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Solitary fibrous tumor of the kidney: a report of two cases

Tumor fibroso solitário do rim: relato de dois casos

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ABSTRACT

Solitary fibrous tumor (SFT) is a rare mesenchymal neoplasm, composed of monotonous spindle cells, intermingled with collagen fibers. Kidney location is sporadic, with few reports in literature. We present two cases of 40- and 48-year-old males, one with incidental detection of a peripheral mass; another with hilar lesion perceived following investigation of hematuria. Both revealed a spindle-cell proliferation, without mitosis and necrosis. Immunohistochemical studies: positivity for CD34, CD99 and B-cell lymphoma 2 (Bcl-2). No sign of disease were evident nine and five months after surgery, respectively. SFT is mostly benign, but can show malignant behavior. Morphologic and immunohistochemical characteristics are essential for diagnosis.

Key words: solitary fibrous tumors; kidney neoplasms; immunohistochemistry.

RESUMO

Tumor fibroso solitário (TFS) é uma neoplasia mesenquimatosa rara composta por células fusiformes monótonas, intercaladas por fibras de colágeno. Localização renal é incomum, com escassos casos descritos. Relatamos dois casos do gênero masculino, com 40 e 48 anos, um com detecção incidental de massa periférica; outro com lesão hilar descoberta na sequência de investigação por hematúria. Ambos revelaram proliferação fusocelular sem mitoses nem necrose. Imuno-histoquímica: positividade para CD34, CD99 e linfoma de células B2 (Bcl-2). Não há evidência de recidiva nove e cinco meses após cirurgia, respetivamente. TFS é maioritariamente benigno, mas pode revelar comportamento agressivo. Características morfológicas e imuno-histoquímicas são essenciais para o diagnóstico.

Unitermos: tumor fibroso solitário pleural; neoplasias renais; imuno-histoquímica.

RESUMEN

Tumor fibroso solitario (TFS) es una neoplasia mesenquimal infrecuente de células fusiformes monótonas, separadas por bandas de colágeno. Localización renal es poco habitual, con escasos casos descriptos. Reportamos dos casos de pacientes masculinos, de 40 e 48 años, el uno con detección incidental de masa periférica; el otro con lesión hilar descubierta en la investigación de hematuria. Los dos revelaron proliferación fusocelular sin necrosis ni mitosis. Inmunohistoquímica: positividad para CD34, CD99 y linfoma de células B2 (Bcl-2). No hay evidencia de recidiva a los nueve y cinco meses de la cirugía, respectivamente. TFS es, por lo general, benigno, pero puede revelar comportamiento agresivo. Características morfológicas y inmunohistoquímicas son esenciales para el diagnóstico.

Palabras clave: tumor fibroso solitario pleural; neoplasias renales; inmunohistoquímica.

INTRODUCTION

A solitary fibrous tumor (SFT) is a spindle-cell neoplasm, of mesenchymal origin, normally occurring in the pleura that shows a hemangiopericytoma-like vascular pattern⁽¹⁾. Extraleural locations may be registered, but SFTs of the kidney are extremely rare, with only 46 cases reported⁽²⁾. The origin of these tumors is not well established, with some authors stating that they arise from the capsule, interstitial tissues or peripelvic connective tissue⁽³⁾. Morphological and immunohistochemical proprieties are essential for the differential diagnosis with renal sarcomas, gastrointestinal stromal tumors and benign peripheral nerve sheet tumor⁽⁴⁾.

MATERIAL AND METHODS

Histology evaluation

Examination was performed on hematoxylin and eosin (HE)-stained slides observed in light microscope (Nikon Eclipse 50i), and images were obtained using a Nikon-Digital Sight DS-Fi1 camera.

Ancillary techniques/immunohistochemistry

Studies were performed on one representative block of the lesion, resorting to avidin-biotin-peroxidase complex method and performed on Ventana Bench Mark Platform ULTRA IHC/ISH using the following antibodies: AE1/3 (AE1/3, Dako, USA); CD10 (SP67, Ventana, USA); CD34 (QBEND/10, Ventana, USA); CD99 (013, Ventana, USA); CD117 (9.7, Ventana, USA); B-cell lymphoma 2 (Bcl-2) (124, Ventana, USA); S-100 (4C4.9, Ventana, USA); paired-box gene 8 (PAX8) (MRQ-50, Ventana, USA) and alpha-smooth actin (1A4, Ventana, USA).

CASE 1

Clinical data

Forty-year-old man subjected to abdominal sonographic study for symptoms of biliary colic. Gallstones were not detected, but a peripheral right renal tumor was visualized and characterized on computed tomography (CT) scan (**Figure 1A**). Radiology studies did not show other lesions, namely pleural tumors. Partial nephrectomy was performed.

Pathologic findings

Gross examination revealed a well demarcated and brownish lesion with 3.2 cm of diameter (**Figure 1B**). Histologically the lesion was densely cellular, well vascularized, composed of elongated cells, with little atypia, low mitotic activity and no necrosis (**Figure 1C**). Among the tumor cells there were hyaline collagen fibers. Immunohistochemistry revealed intense and diffuse positivity for Bcl-2, CD34, and CD99, and negativity for CD10 (**Figure 1D**). Tumor was also negative for AE1/3 and PAX8. The patient is well and free of disease nine months later.

CASE 2

Clinical data

Forty-eight-year-old man, with no relevant personal background, referred to the emergency service for hematuria.

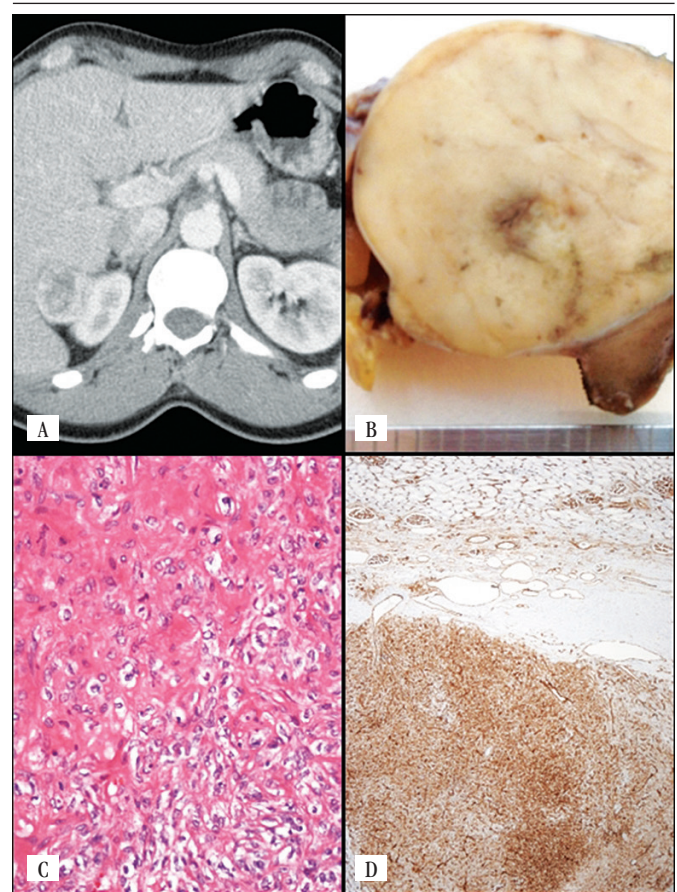


FIGURE 1 – A) peripheral right tumor on CT scan; B) gross examination revealed a brown and capsulated lesion; C) histologically it was composed of elongated cells, with little atypia, intercalated by collagen bundles; D) the tumor cells were positive for CD34
CT: computed tomography.

Sonography showed a central lesion with 8 cm in the right kidney. Preoperative staging resorting to thoraco-abdominal-pelvic CT scan did not show other tumoral lesions. Urinary cytology revealed neoplastic cells, and a nephrectomy was performed.

Pathologic findings

On gross examination there was a renal hilar lesion with 8 cm that compressed the renal pelvis, with a whitish and fascicular cut surface (**Figure 2A**). Histological evaluation showed spindle-cell proliferation, with scanty cytoplasm with elongated nuclei and slight atypia, intercalated by hyaline collagen bundles (**Figure 2B**). The lesion was expansive, densely cellular and vascularized, with no necrosis and mild mitotic activity (3/10HPF). Immunohistochemistry evaluation showed positivity for CD34, CD99 (**Figure 2C**), Bcl-2 (**Figure 2D**), and negativity for AE1/3, PAX8, CD10, CD117, S100 and alpha-smooth actin. The patient is doing well, with no signs of tumor recurrence five months later.

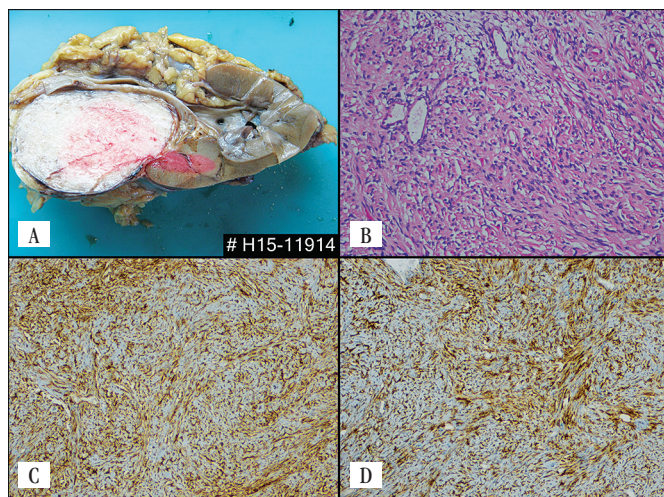


FIGURE 2 – A) gross inspection showed a white and fascicular lesion compressing the renal pelvis; B) microscopically it was well vascularized and composed of slightly atypical spindle cells (HE, 200 \times); C) the tumor cells were positive for CD99, 100 \times ; D) for Bcl-2, 100 \times

HE: hematoxylin and eosin; Bcl-2: B-cell lymphoma 2.

DISCUSSION

SFTs are slow-growing tumors⁽⁵⁾, of mesenchymal origin, with no sex predominance and a mean age of 52 years (range 28-83)⁽⁶⁾. They are normally detected in the pleura, but

extrapleural presentation may arise, affecting any part of the body, including genitourinary tract⁽¹⁾. SFT in the kidney is extremely rare, with the first case described in 1996⁽⁷⁾.

Clinically they can present as an incidental finding on radiology or as a palpable mass, and symptoms, if any, are commonly abdominal pain and hematuria⁽²⁾. On gross examination, SFTs are well circumscribed, pseudoencapsulated and solid, with a gray-white to tan appearance^(2, 4, 8). Histologically, SFTs are composed of bland spindle cells, organized in short fascicles or in a storiform pattern, intercalated by bundles of collagen fibers⁽⁸⁾. Due to the extensive range of differential diagnosis of mesenchymal lesions of the kidney, immunohistochemistry plays a pivotal role, revealing positivity in SFT for CD34, CD99 and Bcl-2^(1, 5, 8); recently a new antibody was released with a high sensitivity for SFT diagnosis – signal transducer and activator of transcription 6 (STAT6)⁽⁸⁾. Ultrastructural characteristics include protruding Golgi apparatus, dispersed intermediate filaments, irregular nuclei, and unfixed number of mitochondria⁽⁶⁾.

Nephrectomy is the treatment of choice⁽¹⁾, with most cases of SFT being benign and having a favorable prognosis; a minor number may present malignant transformation and consequently poorer outcome⁽⁹⁾. Despite benign histology, SFT are regarded as an “Intermediate malignant, rarely metastasizing neoplasm”⁽¹⁰⁾, so a close and long-term follow-up is recommended for all patients in order to determine clinical behavior. Some authors have developed prognostic scores in order to predict malignancy based on cellular density, necrosis and prominent mitotic activity (over four mitosis/10 high-power-field)⁽¹¹⁾, tumor size (more than 10 cm) and mindbomb E3 ubiquitin protein ligase 1 (MIB-1) proliferative index^(12, 13) and even gender (male) and age (over 55 years old)⁽¹⁴⁾; however the scores are still far from ideal and are developed for pleural SFT, lacking validation for extrapleural SFT – the cases of malignant renal SFTs reported in the literature did not show tumor recurrence or metastasis^(9, 10).

In conclusion, renal SFT is a rare tumor, which pathologist needs to be aware of in order to provide the correct diagnosis based on morphology and ancillary studies. The stratification of renal SFTs according to their clinical behavior is not a reality at the moment. More studies are needed, namely molecular biology classification and the development of large cohorts of patients with long-term follow-up.

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