



Multilocular Cystic Nephroma Treated with Laparoscopic Nephron-Sparing Surgery: A Case Report

Busato WFS Jr*, Almeida GL and Ogata D

Department of Urology, Brazilian Society of Urology, UNIVALI, Brazil

Abstract

Multilocular Cystic Nephroma (MCN) is a rare non-genetic benign renal tumor, with approximately 200 cases described in the literature. We report a 61-year-old female patient with cystic nephroma for whom we performed laparoscopic nephron sparing nephrectomy. With 24 months of follow-up, the patient is with no signs of recurrence and normal renal function.

Introduction

Multilocular Cystic Nephroma (MCN) is a rare non-genetic benign renal tumor, with approximately 200 cases described in the literature [1-4]. It has a bimodal incidence with the congenital form commonly seen in boys about 2 years old; and the adult form in postmenopausal women [2]. The MCN definition was initially proposed in 1951 and later subdivided into cystic nephroma and partially differentiated cystic nephroma depending on the presence or absence of blastemal element, respectively [5].

There are at least 20 denominations that include multilocular cystic renal tumor, polycystic nephroblastoma, solitary multilocular cyst, renal cystadenoma and cystic nephroma [1,2]. Most patients are asymptomatic and the tumor is commonly diagnosed incidentally. For decades the standard treatment has been Radical Nephrectomy (RN). We report a case of MCN treated by laparoscopic Nephron Sparing Surgery (NSS). Clinical, radiographic, pathological and surgical aspects will be discussed.

Case Presentation

A 61-year-old woman, asymptomatic, performed a routinely ultrasound scan that revealed a complex renal cyst localized in the right kidney. A subsequent CT scan demonstrated a complex cystic tumor in the lower pole of the right kidney, well defined, with smooth walls, with several internal septa, some thick and with a slight contrast enhancement classified as a lesion Bosniak III (Figure 1A and 1B). No further abdominal lesion or lymphadenopathy was identified.

Based on these findings, a right laparoscopic heminephrectomy was realized. Surgery was performed with the placement of two 5 mm and one 10 mm trocars, dissection of the renal vascular pedicle with placement of a laparoscopic vascular clamp (bulldog) in the renal segmental artery to permit dissection of the cystic mass safely with a small margin of renal parenchyma (Figure 2A). The mass was completely resected using cold scissors, and reconstruction was done closing with running suture anchored by Hem-o-lock clips. The vascular clamp was removed totalizing a warm ischemia time of 14 minutes. With 24 months of follow-up, the patient is with no signs of recurrence

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*Correspondence:

Wilson FS Busato Jr, Department of Urology, Brazilian Society of Urology, UNIVALI, Brazil,

E-mail: wbusato@gmail.com

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Figure 1A: CT scan shows an extensive multicystic mass.



Figure 1B: CT scan shows an extensive multicystic mass.



Figure 2A: Inferior renal pole containing the tumor.



Figure 2B: Inferior renal pole containing the tumor.

and normal renal function.

Macroscopically, the cystic nephrons commonly present as expansive masses, well demarcated from the non-neoplastic renal parenchyma, with a thick fibrous pseudo-capsule. The tumor is completely cystic, with no solid component. The cysts contain clear or hemorrhagic fluid, often biochemically similar to the serum. The septa are thin, translucent and uniform (Figure 2B). Microscopically, cystic nephroma presents cysts coated by a layer of hobnail, cuboidal or flat epithelium. Mitotic figures are very rare or absent. The stromal septa consist of fibrous tissue, ranging from collagenous to myxoid, eventually presenting a similar appearance to the ovarian stroma

(Figure 3) [6].

Discussion

MCN is a rare benign tumor of the kidney, with a bimodal peak of incidence, between 2 and 4 years of age (73% are boys) and between the fourth and sixth decade. Predominating in women in the 1:8 ratio [2,4,7]. Most cases are asymptomatic and are found incidentally in routine radiological exams by pathology or as palpable abdominal mass in children [8]. In 1989 Joshi and Beckwith [8] indicated pathology diagnostic criteria: 1. tumor composed entirely of cysts and septa; 2. mass well delimited; 3. septa are the only solid components without nodular areas; 4. cysts coated by flat or cuboidal epithelium; and 5. septa with fibrous tissue, with well differentiated tubules present [9]. MCN cells are positive for various markers of distal tubule and collecting duct (CK19, EMA, CK AE1/AE3) and proximal tubule (alpha-1 antitrypsin, lysosin, CD10, CD15). These immunohistochemical findings suggest tubular differentiation [7].

The pathogenesis is unknown, but defects in development and even neoplastic origin are suggested [2,4]. Despite presenting diverse biological behavior, the infant and adult subtypes have similar histomorphology characteristics. As the tumor is similar to ovarian stroma, the presence of mullerian-like stromal cells in the kidney is possible [10]. Hormonal theory has been suggested since it is highly prevalent in women and tumor stroma is positive for estrogen and progesterone and some of the male patients with MCN were in therapy for prostate cancer [7].

The diagnosis is usually incidental, but there may be non-specific symptoms such as abdominal discomfort, hematuria, or urinary tract infections. Most are found incidentally in Ultrasound exams or CT scans performed by routine or other cause, and the average size at diagnosis is 10 cm [11]. The differential diagnosis in adults includes polycystic kidney, nephroblastoma, hydronephrotic kidney and cystic renal cell carcinoma [1]. There is no specific radiological method and it is not possible to exclude malignancy in the preoperative evaluation, surgery is necessary for diagnosis and treatment.

The treatment indicated usually is Radical Nephrectomy (NR), without chemo or radiotherapy [1,12,13], however NSS may be an option depending on the size and location of the lesion [1,2,4]. Wilkinson et al. [4] related a series of 6 cases treated by laparoscopy. Although MCN is a benign condition, recurrence has been observed and it is not known whether it results from forgotten foci or sarcomatous degeneration [10]. There are only 4 cases described in the literature of recurrence, all after Partial Nephrectomy (PN) [9]. On the other hand, in a series of 24 MCN patients treated with open PN no recurrence was found at a medium follow-up of 39 months [6].

Most of the cases described are pre date the era of laparoscopic

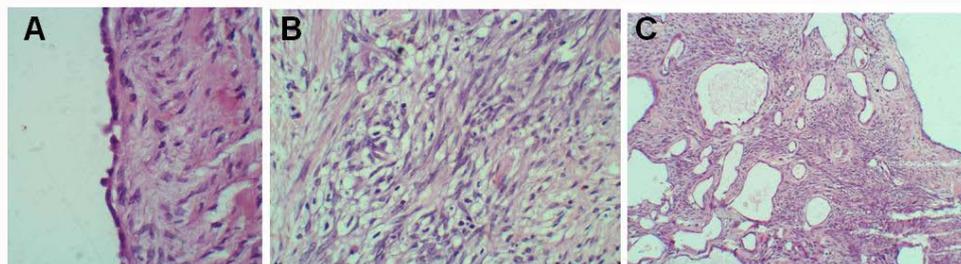


Figure 3: A) Photomicrograph showing simple flat epithelium coating parade of cyst (HE 400x); B) Photomicrography exhibiting ovarian-like stroma, containing spindle cells (HE 400x); C) Photomicrograph revealing numerous cystic structures, separated by ovarian-like stroma (HE 200x).

NSS, with techniques such as selective clamping of vessels, transoperative ultrasound and new sealing equipment. Laparoscopic PN may be a good treatment option if the diagnosis of cystic nephroma is considered preoperative. In the present case, the diagnosis of a Bosniak III cyst suggesting a cystic renal cell carcinoma was treated by laparoscopic NSS and more than 50% of the renal parenchyma was preserved.

Conclusion

MCN is a rare and benign renal tumor, presenting with good prognosis and usually diagnosed incidentally. It should be considered as differential diagnosis with cystic renal cell carcinoma. Although RN is the standard of care, it can be very aggressive for a benign pathology. We present a case treated by laparoscopic nephron sparing surgery, with no signs of recurrence, indicating that this approach may be safe and less aggressive treatment option.

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