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Metachronous Solitary Fibrous Tumors in the Same Kidney: A Case Report and Literature Review

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Abstract

Solitary Fibrous Tumor (SFT) is a rare spindle cell soft tissue tumor. The combination of clinical, biochemical, and radiological features may help in lesion characterization, but only histology can provide the definitive diagnosis. Recurrence and/or metastases have been reported in up to 10% to 15% of cases. Although no clear evidence-based treatment guidelines exist, it is widely accepted that renal SFT need to be removed surgically to avoid the potential of malignant degeneration and metastases. A longer follow-up period might be necessary to definitively evaluate the clinical outcome of a renal SFT. A 36-year-old male, without comorbidities, underwent laparoscopic excision of a 7 cm left juxta-hilar pararenal mass in October 2018. Preoperative abdominal Computed Tomography (CT) was compatible with a renal cell carcinoma. The pathological study revealed an SFT. An abdominal CT scan, in May 2021, identified a new suspicious left kidney nodule. Thereafter, the patient had done a Magnetic Resonance Image (MRI) in June 2021 that confirmed a mesorrenal nodule suspicious of renal cell carcinoma (24 mm × 21 mm × 21 mm) in the left kidney. He was submitted to open excision of two left juxta-hilar pararenal lesions. Histology provided the definitive diagnosis of metachronous Solitary Fibrous Tumors (SFTs). This is a very rare case of metachronous SFTs in the same kidney of the first resection in a young patient.

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Introduction

Solitary Fibrous Tumor (SFT) is a rare entity, representing 2% of all soft tissue tumors [1]. It was first described in 1931 by Klemperer and Rabin as a distinct mesothelial tumor arising from the pleura. SFT was reported later in almost any extrapleural site. It is now distinguished into two entities, pleural and extrapleural. Extrapleural SFTs are observed in middle aged adults (mean age 50 years) and affect both genders equally. Urogenital system localizations are very rare. SFTs are usually asymptomatic when they have a small size, so the diagnosis is often delayed. Imaging features are not specific of SFT which are diagnosed as Renal Cell Carcinoma (RCC) and treated as such. Histologically, is usually a well circumscribed mass, with ill-defined fascicles of ovoid to fusiform cells and hemangiopericytoma-like vasculature, displays immunohistochemical staining for CD34 and STAT-6. Further investigation is needed to assess the clinical behavior of SFT, as well as to establish the appropriate duration and schedule of follow-up. Much of the research on this neoplasm is based on case reports that highlight its potential for clinical aggressiveness. With this same objective, we describe a very rare case of metachronous SFTs in the same kidney of the primary diagnosis.

Case Presentation

In October 2018, a 36-year-old man was referred to our center because of a left pararenal neoplasm measuring approximately 7 cm near the renal hilum. Patient had no comorbidities or usual medication. Due to complaints of nonspecific abdominal pain, an abdominal ultrasound was performed, which detected a solid nodule in the left kidney. Consequently, an abdominal CT scan was performed, which confirmed the presence of a left renal nodule suspected of RCC. He was submitted to laparoscopic excision of pararenal mass. Histological and immunohistochemical aspects fit into a 6.5 cm SFT of the left kidney. It was observed a multinodular neoplasm with well-defined borders. It was constituted by the proliferation of spindle cells without cytological atypia or only with mild atypia. Cells had round or elongated nuclei, with a thin chromatin, without prominent nucleoli, and mitotic figures were rare. There were no areas of necrosis. The cells were arranged in bundles that intersect a predominantly fibrous stroma. The neoplasm showed intense and diffuse positivity for

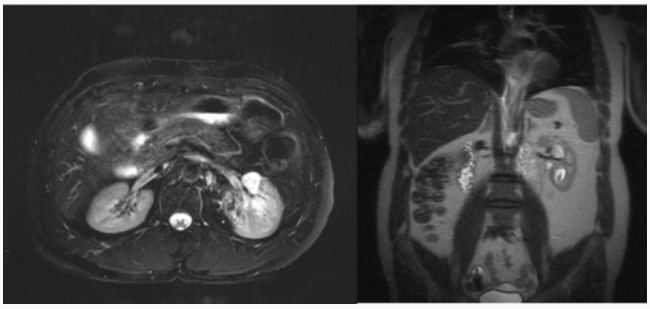

Figure 1 and 2: Renal MRI images in June 2021 before surgical resection of metachronous solitary fibrous tumors.

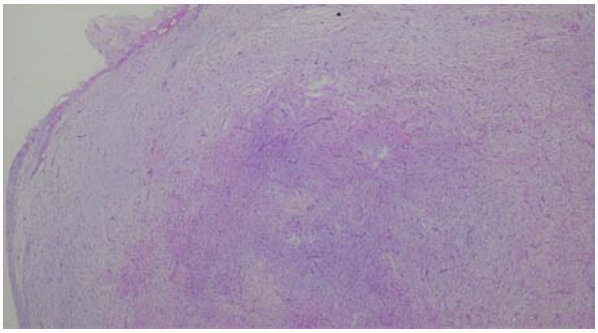

Figure 3: HE 20x: Unencapsulated and well circumscribed lesion.

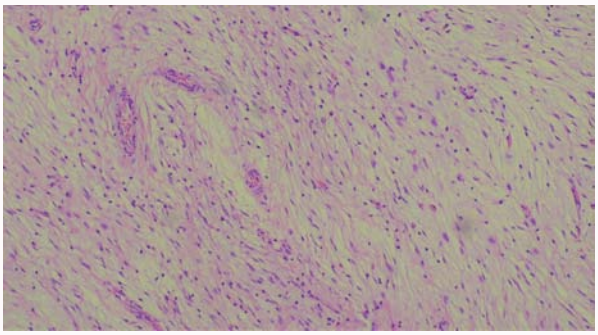

Figure 4: HE 100x: Fusiform spindle cells with indistinct borders, dilated thin wall vessels and stroma with prominent myxoid.

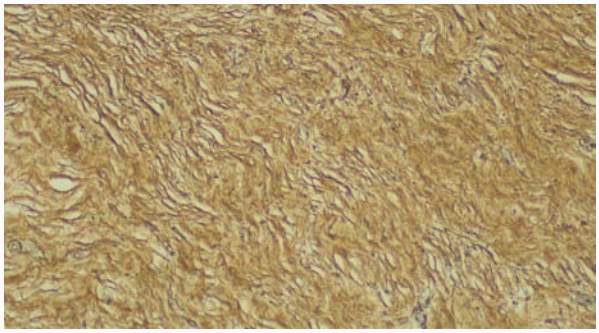

Figure 5: Immunohistochemistry 100x: Strong and diffuse staining for CD34.

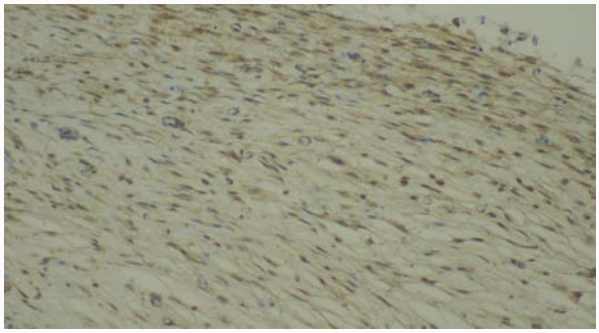

Figure 6: Immunohistochemistry 200x: Diffuse nuclear staining for STAT-6.

vimentin, CD34 and Bcl-2. There was very slight positivity for CD99. It was verified absence of positivity for c-kit, DOG1, smooth muscle actin, desmin, protein S100, and β-catenin. The expression of Ki67, the proliferative index, was around 5%.

The patient underwent imaging control every 6 months (abdominal ultrasound or CT). On May 2021, in an abdominal CT scan it was identified a new 26 mm left kidney nodule, with low uptake of contrast, not present in the previous exam. Consequently, the patient performed a renal MRI in June 2021 that revealed in the middle third of the left kidney, in an anterior location, a cortical nodule measuring 24 mm × 21 mm × 21 mm compatible with RCC (Figure 1, 2). In September the patient underwent open mass excision. Intraoperatively, two suspicious hilar nodules were identified, a 2.4 cm lesion identified in the CT and MRI scans and a 1 cm independent lesion. The resections were macroscopically complete.

Both nodular lesions had overlapping morphological characteristics, with expansive growth and low cellularity. It was observed a neoplasm formed mainly by monotonous spindle cells, but cells with a vaguely histiocytic morphology with oval nuclei were also identified. No areas of necrosis, mitotic figures or cytological atypia were identified (Figure 3, 4). Immunohistochemical techniques revealed strong and diffuse positivity for CD34 (Figure 5) and nuclear staining with weak to moderate intensity for STAT-6 (Figure 6). The definitive diagnosis was metachronous SFTs (two lesions) with 1.2 and 2.4 cm in the left kidney hilum, with apparently complete excision.

Discussion

SFT clinical symptoms could be a palpable mass, a flank pain or hematuria but SFT is usually asymptomatic when it has a small size. Although, imaging is useful for the evaluation of renal tumors, ultrasonography, CT, and MRI features are not specific for the diagnosis of SFT [1]. Usually, the differential diagnosis of SFT is difficult, and in most cases the renal mass is interpreted as suspected of RCC and patients undergo surgery [2]. Diagnosis of kidney SFT is mainly based on the histopathological and immunohistochemical findings. SFT is a neoplasm characterized by the proliferation of spindle cells originating from fibroblasts and mesenchymal cells. On gross examination, tumors are usually tan-gray, homogenous, lobulated, well-circumscribed, and most often involve the renal capsule. Microscopically, spindle cells are surrounded by thick, keloid-like collagen bands with patternless arrangements, sometimes with predominant myxoid changes. It is currently recognized to originate from CD34-positive dendritic interstitial cells and therefore show staining for this immunohistochemical marker (90% to 95% of cases). Immunohistochemical staining with Bcl-2, vimentin, and CD99 can also aid in the diagnosis of SFT. Focal and limited reactivity for S100 protein, cytokeratins and/or desmin had also occasionally been reported [3]. The STAT-6 immunohistochemistry constitutes nowadays the most useful diagnostic tool in daily practice, since it is

a surrogate marker for the NAB2-STAT6 gene fusion, present in the vast majority of SFT [4].

It is estimated that 10% of extrapleural SFTs will recur and/or metastasize [5]. SFT are categorized as having intermediate biological behavior, locally aggressive and rarely metastasizing according to the 2020 WHO classification. The diagnostic criteria for malignant SFT were proposed by England in 1989: Mitotic count >4/10 high-power fields; diversity of shapes of cells; many focal points in cells; and necrosis of some cells [6]. In the latest classification of SFTs formulated by WHO, the above criteria were still used for the diagnosis of extrapleural malignant SFT [7]. Malignant transformation of SFT may result from 2 physiopathologic mechanisms according to literature: *De novo* occurrence and dedifferentiation from preexisting benign SFT [8]. With malignant transformation, SFT may lose CD34 expression [9].

New studies proposed multiple prognostic factors for SFTs, but the lack of large-scale studies worldwide fall short to assess their significance. A recent study in a multicenter cohort from the French Sarcoma Group database developed a risk stratification model (prognostic groups for overall survival and recurrence) [10]. The core idea of risk assessment models is the standardization of the treatment protocol by allocating patients in different subgroups in which therapeutic lines depend on the calculated risk for local and metastatic recurrences.

There are very few series of patients with SFT and much of the literature only describes case reports. Fu et al. [11] retrospectively analyzed 10 adult patients with renal SFT. Pathological diagnosis revealed that the tumors in 8 patients were benign, while those in the other 2 patients were malignant. No recurrence occurred in a mean follow-up of 47.3 ± 21.5 months. Cheung et al. [12] described a 49-year-old female with a left renal SFT (16 cm × 11 cm × 9 cm) who after undergoing a nephrectomy, presented 8 years later with a metachronous contralateral kidney SFT (exophytic nodule, 4.3 cm, in the lower pole submitted to partial nephrectomy). Sfoungaristos et al. [13] report a case of a 72-year-old male with a large retroperitoneal SFT recurrence (12 cm × 11 cm) 3 years after nephrectomy to excise the primary tumor. The pathologic diagnosis of the nephrectomy specimen was SFT with negative surgical margins. The patient follows a strict surveillance, being well without evidence of recurrence or metastasis 9 months after the re-operation. Sammoud et al. [8] described a case of SFT with two local recurrences in a 53-year-old man. In the second time of local recurrence the patient had lung metastasis. As in the cases described in the literature, in our case there was a recurrence of the disease even though the first resection specimen had negative margins.

The follow-up time for patients with SFT is not defined. Salas et al. [10] concluded that recurrence increased between 10 and 20 years, suggesting that long-term monitoring is useful. In our case, the recurrence was after 3 years of the primary tumor diagnosis. The patient will maintain imaging control, but there is no defined protocol.

Conclusion

In our case report we describe a young man with diagnosis of a very rare case of metachronous SFTs in the same kidney of the first resection. Diagnosis was based on the histopathological and immunohistochemical findings. The evaluation of SFT mainly comes from case reports. Future large population studies are required to enrich our understanding of this tumor type and its behavior and to improve its diagnosis, treatment, and follow-up.

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