SOLITARY FIBROUS TUMOR OF THE KIDNEY

P. Tritschler¹, B. Coulier¹, I. Gielen²

Solitary fibrous tumor (SFT) is an unusual spindle cell neoplasm rarely described in the kidney. Usually occurring in the pleura, it has also been described in various extrapleural sites. We report a rare case of SFT of the kidney fortuitously found in a 55-year-old patient. The imaging features are illustrated. The definite diagnosis was made through histological and immunohistochemical study after radical nephrectomy.

Key-word: Kidney, neoplasms, diagnosis.

Solitary fibrous tumor (SFT) is a rare lesion representing 2% of all soft tissue tumors (1). Described for the first time in the pleura by Klemperer and Rabin in 1931 (2), SFTs can also rarely occur in intra- or extrathoracic site. Imaging features on SFT are mostly non specific and essentially show a highly vascularised tumor. The definite diagnosis can only be made through histological and immunohistochemical study. We report a new case of solitary fibrous tumor of the kidneys fortuitously found in a 55-year-old patient.

Case report

A 55-year-old patient was referred to our radiology department with a few weeks history of right upper quadrant pain.

Nausea, vomiting, diarrhea or hematuria were absent. Abdominal physical examination and blood tests were normal. The patient had no previous medical or surgical history.

Abdominal ultrasound demonstrated uncomplicated gallstones but extended examination revealed a central mass in the right kidney (Fig. 1A,C). There was no pyelectasy. This well circumscribed solid mass was developing in the right renal sinus (Fig. 1C, D, E). The tumor appeared separated from the renal cortex. Areas of parenchyma atrophy of the renal cortex were visible adjacent to the tumor. No delay of renal excretion was seen.

The arterial phase of contrast enhanced mdCT confirmed a 5 x 7 cm hypercellular solid tumor made by spindles cells with rounded or oval nucleus, arranging in all directions with formation of vortices hemangiopericytoma like (Fig. 2B). The cytoplasm was moderately abundant with precise boundaries. No mitotic activity nor atypia were found. This tumor invades the hilar fatty tissue (Fig. 2A).

Immunohistochemical examination revealed a positivity on numerous cells for the CD99, Bcl2, CD34, vimentine and a minority of cells for the actine. Desmine and EMA are negative. The Ki67 nuclear proliferation index is low 3%.

A diagnosis of solitary fibrous tumor of the kidney is then evoked.

Discussion

Solitary fibroma tumors (SFTs) are mesenchymal tumors, now considered a variant of hemangiopericytomas (3). SFTs were described for the first time in the pleura but they can affect all the body at any location (4). Cases have been described in upper respiratory tract, lung, nasal cavity, parasanal sinuses, orbits, mediastinum, salivary glands, breast, meninges, liver and urogenital organs (3). Thus, there are intra and extra-thoracic locations but most of cases are intra-thoracic (5).

Intra-thoracic and extra-thoracic SFTs are often considered benign neoplasms but the incidence of aggressive forms manifested by local invasion or distant metastases (6) is about 10-15% for intra-thoracic SFTs and more than 10% for extra-thoracic SFTs (7). Therefore all patients with SFT need to be on long-term follow-up (3).

SFTs of kidney is very rare with only about 40 cases described in the literature (1). The origin of most cases kidney SFTs is difficult to determine and may originate from the capsule, interstitial tissues, or peripelvic connective tissues (3). The average age of presentation is 51 years (1), with a small female predilection (6) but usually the addition of intra and extra-thoracic forms revealed no preference, male and female being equally affected.

Clinical examination can show a palpable mass, a flank pain and often hematuria. Blood test are non contributive. Sometimes hypoglycaemia is described for extra or intra thoracic forms but never for kidney SFTs (7). The diagnosis is often delayed because these tumor are usually asymptomatic when they have a small size.

Histopathological nature of SFTs is distinguished by a hypercellular stroma of bland spindles cells with a patternless architecture. Typical immunohistochemical characteristics are a high positivity for CD34 – regarded as an indispensable finding in the diagnosis (3) and to a lesser extent for Bcl2, vimentin and CD99 (1, 8).

The differential diagnosis is large and includes highly vascularized tumors as renal cell carcinoma, sarcomatoid renal cell carcinoma, angiomylipoma, fibroma, fibrosarcoma, and leiomyosarcoma (3).

The therapeutic management is surgery with a high survival rate approaching 100% at 5 years (1).
Finally the final diagnosis is always based on histopathology and immunohistochemical study. Radiologists could only consider SFT in the differential diagnosis of a renal mass if the lesion is a large, situated in the renal sinus with lobulated margins and highly vascularized by large vessels (4).
Fig. 2. – A. Photomicrograph (hematoxylin-eosin stain) shows at low power field view (×10) a rather well circumscribed hypercellular tumor (white star) invades the sinusal fat (black star). B. Photomicrograph (hematoxylin-eosin stain) shows at higher power field view (×40) the hypercellular tumor appears composed of numerous bland uniform spindle cells with a patternless architecture.

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