Benign Fibromatous Tumor (Fibroma) of the Kidney: A Case Report

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Summary

Benign fibromatous tumor (fibroma) of the urogenital tract is a distinctive pathologic entity occurring in the testis, paratesticular structures and renal peripelvis. We report a well-circumscribed tumor replacing the cortex and the medulla of more than half of the upper kidney, radiographically thought to be renal cell carcinoma. Histologically, the tumor was characterized by a variable cellularity and was composed of bland spindle-shaped cells loosely dispersed in a fibromyxoid to densely fibrous stroma in which calcifications and chronic inflammation were not observed. Immunocytochemistry showed that cells were positive for vimentin and, only focally, stained positive for desmin and α-smooth muscle actin. Differential diagnosis included a wide spectrum of benign and malignant spindle cell tumors. The clinicopathologic features were consistent with benign fibromatous tumor (fibroma) of the kidney.

To our knowledge, this is the first case of benign fibromatous tumor (fibroma) of the kidney reported in the English literature.

Key words: Fibromatous tumor – Fibroma – Kidney

Introduction

Benign fibro-proliferative lesions of the urogenital tract, forming uninodular or multinodular firm masses, have been reported in the epididymis, spermatic cord, paratesticular structures, testis and renal peripelvis under a variety of terms including fibrous tumor, fibroma and fibrous pseudotumor [8, 9, 14]. Recently, on the basis of clinical, gross, histologic and immunohistochemical findings, a distinction between true fibrous tumors, also described by the term “fibromas,” and fibrous pseudotumors has been proposed [9].

We report a unique case of benign fibromatous tumor (fibroma) of the kidney, with gross, histologic and immunohistochemical findings similar to those previously described in the fibroma of the testis, paratesticular soft tissues and renal peripelvic connective tissue [8, 9, 14]. The tumor presented as a large mass replacing the cortex and medulla of more than half of the upper kidney, clinically producing an intermittent gross hematuria in a 74-year-old woman. The peculiar clinicopathologic findings are discussed.

Case Report

A 74-year-old woman presented with a one-week history of painless, intermittent gross hematuria. Neither previous trauma nor surgery was reported in the patient’s history. The medical history and review of systems were no contributory factors. Physical examination was within normal limits. Urine cytology, performed on several occasions, was negative. Ultrasonography of the abdomen and pelvis revealed a solid echogenic mass in the upper pole of the left kidney. Computerized tomography confirmed the presence of an 8 cm heterogeneous, solid, spherical mass which was considered most consistent with a renal cell carcinoma. Neither local invasion nor lymphadenopathy was identified. Renal arteriography was not performed. The patient underwent a left radical nephrectomy and paracaval lymph node dissection. Two years after surgery, the patient is well with no evidence of tumor.

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Pathologic Findings

On gross examination, more than half of the upper kidney was replaced by a well-circumscribed, spherically shaped mass, protruding from the surface with a smooth and pushing border, and covered by a smooth, glistening capsule continuous with the renal capsule. On cut section, a firm, white, sharply circumscribed mass, measuring 8 × 7 × 5.5 cm, completely replaced the cortex and the medulla and compressed the pelvis (Fig. 1). The cut surface also contained some cystic cavities of variable size. Grossly, the kidney was unremarkable. The resected kidney was immediately fixed in 10% buffered formalin. Multiple samples of the mass were taken and paraffin-embedded. Sections were cut and stained with hematoxylin and eosin, periodic acid-Schiff (PAS) staining (with or without diastase digestion), Masson's trichrome and with van Bieson for reticulin.

Histologically, the mass was composed of medium-sized, spindle-shaped cells loosely dispersed in a fibromyxoid to densely fibrous stroma with low vascularization (Fig. 2). Collagenous bands were chiefly arranged in a parallel manner. The cells had oval and normochromatic nuclei. Cellularity ranged from moderate to very low, especially in the more fibrotic stroma (Fig. 2). Cellular pleomorphism, nuclear atypia and mitotic figures were not observed. Necrosis, hemorrhage and calcification were absent. Lymphocytes were only rarely detected in some areas. Small groups of atrophic and cystically dilated tubular structures, lined with a single layer of cuboidal epithelium, were entrapped throughout the fibrous stroma (Fig. 2). Infiltrating borders into the adjacent renal parenchyma were not observed.

Fig. 1. A well-circumscribed, white mass is located and replaces the cortex and the medulla of more than the upper half of the kidney.

Immunocytochemistry (streptavidin-biotin immunoperoxidase), using a panel of antibodies, including vimentin (Dako, dilution 1:100), α-smooth muscle actin (Dako, dilution 1:200), desmin (Dako, dilution 1:500), S-100 protein (Dako, dilution 1:300), CD34 (Immunotech; prediluted), pan-cytokeratin (Dako, prediluted) and epithelial membrane antigen (EMA) (Dako, dilution 1:150) showed that spindle-shaped cells were diffusely positive to vimentin and, only focally, to desmin and α-smooth muscle actin (Fig. 2C).

Discussion

We report a unique case of benign fibromatous tumor of the kidney which, on the basis of gross histologic and immunocytochemical examinations, is similar to the fibromatous tumor (fibroma) described in the testis, paratesticular soft tissues and peripelvic renal tissues [8, 9, 14]. Grossly, the tumor presented as a firm, white and well-circumscribed mass, completely replacing the cortex and medulla of more than half of the upper kidney. Histologically, the tumor was composed of a low to moderate number of spindle-shaped cells sparsely distributed in a fibromyxoid to densely fibrous stroma. Immunocytochemistry revealed that neoplastic cells were positive for vimentin and focally for desmin and α-smooth muscle actin, suggesting the fibroblast-myofibroblastic nature of the tumor. On the basis of gross, histologic and immunocytochemical findings [9, 14], the diagnosis of benign fibromatous tumor (fibroma) was made.

Differential diagnosis included benign (inflammatory myofibroblastic tumor, desmoid-type fibromatosis, leiomyoma, schwannoma/neurofibroma, perineurioma, benign fibrous histiocytoma, solitary fibrous tumor, adult mesoblastic nephroma and renal medullary fibroma) as well as malignant fibrous spindle cell tumors (low-grade fibromyxoid sarcoma).

The term “inflammatory myofibroblastic tumor” [2] usually refers to a wide spectrum of spindle cell lesions which, on the basis of their predominant histological features, are also described by several terms including inflammatory pseudotumor, pseudosarcomatous myofibroblastic proliferation and fibrous pseudotumor [2, 14]. Unlike our case, this category of spindle cell lesions usually presents as a multinodular or diffuse mass composed of a highly variable admixture of bland-looking (myo)fibroblast-like cells, myxoid to hyalinized stroma, conspicuous polymorphous inflammation (lymphocytes, plasma cells), calcifications and bone formation [2, 9, 14]. Desmoid-type fibromatosis is a fibroproliferative lesion which may mimic benign fibromatous tumor histologically. The absence of both infiltrating margins at the periphery of the tumor and the typical growth pattern, showing a formation of interlacing
Fig. 2. The tumor shows spindled hypocellular (A) and moderately cellular fibrous (B) stroma (H & E, ×80). (C) Spindle cells are focally stained with α-smooth muscle actin (×125). Atrophic renal tubules are entrapped throughout the fibrous stroma (A, C).
bundles and fascicles composed of uniform spindle-shaped cells with a parallel array [4, 11], were all features useful for excluding desmoid fibromatosis. Leiomyoma and schwannoma/neurofibroma are rarely neoplasms of the kidney [12, 17]. They are easily distinguishable from benign fibrous tumor because of their characteristic histologic and immunohistochemical findings which are identical to those of similar tumors seen at other body sites [12]. Perineurioma is a rare benign spindle cell tumor of the kidney occurring in both childhood and adult [10, 18]. Although this tumor may show a variable degree of cellularity and sclerosis, it is usually characterized by hypercellular areas containing EMA-positive fusiform cells arranged in a fascicular, lamellar, whorled or storiform pattern [10, 18]. Benign fibrous histiocytoma, a tumor rarely reported in the kidney, has rarely been reported [6, 7]. Unlike our case, solitary fibrous tumor exhibits a variety of growth patterns, including a storiform, hemangiopericytoma-like, angio-fibroma-like pattern and is strongly positive for CD34 [6, 7].

Mesoblastic nephroma is a renal tumor of early infancy which rarely occurs in adults [3]. Histologically, two distinct growth patterns, often mixed in the same tumor, are recognized: the classic and the cellular pattern. In adults, this tumor virtually always shows the classic patterns characterized by intersecting fascicles of spindle cells that infiltrate renal and perirenal tissues, entrapping glomeruli and tubules [3]. This fibromatosis-like growth pattern is missing in our case.

Renal medullary fibroma, also known under the term interstitial cell tumor, is a distinctive pathologic entity occurring as single or multiple, small white nodules located exclusively in the medulla and rarely exceeding a few millimeters in diameter [12]. The medullary location without extension toward the cortex is a peculiarity of this tumor, as confirmed by all cases of renal medullary fibromas, including the exceptionally large ones [15] reported in the more recent literature [13]. Although some reports of cortical fibromas have been recorded in the older literature, it is likely that these tumors, diagnosed without special stains, were not true fibromas [11]. Low-grade fibromyxoid sarcoma differs from benign fibrous tumor in that bland, innocuous-appearing fibroblastic cells are arranged in a swirling and whorled growth patterns [5].

Although in most cases of benign fibromatosus tumors of the urogenital tract, the testicular tunics [9, 14], gonadal stroma [9] and renal peripelvis connective tissues [8] are considered to be the site of origin of the tumor, in our case, with the large size of the tumor given, it was difficult to decide whether it has arisen from the renal capsule, renal parenchyma or renal pelvis. Nevertheless, we underline the possibility that benign fibrous tumor may present as a large corticomedullary mass replacing a large portion of the kidney. The large size, location of the tumor, clinical symptoms such as hematuria in an elderly patient, and the aspecific features, shown both ultrasonography and computed tomography, led us to make a preoperative diagnosis of renal cell carcinoma with subsequent radical nephrectomy. With the increasing use of radiologic imaging of the abdomen, an increased number of both incidental and symptomatic benign renal tumors is expected. Benign fibromatous tumor should be considered in the differential diagnosis of fibrous spindle cell tumor or tumor-like lesions of the kidney to ensure that the patient receives appropriate prognostic information and therapy.

References

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