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Solitary Fibrous Tumors of the Kidneys: Presentation, Evaluation, and Treatment

Key Words

Kidney · Solitary fibrous tumor · Nephrectomy

Abstract

Introduction: Renal solitary fibrous tumors (SFTs) are spindle cell neoplasms of mesenchymal origin, and very rare with only 46 cases reported worldwide to date. It is crucial to differentiate this tumor from other tumors of the kidney, so as to avoid unnecessary treatment. Therefore, our objective was to review reports of renal SFTs, their clinical presentations, imaging methods, and surgical management, updated to 2013. **Material and Methods:** We retrospectively reviewed articles published in the USA, Europe, and Asia from 1996 to date using PubMed, Medscape, Medline, and several major journals. We report on areas of controversy and well-established guidelines. **Results:** We reviewed 58 articles which confirmed, with a high level of evidence-based medicine, that the male-to-female ratio is equal and their most common presentation is an incidental finding on a radiological study, in which it is difficult to differentiate them from renal cell carcinoma. Nephrectomy is the gold standard treatment, with almost no recurrence. **Conclusions:** In symptomatic patients, complete surgical resection of renal SFTs may provide a very good outcome, with almost no recurrence.

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Introduction

A solitary fibrous tumor (SFT) is a spindle cell tumor originating from mesenchymal cells that was originally discovered in the pleura of the respiratory system [1]. It has also been described in extrapleural sites, including the genitourinary system. The presence of an SFT in the kidney is extremely rare. Only 46 cases of an SFT in the kidneys have been reported in the literature (table 1). The first case was described by Gelb et al. [2] in 1996. Even with the increased availability of high-resolution ultrasonography (US), computed tomography (CT) scanners, and magnetic resonance imaging (MRI), renal SFTs remain rare neoplasms. Previous data were found on gender distribution, age, presenting symptoms, optimal diagnostic tests, and treatment based on size and location; however, those reports seem to have limitations and shortcomings concerning the selection criteria related to the surgical management of these tumors, with areas of controversy leading to different management when several factors were involved, e.g. size, exact location, and natural evolution of these tumors.

At the American University of Beirut Medical Center, we recently treated a patient with a renal SFT, and subsequently reviewed in detail previous reports to describe the contemporary approach to this tumor. Furthermore,

Table 1. Cases of SFTs reported in the literature updated to 2013

Case No.	Year	Authors	Age/gender of patient, years	Site of SFT	Size of tumor, cm	Treatment	Follow-up
1	1996	Gelb et al. [2]	45/M	Rt. kidney	3.0×2.5×1.5	radical nephrectomy	died (3 mo)
2	1996	Fain et al. [33]	45/M	Rt. kidney	6×5×3.5	radical nephrectomy	8 mo; NED
3	1996	Fain et al. [33]	46/F	Rt. kidney	7.2×6×5.5	radical nephrectomy	33 mo; NED
4	1996	Fain et al. [33]	51/M	Lt. kidney	4.5×4×2.5	radical nephrectomy	2 mo; NED
5	1997	Fukunaga et al. [34]	33/F	Rt. renal peripelvis	3×2.5×2.5	Rt. nephrectomy	2 mo; NED
6	1997	Fukunaga et al. [34]	36/F	Lt. renal peripelvis	2×1.5×1.5	Lt. nephrectomy	12 mo; NED
7	1999	Hasegawa et al. [35]	64/M	Rt. kidney	4.5	radical nephrectomy	8 mo; NED
8	2000	Morimitsu et al. [27]	72/F	Lt. kidney	8.0	radical nephrectomy	10 mo; NED
9	2000	Leroy et al. [36]	66/F	Rt. renal mass extending to renal vein causing thrombus	9×7×6	Rt. radical nephrectomy	9 mo; NED
10	2001	Yazaki et al. [37]	70/M	Rt. renal pelvis	6×4.5×4	radical nephrectomy	60 mo; NED
11	2001	Wang et al. [9]	41/M	Lt. kidney	14×12×7	Lt. nephrectomy	4 years; NED
12	2001	Cortes-Gutierrez et al. [38]	28/F	Lt. renal capsule	15×11	Lt. radical nephrectomy	12 mo; NED
13	2001	Wang et al. [9]	72/M	Rt. renal	13×9×7	Rt. nephrectomy	5 mo; NED
14	2002	Magro et al. [18]	31/F	Rt. kidney	8.6	radical nephrectomy	8 mo; NED
15	2003	Llarena Ibaguren et al. [10]	51/F	bilateral	25 (Lt.) 2 (Rt.)	resection	NED
16	2003	Durand et al. [39]	35/M	Rt. kidney hilar mass	17	Rt. radical nephrectomy	6 mo; NED
17	2003	Bugel et al. [40]	50/F	Rt. kidney	11	Rt. nephrectomy	48 mo; NED
18	2004	Kunieda et al. [41]	53/M	Rt. renal	14×13×10	Rt. nephrectomy	NED
19	2004	Yamada et al. [42]	59/M	Lt. renal capsule	6.8×4.4	Lt. nephrectomy	4 years; NED
20	2004	Gres et al. [43]	83/M	Rt. kidney	9.0	Rt. radical nephrectomy	18 mo; NED
21	2005	Yamaguchi et al. [44]	51/F	Lt. kidney	10×5×10	Lt. nephrectomy	NED
22	2005	Johnson et al. [45]	51/F	Rt. kidney	11.0	radical nephrectomy	NED
23–28	2005	Pierson et al. [46] (poster presentation)	29–79 (mean 52.6)	6 cases intrarenal/ 1 case perirenal	2.2–10.1 cm/ mean 5.7 cm	nephrectomy	–
29	2006	Alvarez Mugica et al. [47]	36/M	Rt. kidney	–	radical nephrectomy	NED
30	2006	Ferrari et al. [48]	4/M	Rt. middle and lower pole kidney	8.5×6.0×15.0	Rt. nephrectomy	–
31	2006	Fine et al. [49]	76/M	Lt. kidney	12.0	Lt. radical nephrectomy	malignant
32	2007	Znati et al. [50]	70/M	Lt. kidney	15×12×4	radical nephrectomy	6 mo; NED
33	2006	Kohl et al. [17]	85/F	Lt. renal hilum	4.5	Lap. radical nephrectomy	–
34	2006	Koroku et al. [51]	18/F	upper calyx of left kidney	3.0	Lap. Lt. nephrectomy	15 mo; NED
35	2007	Constantinidis et al. [52]	26/M	Rt. upper and mid-pole attached to renal capsule	–	Rt. nephrectomy	6 mo; NED
36	2007	Bozkurt et al. [53]	51/F	Lt. kidney/mid-lower renal pole adjacent to renal pelvis	4.0×3.5×3.5	Lt. radical nephrectomy	malignant 10 mo; NED
37	2008	Magro et al. [54]	34/F	Lt. kidney upper pole	9.0	Lt. nephrectomy	malignant 21 mo; NED
38	2009	Petrella et al. [55]	–	synchronous SFT in the left parietal pleura and left kidney	–	diagnosed by echo-guided histological biopsy, was closely monitored by CT scan and US	renal lesion remained stable
39	2009	Hirano et al. [56]	75/F	Lt. kidney lower pole	4.5×3.5	Lap. Lt. nephrectomy	malignant 9 mo; NED
40	2009	Makris et al. [32]	35/M	Rt. kidney compressing upper pole leading to mild hydronephrosis	8.0	Rt. partial nephrectomy followed by autotransplantation	NED on MRI follow-up
41	2010	Hsu et al. [57]	51/F	Lt. palpable renal mass in retroperitoneum	10×1×8 in mid- and upper portion	Lt. radical nephrectomy	18 mo; NED
42	2011	Naveen et al. [14]	52/F	Rt. kidney	18.0×9.0×10.0	Rt. radical nephrectomy with renal vein thrombectomy	6 mo; NED
43	2011	Marzi et al. [58]	72/F	Lt. hemiabdomen mass	19.0	Lt. radical nephrectomy	–
44	2011	Hsieh et al. [8]	50/F	palpable Rt. flank mass; Rt. perirenal space	9.0×9.0×6.0	Rt. radical nephrectomy	30 mo; NED
45	2012	Sfoungaristos et al. [5]	72/M	upper pole of left kidney	7.0×6.0	Lt. radical recurrence	recurrence in retroperitoneum after 2 years 9 mo; NED
46	2012	Guo et al. [20]	60/M	Rt. renal middle portion invading renal capsule and mucosa of renal pelvis	–	Lap. radical nephroureterectomy	malignant; 3 mo later low back pain; thoracic vertebral metastasis; resection of intracranial tumor; 2 mo later, systemic multiple metastases; deceased
Our case	2012		49/F	Rt. kidney	5.0×5.0×2.5	Rt. Lap. radical nephrectomy + paracaval LN dissection	NED

Rt. = Right; Lt. = left; Lap. = laparoscopic; LN = lymph node; NED = no evidence of disease; mo = months.

our patient presented in the context of acute pyonephrosis, which, to our knowledge, is the first case of a renal SFT presenting as acute pyonephrosis in the English literature. Therefore, the objective of the present review was to detail renal SFTs, their incidence, epidemiology, etiology, clinical presentation, optimal diagnostic methods, and pathology, and to finally clarify the best therapies as of 2013.

Material and Methods

Between 1996 and 2013, the information assessed included a thorough review of previous reports, including 58 articles published in the USA, Europe, and Asia, on renal SFTs. We provide the most up-to-date information and well-established guidelines. The information covered was retrieved from PubMed, Medscape, *Annals of Internal Medicine*, *Clinical Imaging*, *Histopathology*, *Archives of Surgery*, *JACS*, the AUA and *Journal of Urology* website, *BMJ*, Medline, and Springer Link.

We also included, for illustrative purposes, retrograde pyelography, US, and CT findings, in addition to the gross macroscopic and microscopic images of our patient who was recently treated in our medical center for renal SFTs presenting as acute pyonephrosis.

Results

Incidence, Origin, and Etiology

SFTs, formerly classified as hemangiopericytomas, are pleural-based tumors of mesenchymal origin. An SFT is characterized by patternless proliferation of spindle cells, originating from fibroblasts and primitive mesenchymal cells [3]. Extrapleuristic locations have been described in the literature (table 2). Among these locations, the kidney represents one of the rarest locations [4].

Microscopically, around 90% of the cases are benign, whereas the remaining are malignant [5]. Mesenchymal tumors are relatively uncommon in the kidney. Benign mesenchymal tumors that may arise in kidneys are angiomyolipomas, myxomas, SFTs, and benign peripheral nerve tumors such as solitary neurofibromas [6].

Epidemiology

Epidemiological studies have shown that age of presentation is between 28 and 83 years, with a mean age of 52 years, and a male-to-female ratio approximately equal to 1. The right kidney is commonly more affected than its counterpart [7]. Within the kidney, the renal capsule has been found to be the most common site of origin of renal SFTs [5].

Table 2. Extrapleuristic locations of SFTs

Orbital cavities
Thyroid
Sublingual gland
Liver
Pancreas
Retroperitoneum
Skin
Kidney
Periosteum
Bone
Mammary gland
Nasal cavity
Paranasal sinuses
Meninges
Adrenals
Urinary bladder
Prostate
Spermatic cord
Testes

Clinical Presentation

Clinical presentation varies between an incidental finding of a mass on radiological imaging to a palpable mass [8]. Flank pain or abdominal pain, along with gross hematuria, are among the common symptoms. Rarely, SFTs cause paraneoplastic syndromes, such as hypoglycemia related to secretion of insulin-like growth factors [21], but so far this has never been reported in the literature in the presence of renal SFTs. The patient who presented at our medical center is among the rarest cases of renal SFT, in which the tumor was obstructing the pelvicalyceal system, and the first case in the literature leading to pyonephrosis and urosepsis.

Gross Macroscopic Appearance and Histopathology

SFTs can range in size from 2 to 25 cm, with a mean diameter of 8.75 cm [7]. Grossly, the tumor is usually well-circumscribed and pseudoencapsulated, lobulated, rubbery or firm, and exhibits a homogeneous gray or tan-white surface. Necrosis, cyst formation, and hemorrhage are usually absent, except in malignant cases [9]. One case of a bilateral renal SFT has been reported in the literature [10].

Microscopic examination of the tumor cells shows a variety of architectural patterns, with the most frequent being an intermingling of tumor cells and collagen in a random fashion, the so-called 'patternless pattern' [11]. The second most common pattern consists of alternating

Table 3. Markers and immunostaining of renal SFTs and their corresponding frequency

Markers and immunostaining of SFTs	Frequency
CD34	90–95%
CD99	70%
Bcl-2	20–35%
Epithelial membrane antigen	20–35%
Smooth muscle actin	rare focal activity
S-100 protein	rare focal activity
Cytokeratins	rare focal activity
Desmin	rare focal activity

hyper- and hypocellular areas with a hemangiopericytoma appearance [12]. Because of the major histologic overlap between SFTs and hemangiopericytomas on light and electron microscopic examination, and lack of clear criteria to distinguish between them, pathologists gradually abandoned the term hemangiopericytoma in favor of SFT. Tumor cells have a scanty cytoplasm with elongated nuclei with finely dispersed chromatin. Mitotic activity is limited, and cellular atypia and necrosis have not been observed in benign cases [13]. These histopathological findings are similar regardless of the organ involved, whether pleural or extrapleural [11].

On electron microscopy, SFTs are characterized by fibroblast-like cells with a well-developed rough endoplasmic reticulum, surrounded by collagen fibers [14]. A primordial way to differentiate SFTs from other spindle cells tumors is their high positivity for CD34, CD99, and bcl-2. In fact, CD34, which is a stem-cell adhesion marker, is considered a specific immune-peroxidase marker for SFTs [7]. Among all SFTs, 70% express CD99 and bcl-2. Immunostaining for epithelial membrane antigen and smooth muscle actin is positive in approximately 25% of these neoplasms, and immunostaining for S100 protein, cytokeratins, and/or desmin is usually absent or only focal and limited [15] (table 3).

Radiological Modalities

On US, renal SFTs have been described as hypo- or heterogeneous echogenic masses, with relatively well-defined margins, and as hypoechoic masses with intratumoral vascularity on Doppler US. Plain CT scans can show SFTs as well-circumscribed smooth lobulated lesions. Enhanced CT scans show a strong enhancement associated with cysts and regions of hemorrhage or necrosis.

MRI shows predominately low-to-intermediate signal intensity on both T1- and T2-weighted images, mostly related to the high content of fibrous collagenous tissue, hypocellularity, and relatively small number of mobile protons contained within these tumors [11]. Kim et al. [16] reported an important correlation stating that signal intensities of these tumors on T2-weighted images decrease as the collagenous component increases. Intense enhancement after intravenous contrast injection has also been reported due to the high vasculature of these tumors, and persistent prolonged enhancement on delayed images was also occasionally noticed [11].

Differential Diagnosis

The differential diagnosis of an SFT encompasses both benign and malignant spindle cell lesions [17], and includes hemangiopericytomas, fibroepithelial polyps of renal pelvis, angiomyolipomas, fibromas, inflammatory myofibroblastic tumors (pseudotumors), leiomyomas, benign fibrous histiocytomas, benign peripheral nerve sheath tumors (schwannomas/neurofibromas), renal mixed epithelial/stromal tumors, monophasic fibrous-type synovial sarcomas, fibrosarcomas, sarcomatoid renal cell carcinomas, leiomyosarcomas, and malignant fibrous histiocytomas [18]. The presence of the above immunohistochemical properties can differentiate SFTs from other malignancies, such as neurofibroma [19]. In addition, the presence of certain cytogenetic abnormalities, such as translocation t(X;18), can help rule out synovial sarcoma [17].

Prognosis

The vast majority of renal SFTs exhibit benign behavior, with only 5 malignant cases reported in the literature with secondary metastasis, such as to the thoracic vertebral spine [8, 20]. Although the clinical course of SFTs is rather unpredictable, the prognosis of SFTs is generally favorable. It is estimated that 10–15% of intrathoracic SFTs and up to 10% of extrathoracic SFTs will recur or metastasize [21–23]. Therefore, an SFT is considered an ‘intermediate malignant, rarely metastasizing’ neoplasm [21]. A malignant SFT is postulated to develop via two pathways: (1) de novo occurrence or (2) dedifferentiation or sarcomatous overgrowth from a preexisting histologically benign SFT [8].

The diagnostic criteria for malignant extrathoracic SFTs are purely microscopic, and include increased cellularity, pleomorphism, and a mitotic count more than 4 per 10 high-power fields [24]. Currently, the impact of tumor characteristics (size, hemorrhage, necrosis, and lo-

cation) in predicting clinical malignancy in extrathoracic SFTs remains to be investigated. Additionally, the presence of hemorrhage/necrosis and Ki-67 labeling index above 20% further supports a diagnosis of malignant renal SFTs [24]. Additional factors conferring a worse prognosis in SFTs is dedifferentiation or sarcomatous overgrowth, which represents an abrupt transition to a morphologically anaplastic component [25]. Studies have also shown that deep-seated locations of the tumor, overexpression of P53 and P16, loss of CD34 immunostaining, and the presence of dedifferentiated areas may indicate more aggressive behavior than its more common counterpart [25].

Therapies

Surgical resection of the renal SFT is the gold standard treatment, with a prognosis similar to its pleural counterpart [26, 27]. In 2009, Makris et al. [32] reported a case in which partial nephrectomy was done, and a follow-up MRI after 1 year showed no recurrence of the disease. To date, however, there is no evidence whether or not partial nephrectomy has the same oncological outcome as radical nephrectomy. Laparoscopic nephrectomy of the renal SFT has emerged as an alternative to open radical nephrectomy, with a comparable short-term oncological outcome.

Case Profile

We recently treated a 49-year-old woman, previously healthy, who was admitted to our institution through the emergency department, with a 1-day history of high-grade fever (40°C), chills, severe right flank pain, and lower urinary tract symptoms. On her physical exam, she had high-grade fever with right costovertebral angle tenderness. The laboratory tests showed a high white blood cell count with 85% polynuclear neutrophils and normal electrolytes, urine analysis showed a highly positive leukocyte esterase and numerous white blood cells, and urine cytology was negative for malignant tumor (repeated 3 times). US of the abdomen revealed a well-defined soft tissue mass with a heterogeneous predominantly hyperechoic echotexture, with its epicenter arising from the right renal sinus, measuring 5.6×4.2 cm (fig. 1). Enhanced CT scan of the abdomen and pelvis revealed a $5.6 \times 4.2 \times 4.0$ cm enhancing soft tissue mass (fig. 2, 3), located within the pelvis of the right kidney, in continuity with the parenchyma, associated with severe right hydronephrosis and a dilated right ureter. Delayed images (fig. 4) showed a hold-up of contrast within the right renal calyces and a right 'renal pelvis' mass compressing the ureter and displacing it anteriorly.

A few hours after presentation, the patient became septic, and a right nephrostomy tube was inserted. The right nephrostogram (fig. 5, 6) showed a splaying and dilatation of the right pelvicalyceal system secondary to an extrinsic compression of the right

Fig. 1. US of the abdomen revealing a well-defined soft tissue mass with heterogeneous predominantly hyperechoic echotexture, with its epicenter arising from the right renal sinus, measuring 5.6×4.2 cm.

Fig. 2. Enhanced CT scan of the abdomen and pelvis revealing a $5.6 \times 4.2 \times 4.0$ cm enhanced soft tissue mass located within the pelvis of the right kidney, in continuity with the parenchyma, associated with severe right hydronephrosis and a dilated right ureter.

renal pelvis. Two days later, the patient defervesced, and became hemodynamically stable. Repeat US showed an improvement of the hydronephrosis, with persistence of the right renal mass with flow on color Doppler. A ureteral stent was inserted at that time. Six weeks later, the patient underwent a laparoscopic right radical nephrectomy with paracaval lymphadenectomy, and she had an uneventful postoperative course.

Grossly, a circumscribed homogeneous gray-white mass $5 \times 5 \times 2.5$ cm was noted at the hilum of the kidney involving the renal sinus and compressing the renal pelvis (fig. 7, 8). No hemorrhage

Fig. 3. Enhanced CT scan of the abdomen and pelvis revealing a $5.6 \times 4.2 \times 4.0$ cm enhanced soft tissue mass located within the pelvis of the right kidney, in continuity with the parenchyma, associated with severe right hydronephrosis and a dilated right ureter.

Fig. 4. CT scan of the abdomen and pelvis with delayed excretory phase revealing a hold-up of contrast within the right renal calyces, and a right 'renal pelvis' mass compressing the ureter and displacing it anteriorly.

3

4

5

6

Fig. 5. Right nephrostogram showing a splaying and dilatation of the right pelvicalyceal system secondary to an extrinsic compression of the right renal pelvis.

Fig. 6. Right nephrostogram showing a splaying and dilatation of the right pelvicalyceal system secondary to an extrinsic compression of the right renal pelvis.

or necrosis was present. Microscopically, the tumor was composed of tightly packed round to fusiform cells, with clear indistinct cytoplasmic borders and forming ill-defined fascicles. A rich vascular network with staghorn configuration was seen in the background. No mitotic figures were noted (fig. 9, 10).

Lymphovascular and perineural invasion were absent. The surgical margins, ureter, and renal veins were all free of tumors. The tumor cells stained diffusely positive for CD34, bcl-2 (fig. 11, 12), and vimentin, and negative for desmin, smooth muscle actin, S100, HMB-45, epithelial membrane antigen, CD31, CD56, and WT-2. To our knowledge, this is the first case of a renal SFT presenting as acute pyonephrosis described in the English literature.

Discussion

Renal SFTs are extremely rare. Only 46 cases of SFTs in the kidneys have been reported in the literature to date (table 1), with Gelb et al. [2] describing the first one in 1996. We report a 47th case from 2013 – the first in the literature to present as acute pyonephrosis. SFTs are spindle cell tumors originating from mesenchymal cells and were originally discovered in the pleura of the respiratory system [1]. Some other extrapleural sites have been discovered, and a clear etiology still needs to be elucidated.

Color version available online

Fig. 7. Macroscopic gross picture of the specimen showing a well-circumscribed homogeneous gray-white mass, $5 \times 5 \times 2.5$ cm, at the hilum of the kidney involving the renal sinus and compressing the renal pelvis.

Color version available online

Fig. 8. Macroscopic multi-cross section of the specimen showing a well-circumscribed homogeneous gray-white mass, $5 \times 5 \times 2.5$ cm, at the hilum of the kidney involving the renal sinus and compressing the renal pelvis, pressing the renal pelvis.

Color version available online

Fig. 9. Microscopic picture of a renal SFT ($\times 10$) showing tightly packed round to fusiform cells, with clear indistinct cytoplasmic borders and forming ill-defined fascicles.

Color version available online

Fig. 10. Microscopic picture of a renal SFT ($\times 40$) showing tightly packed round to fusiform cells, with clear indistinct cytoplasmic borders and forming ill-defined fascicles, with no evidence of mitotic figures.

SFTs were previously classified as hemangiopericytomas, but we currently know that SFTs are characterized by spindle cells, originating from fibroblasts and primitive mesenchymal cells, and that they proliferate in a 'patternless' architecture [3]. We cannot currently predict their propensity for malignancy based on any tumor

marker or blood test, but we can attest that statistically 90% of renal SFTs are benign and 10% are malignant [5].

We still need more randomized prospective studies and molecular biologic experiences to elucidate the reason why certain tumors are malignant from the beginning, and how to predict the unfortunate progression of

Color version available online

Fig. 11. Microscopic picture of a renal SFT showing diffusely positive staining for CD34.

Color version available online

Fig. 12. Microscopic picture of a renal SFT showing diffusely positive staining for bcl-2.

a benign tumor to a malignant cancer. The differential diagnosis of renal SFTs seems to be restricted to extremely rare tumors as well, including lipomas, myxomas, and solitary neurofibromas [6], and they can be easily differentiated on histopathological studies with recent appropriate immunostaining techniques. The age of presentation fluctuates between 28 and 83 years, with a mean age of 52 years, confirming that the pediatric population is not affected with this disease.

More cytogenetic studies need to be done in order to assure that no chromosomal abnormality can occur in the first two decades of life. However, a recent case report published by Choy et al. [28] in 2012, revealed that a mixed epithelial and stromal tumor of the kidney was detected in a 14-year-old boy. It seems that there is no sex-linked chromosomal defect since the male-to-female ratio is around 1. An anatomical predisposition or a certain venous drainage may be the cause behind a right-sided preponderance, as shown in the series of Katabathina et al. [6]. The reason why the capsule is mostly affected, within the kidney, as demonstrated by Sfoungaristos et al. [5], is still not very clear.

Similar to renal cell carcinoma, renal SFTs may present with the typical triad of flank pain, a palpable abdominal mass, and gross hematuria. Again, however, these symptoms are rare, and renal SFTs are found mostly on an incidental basis during radiologic studies, as shown by Hsieh et al. [8]. However, renal SFTs, as opposed to renal cell carcinomas, cause almost no paraneoplastic

syndromes. The patient who presented at our medical center represents the first case of renal SFT in the literature in which the tumor was obstructing the pelvicalyceal system with pyonephrosis leading to urosepsis.

SFTs can range in size from 2 to 25 cm, with a mean diameter of 8.75 cm as shown in the series of Katabathina et al. [6]. This means that it may be technically challenging to remove those tumors exclusively by laparoscopy or robotic surgery.

One case of a bilateral renal SFT has been reported in the literature by Llarena Ibarguren et al. [10], and more studies need to be conducted in order to elucidate if renal SFTs are associated with syndromal disorder.

We now know that there is a very precise way to differentiate SFTs from other spindle cell tumors: they are highly positive for CD34, CD99, and bcl-2. Katabathina et al. [6] demonstrated that CD34 is considered a specific immune-peroxidase marker for SFT; however, serum tumor markers have yet to be detected, and it will be easier to have a preoperative diagnosis in order to identify those tumors and for the regular postoperative follow-up.

Radiologic studies remain one of the best tools to identify, classify, and study the extent of these tumors. In fact, they represent the most common method of detection. US is a reliable, fast, and inexpensive diagnostic tool for detecting renal SFTs. They present as a hypo- or heterogeneous echogenic mass with relatively well-defined margins. CT scans are more accurate in terms of size, nearness to the renal hilum, vascular supply, and local

extent, and are an excellent tool in the armamentarium of the urologic surgeon before the surgical intervention. Plain CT scans may show a well-circumscribed smooth lobulated lesion; however, enhanced CT scans can confirm the diagnosis with a strong enhancement associated with cysts and regions of hemorrhage or necrosis. MRI is rarely ordered and remains a radiologic test to be done in case of uncertainty or in case of renal insufficiency limiting the clinician in the administration of intravenous contrast dye. Since these tumors are rich in collagen, due to their fibrous component, MRI shows a low-to-intermediate signal intensity on both T1- and T2-weighted images [11]. Kim et al. [16] demonstrated that signal intensities of these tumors on T2-weighted images decrease as the collagenous component increases, and therefore MRI is gaining specificity in that context.

The differential diagnosis of renal SFTs is very vast as well, and includes hemangiopericytomas, fibroepithelial polyps of renal pelvis, angiomyolipomas, fibromas, inflammatory myofibroblastic tumors (pseudotumors), leiomyomas, benign fibrous histiocyctomas, benign peripheral nerve sheath tumors (schwannomas/neurofibromas), renal mixed epithelial/stromal tumors, monophasic fibrous-type synovial sarcomas, fibrosarcomas, sarcomatoid renal cell carcinomas, leiomyosarcomas, and malignant fibrous histiocyctomas [18], but only the final histopathological markers are able to differentiate them. This is why some authors would advocate the role and importance of renal biopsy in case of an inability to confirm the diagnosis as outlined by Ingels et al. [29].

Regarding their prognosis, renal SFTs are almost exclusively benign tumors, although 5 malignant cases have been described in the literature. Therefore, a full metastatic work-up should always be done in the preoperative setting, assuming that the tumor seen on radiology might have a very rare extension to bone or liver, as shown in the series of Hsieh et al. [8] and Gang et al. [20]. The final histopathological result can confirm their probable extremely rare malignant component in the presence of hemorrhage, necrosis, sarcomatous overgrowth, and a 20% Ki-67 proliferative index as described by Guillou et al. [24, 25]. We lack studies that can confirm, with a high index of suspicion, P53 or P16 overexpression should indicate a malignant renal SFT. More randomized prospective studies need to be conducted in order to elucidate the role of these markers and their relation to apoptosis.

Surgery remains the single most curative therapeutic modality. Radical nephrectomy is the treatment of choice, although partial nephrectomies have been rarely

attempted, as reported in the series of Makris et al. [32] in 2009. An appropriate 'RENAL' nephrometry score may be calculated to evaluate the complexity of such tumors, but to date we are not sure whether or not partial nephrectomy has the same oncological outcome as radical nephrectomy. Nowadays, laparoscopic radical nephrectomy or robotic radical nephrectomy, in the hands of excellent surgical expertise, may be considered the first-line surgical technique, and the upper size limit of 7–10 cm may be utilized [30]. However, another debate in the management of the renal SFTs is the value of lymph node dissection, especially when lymph nodes are not involved on the imaging studies in the preoperative setting.

Finally, insights into new treatment options for SFTs are emerging from the presence of hemangiopericytomas on histological examination, paving a new way for the antiangiogenic therapy as a new treatment option for SFTs; however, this is still under clinical investigation [31].

Conclusion

SFTs of the kidney are very rare entities. They are almost exclusively benign lesions that present as an incidental finding; therefore, patients are rarely symptomatic. They have specific characteristics on radiologic studies and although their differential diagnosis may be vast, their final histopathological criteria and results are exclusive. For the time being, as radical surgical resection is the cornerstone gold standard management of renal SFTs, future clinical trials are needed to demonstrate the benefit of additional chemotherapy or tyrosine-kinase inhibitors, and also to define the proper protocol in the follow-up of these patients since there is the possibility, although rare, of malignant transformation.

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