in the ages between 5 and 30 years old (8). There are several etiological theories for the development of CN. There is the hypothesis that its origin could be neoplastic, probably developmental defect of the ureteric bud or a renal dysplasia, probably related to polycystic kidney disease (9). Steele *et al*. reported a case of a renal multilocular cyst with a Müllerian (ovarian)-like stroma and they suggested that this discovery would support a dysontogenetic origin for the lesion (10). Joshi and Beckwith suggested 5 criteria for the diagnosis of a CN: 1) the composition of the lesion consists entirely of cysts and their septa; 2) it forms a well circumscribed mass with clear distinction from the non-cystic renal parenchyma, 3) the only solid part of the lesion is the septa, without solid expansile nodules at the outlines of the cysts 4) cuboidal flattened or hobnail epithelium is the lining of the cysts and 5) the composition of the septa is fibrous tissue usually with presentation of well-differentiated tubules. There are no presentations of poorly differentiated tissues and blastemal cells in the septa (11). Our patient was a 52-year-old female, with intermittent right-flank pain and no other symptoms. Usually,

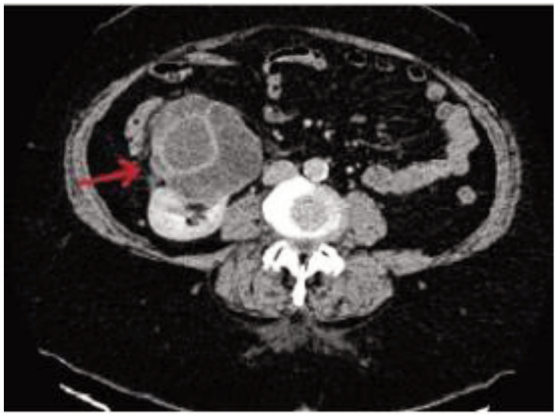

Figure 1. *Preoperative abdominal CT scan. The image demonstrates a multilocular, well-circumscribed cyst containing numerous calcifications on the walls (red arrow) located at the right kidney. CT: Computed tomography.*

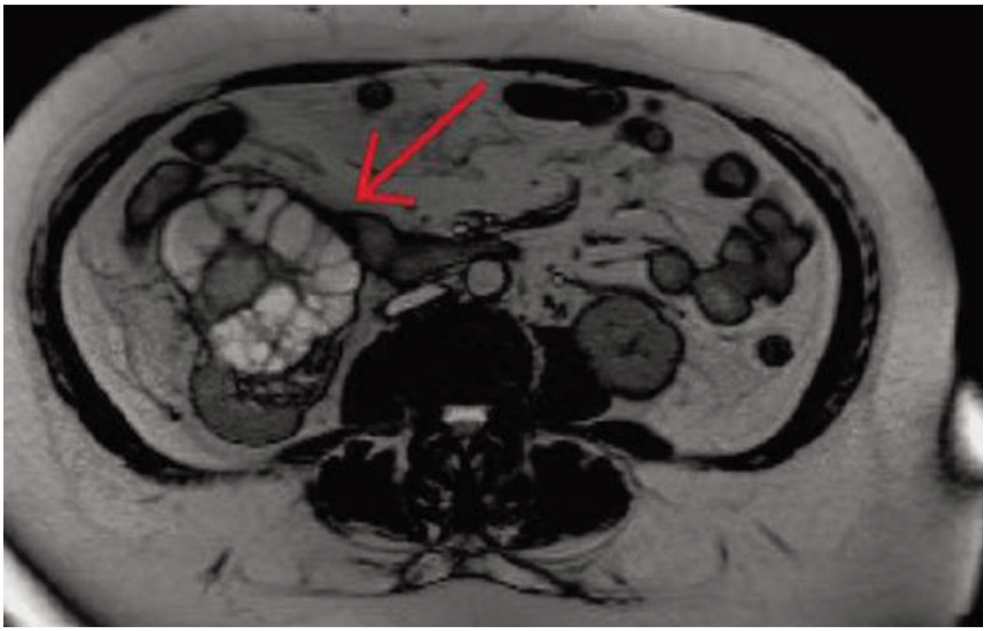

Figure 2. *Preoperative T2-weighted axial MRI of the abdomen. The image shows a large multilocular cystic mass with fluid-equivalent and atypical cysts (red arrow), located on the right kidney. MRI: Magnetic resonance imaging.*

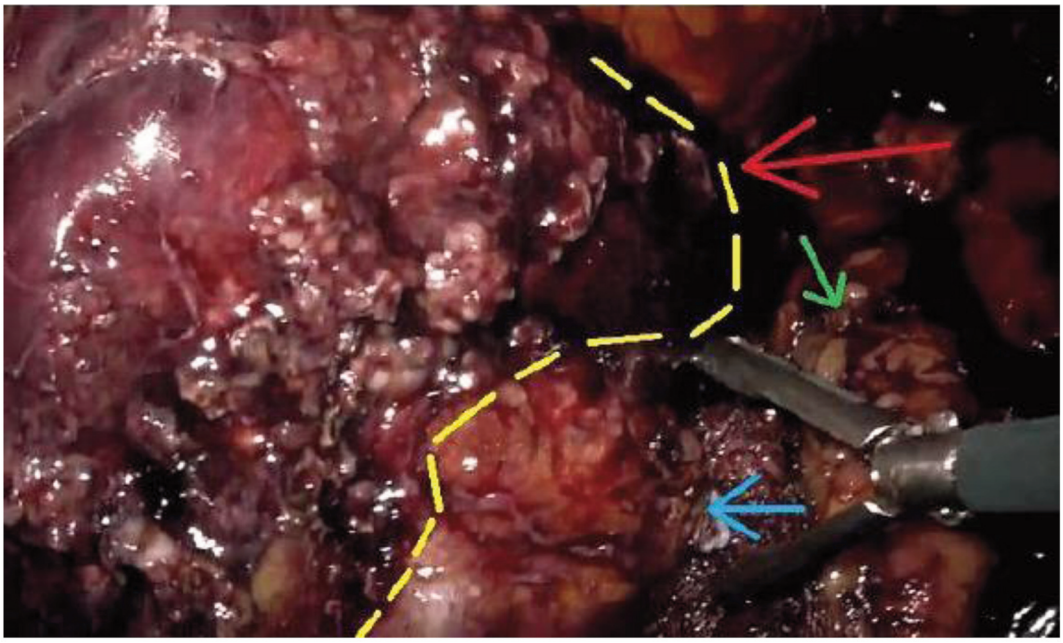

Figure 3. *Intraoperative image. The intraoperative image during the gradual resection of the neoplasm (between yellow dotted lines-red arrow) from the normal renal parenchyma (blue arrow). Ascending colon (green arrow).*

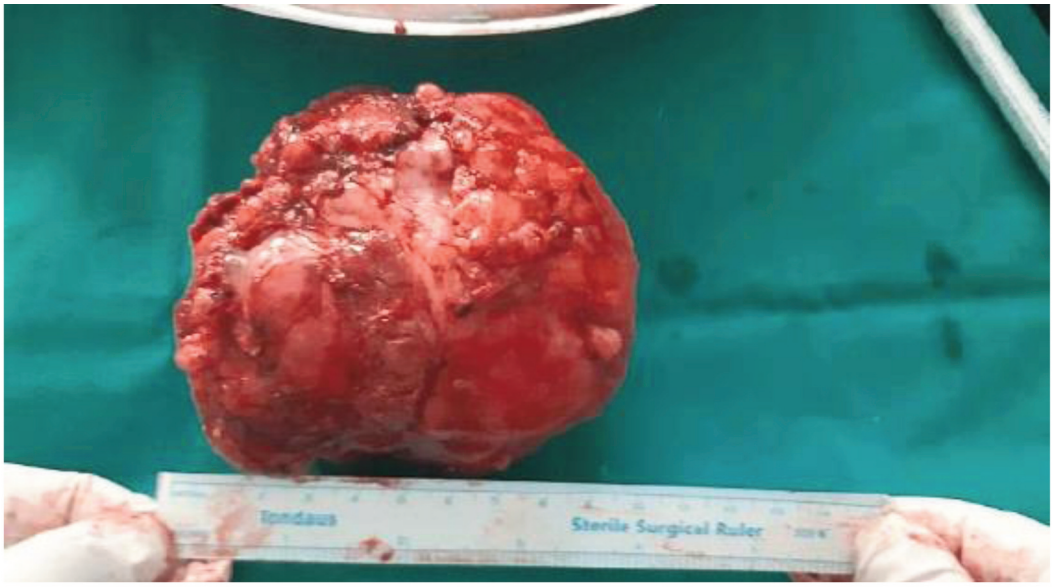

Figure 4. *Image of the resected renal neoplasm.*

CN is a mass of the kidney with no symptoms, and like other lesions of the renal parenchyma, they are incidental findings during radiological examinations. Differential diagnosis includes the cystic partial differentiated nephroblastoma, multicystic dysplastic kidney, malignant necrotic and hemorrhagic mass lesions of the kidney (renal cell carcinoma), and cystic mesoblastic nephroma (12). Imaging studies (CT and MRI) have low sensitivity in the preoperative diagnosis of CNs and the Bosniak classification is unable to differentiate CNs from RCCs. Eventually the diagnosis is histopathological.

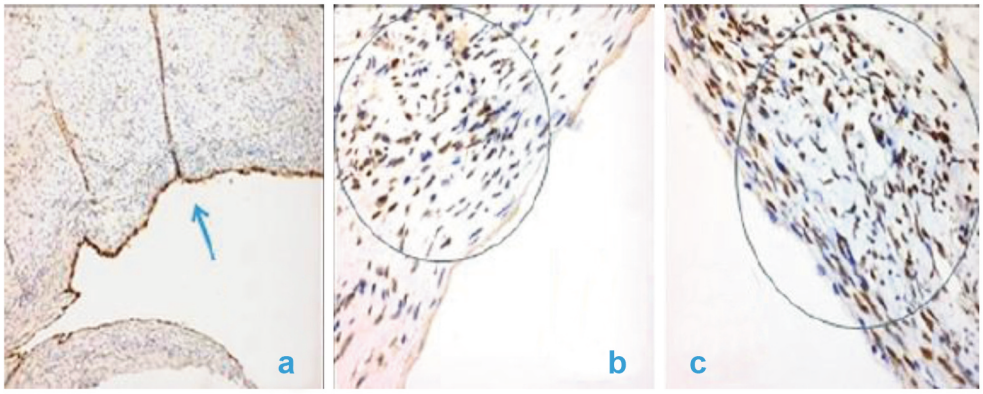

Figure 5. *Immunohistochemical study of the case. (a) Epithelial cells were strongly positive for PAX-8 (blue arrow) and stromal cells were strongly positive for ER (b) and PR (c). ER: Estrogen receptor; PR: progesterone receptor.*

Surgical treatment with resection of this benign lesion is the treatment of choice and no reports of relapse or metastasis are published to date (13).

Conclusion

CNs are rare benign tumors which should be included in the differential diagnosis when treating large multiloculated renal cysts. Preoperative diagnosis is very difficult and specific imaging features may lead the treatment towards nephron sparing procedures.

Conflicts of Interest

The Authors have no conflicts of interest to declare in relation to this study.

Authors' Contributions

ST, FG, AT, CB, NF – performed the surgical procedures; MZ, SP, AS, CB – performed literature review; ST, MZ, SP and NF prepared the draft of the manuscript; ST and NF reviewed the final version of the manuscript. All Authors read and approved the final version of the manuscript.

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