Bilateral renal involvement by solitary fibrous tumor — Report of a case in the post-WHO/2016 era

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Oncology

1. Introduction

Solitary fibrous tumors (SFT) are uncommon neoplasms that typically arise from the pleura and have unpredictable behavior. Extrapleural SFTs are rare in the genitourinary tract, where they more often involve the kidney. Definition between primary or metastatic SFT is critical for patient management, impacting prognosis and treatment. SFTs share many similarities to hemangiopericytoma (HPC), but recent molecular improvements place most HPC as cellular variants of SFT. We describe the case of bilateral renal masses in a young male, exemplifying why such a shift in terminology is important, especially when molecular knowledge can be translated into accessible diagnostic tools.

In 1870 Wagner described a fibrous or reticular tumor (“retikuliert tuberkel”) with malignant features involving the pleura. The term “solitary” was proposed later to differentiate from mesothelioma. SFT and HPC have since been separate entities by the WHO Classifications of several organs. However, over time, the vascular pattern characteristic of HPCs was found in 15% of all soft-tissue tumors, suggesting that this feature represents a histologic pattern instead of a clinicopathologic entity. Furthermore, recent research showed that these lesions share the chromosomal fusion NAB2-STAT6, leading the following WHO editions to incorporate most HPC as part of the spectrum of SFT (Fig. 1).

2. Case presentation

A 39 year-old male complained of abdominal discomfort and enlargement of the right scrotum for 6 months, with no urinary symptoms or weight loss. Initial investigation at the community health facility diagnosed varicocele and indicated surgery. Palpation of the abdomen under anesthesia revealed a large mobile mass (left hypochondrium to iliac region) and a transcutaneous biopsy was preferred. Histology revealed a monotonous spindle cell proliferation, with bland nuclear features and low proliferation rate, in a collagenous background, with no necrosis and areas of CD34 immunoexpression, suggestive of SFT. He then reported to us for a second opinion.

Aside from surgical excision of a meningioma 7 years ago, there was no relevant history. He had discrete anemia (hemoglobin: 11.7 mg/dL; hematocrit: 33%) and preserved renal function. Abdominal computed tomography (CT) revealed nodular heterogeneous confluent masses in the retroperitoneum, connected to the left kidney, and smaller similar lesions in the contralateral kidney (Fig. 2). Radical left nephrectomy was performed. There were no peritoneal implants or visible adenopathy.

Cross evaluation (Fig. 2) evidenced an exophytic lesion invading the renal parenchyma and hilum. Histology (Fig. 3) showed variable cellularity of spindled to ovoid cells in a collagenous stroma that produced areas of fibrous background.
Hemangiopericytoma-like vasculature and ectatic vessels were predominant. Despite cystic degeneration, necrosis was absent. Mitotic index was 3.8/10HPF. Immunohistochemistry was diffusely positive for CD99 and BCL2, and focally positive for CD34. The lesion also showed nuclear expression of STAT6 - a surrogate for NAB2-STAT6 fusion. Cytokeratins, WT1, FLI-1, SMA and S-100 were negative, ruling out sarcomatoid carcinoma, synovial sarcoma, Wilms tumor, rhabdomyosarcoma, leiomyosarcoma and melanoma. A diagnosis of fibroblastic lesion of low malignant potential in the HPC/SFT spectrum was made.

One month post-operatively, ultrasound revealed enlargement of the nodules in the remaining kidney. Given such rapid progression, a metastatic nature was suspected and the previous brain tumor was retrieved for pathology review. The meningeal lesion, a metastatic nature was suspected and the previous brain metastasis allowed for observation of larger series, some argue that the name metastatic SFT carries prognostic information by itself. At least in the HPC/SFT spectrum was made.

Fig. 2. Anatomy of left kidney lesion. Grossly, the tumor was predominantly exophytic with invasion of renal parenchyma and hilum. Residual kidney is highlighted in red. Inset: correlation with coronal CT view. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

Clinical presentation of renal SFT is non-specific: pain can present on larger masses and hematuria may indicate invasion of the pyelocalicial system. While the lung is the most frequent metastatic site of primary renal SFT, the kidneys have been reported as a frequent metastatic site for meningeal HPC/SFT. Except for few isolated reports, data is imprecise with regards to how often SFT metastasizes to the parenchyma or retroperitoneal adipose tissue. We believe parenchymal invasion should raise the possibility of a metastatic nature. Most tumors are indolent, whereas aggressive behavior has been reported in histologically benign appearing lesions, therefore grading schemes are imprecise. While a uniform nomenclature allows for observation of larger series, some argue that the name HPC carries prognostic information by itself. At least in the meninges, former-HPCs have higher metastatic potential than classic SFTs, frequently as late recurrences, like our case. We found three previous reports of meningeal HPC/SFT, all males between 37 and 58 years old, progressing after 10–14 years of primary excision.

The fibroblastic variant of meningioma is particularly important in the differential of HPC/SFT and histological identification of typical meningothelial areas and calcified psammoma bodies aid distinction. Reticulin stain and STAT6 immunohistochemistry were helpful in our experience. Retroperitoneal dedifferentiated

3. Discussion

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liposarcomas should be in the differential of “fat containing” SFTs since they may show STAT6 positivity. Sarcomatoid renal cell carcinoma should also be excluded, since most extrapleural SFTs express PAX8.

4. Conclusion

Advanced and bilateral renal SFTs should prompt investigation of extra-renal primary sites. The NAB2-STAT6 fusion has been shown to be specific for SFTs, encouraging the possibility of targeted therapy. STAT6 expression is a surrogate marker that enhances contemporary diagnostic accuracy.

Consent

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Conflicts of interest

All the authors declare no conflict of interest regarding this scientific communication.

Appendix A. Supplementary data

Supplementary data related to this article can be found at https://doi.org/10.1016/j.eucr.2017.12.003.

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