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Renal Leiomyoma: A Case Report and Review of the Literature

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ABSTRACT

Leiomyoma is an uncommon tumor of the kidney. The authors report a rare case of renal leiomyoma in a 39-year-old male patient who presented with a right flank mass. Laparoscopic nephrectomy was performed. The histopathology and immunohistochemistry confirm the diagnosis of renal leiomyoma. The review of literature in the clinicoradiopathological correlation was illustrated.

Introduction

Leiomyoma is an uncommon primary renal tumor with smooth muscle differentiation.[1,2] In the genitourinary system, a urinary bladder is the most common site of occurrence. Primary renal leiomyoma is first described by Busse in 1899. Histopathological diagnosis is difficult, and always needs immunohistochemical stain. The purpose of this report is to illustrate a case of a renal leiomyoma and review the literature.

Case presentation

A 39-year-old male patient presented with a right flank mass for 3 months. Ultrasonogram showed an 11-cm solid-hypoechoic mass in the right kidney. Abdominal computed tomography (CT) scan showed a well-defined hyperattenuating solid mass (Fig. 1A, B, C). A laparoscopic right radical nephrectomy was performed. The right kidney weighed 454.4 g. The cut surface showed an 11 × 9.5 × 9.5 cm well-circumscribed unencapsulated firm whorling white mass (Fig. 1D). The histopathology revealed spindle cells arranged in intersecting fascicles (Fig. 2). The tumors demonstrated scanty cytoplasm, vesicular oval nuclei, and inconspicuous nucleoli. In immunohistochemical staining, diffuse expression of vimentin, muscle specific actin, smooth muscle actin, h-caldesmon, and desmin were detected in the spindle cells (Fig. 2). Immunohistochemical stains including HMB45, epithelial membrane antigen (EMA), cytokeratin AE1/AE3, CD10, renal cell carcinoma (RCC) antigen, PAX2, PAX8, sarcomeric actin, MyoD1, myogenin, WT1, S100, CD34, CD117, and Bcl2 were not expressed in the spindle cells. Ki67 proliferation index was 1%. Epstein-Barr virus (EBV) in situ hybridization was negative in the tumor cells. The final diagnosis was renal leiomyoma. No adjunctive treatment was given. He remains healthy at 6 years post-operation.

Discussion

Renal leiomyoma is a rare neoplasm that accounts for 4.2% to 5.2% of autopsy specimens and 0.29% of primary renal neoplasms.[3] It may originate from tunica media of the renal cortical vasculature, smooth muscle cells in the renal capsule, or muscularis of the renal pelvis, which can be grossly classified as subcapsular, capsular, or subpelvic type, respectively.[1,3] This tumor shows a female predilection (2:1).[2] The ages of patients range from 6 to 82 years with a mean age of 47 year-old.[2] Renal leiomyomas are found equally in both kidneys and are located more frequently in the lower pole (75%). Renal leiomyomas usually remain asymptomatic until they produce a mass effect. The frequently presenting symptoms are palpable flank mass, flank pain, and hematuria.[1–3]

The imaging procedures such as ultrasonography and CT scan may allow early recognition of renal tumors. The pattern approach depends on imaging findings and anatomic distribution. According to its benign nature, leiomyoma has a well-defined margin, without

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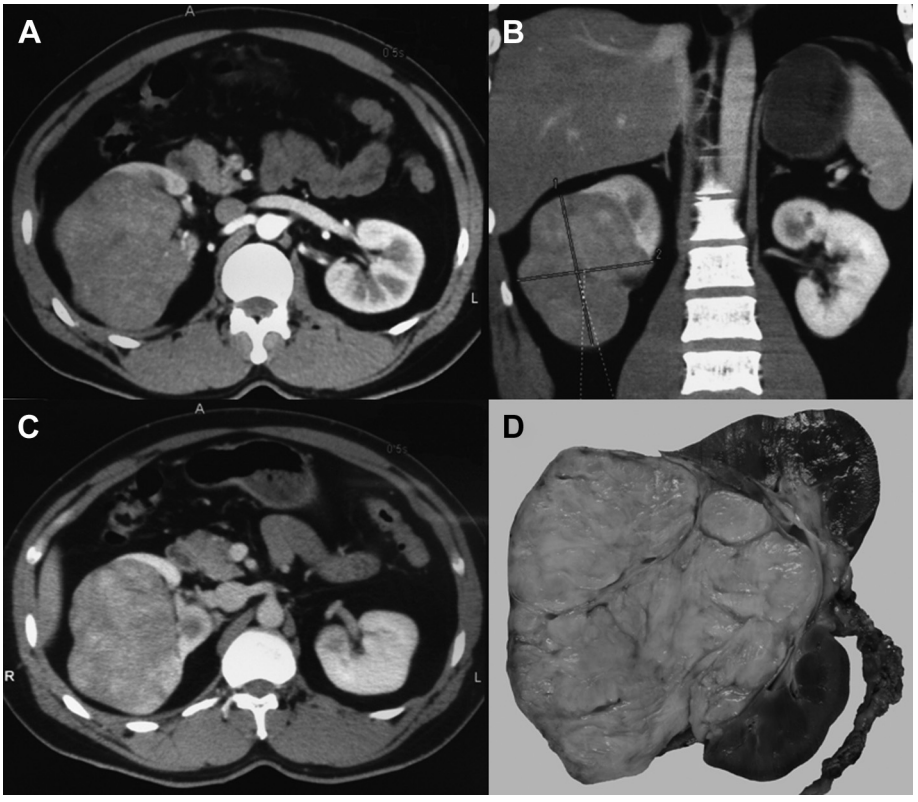

Figure 1. Post-contrasted CT scan on the axial and coronal views: There is a well-defined, exophytic mass at the interpolar region of the right kidney, and associated buckling of the renal cortex. After contrast material injection, the mass is hypodense compared to the renal cortex on the corticomedullary phase (A, B). It shows progressively enhancement on the later phase (C). The gross pathology shows a well-circumscribed unencapsulated firm whorling white mass (D).

signs of invasion into the surrounding parenchyma.[4] On non-contrast CT, leiomyoma is hyperdense compared to the renal parenchyma, with the density equals to muscle. After contrast material administration, leiomyoma shows a lower enhancement than the surrounding parenchyma at the corticomedullary phase.[4] It typically demonstrates relatively homogeneous enhancement.

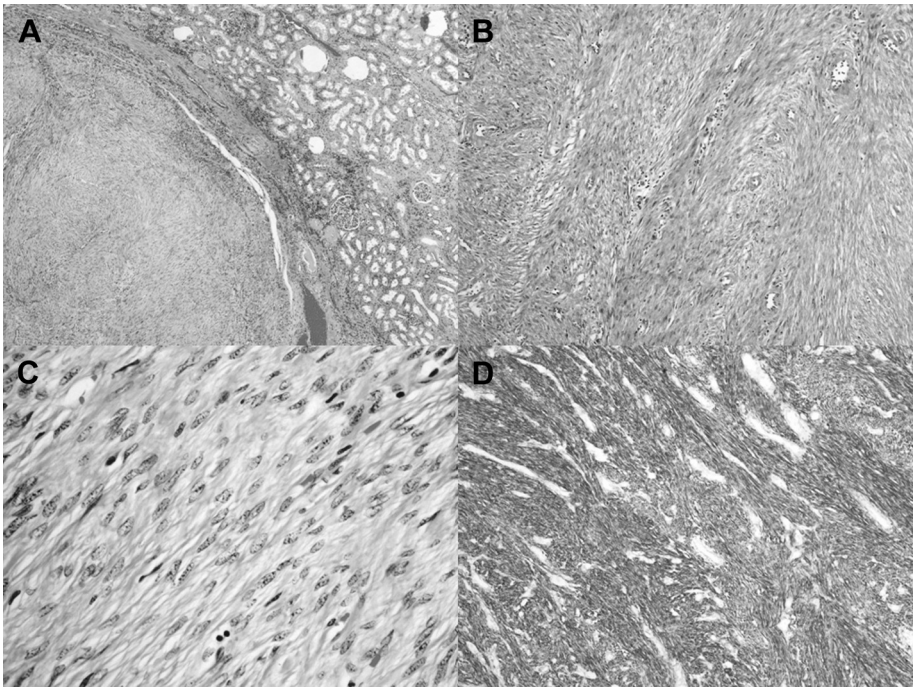

Figure 2. Presence of a well-circumscribed mass with proliferation of short spindle cells (A) arranged in intersecting fascicles (B) of monomorphic fusiform cells (C). The immunohistochemistry was positive for smooth muscle actin (D).

Large tumors may show heterogeneous areas of enhancement due to hemorrhage and cystic or myxoid degeneration.[4] Leiomyoma shows progressive and homogeneous enhancement on later phase of dynamic scanning.[4] These characteristics of enhancement are useful in distinguishing leiomyoma from the conventional RCC, which often shows marked, heterogeneous enhancement on the corticomedullary phase and decreased enhancement on the nephrographic phase.[4] However, chromophobe RCC has pattern of enhancement similar to leiomyoma.[4]

The macroscopic findings of leiomyoma are typically well-circumscribed, firm, bulging, whirling fibrous trabeculated white appearance. Focal calcification or cystic degeneration may be presented. Tumor size ranges vary from 0.5 to 57.5 cm with a mean size of 12.3 cm.[3] The largest tumor weighs of 37.2 kg.[1] The histopathology shows spindle cells arranged in intersecting fascicles. Immunohistologically, the tumor cells show positive immunostaining for vimentin and smooth muscle markers.[1,2] Moreover, the capsular leiomyoma frequently contains a population of cells strongly positive melanoma markers.[2]

The pathogenesis of leiomyoma remains enigmatic; although initially the tumors were thought to represent a hamartomatous process by the evidence of some renal leiomyomas have been associated with tuberous sclerosis and positive immunostaining for melanoma markers. Furthermore, there is a report in the literature documenting the combined losses of chromosomes 4, 6, 12 and 14, suggesting a different molecular pathogenetic mechanism, which has not been previously described in renal tumor.[5] Renal leiomyoma has clonal chromosomal aberrations that are acquired through somatic mutations. Moreover, a pediatric post-transplant patient previously published with renal leiomyoma had been associated with EBV infection.[2] The tumorigenesis of renal leiomyoma remains to be elucidated.

The differential diagnoses of renal leiomyoma include angiomyolipoma (AML) and sarcomatoid RCC. AML is a benign mesenchymal tumor composed of a variable proportion of adipose tissue, smooth muscle cells and abnormal thick-walled blood vessels. AML is classified in a family of hamartomatous lesions and is characterized by a co-expression of melanoma and smooth muscle markers. AML is excluded as it lacks the typical triphasic histopathology, and does not immunohistochemically express melanoma marker.[2] RCC may show sarcomatous feature consisting of spindle cells. Negative results of immunohistochemical stains for epithelial marker and RCC marker may be helpful in exclusion[1,2]

Leiomyomas generally behave in an indolent manner and generally do not recur after complete excision. Laparoscopic nephrectomy seems to be safe and advantageous to obtain a radical excision of the renal tumor. A correct histological diagnosis is essential to avoid detrimental treatments. Given the documented potential for local recurrence, we recommend careful evaluation of the patient to rule out recurrence as well as long-term follow-up.

Conclusion

A case of primary renal leiomyoma of the right kidney in a 39-year-old man clinically presenting with a right flank mass is reported. He underwent a laparoscopic right radical nephrectomy. Pathological examination of the right renal tumor characterizes macroscopically by a cortical mass and histologically by bland spindle cells. Immunohistological stainings are positive for vimentin, muscle specific actin, smooth muscle actin, h-caldesmon, and desmin. Renal leiomyoma is a rare mesenchymal tumor and should be included in the differential diagnosis of renal mass. Clinical and pathological features with a brief review of the relevant literature are discussed. Surgical resection remains the cornerstone of management and is recently facilitated by a laparoscopic approach.

Conflict of interest

The authors declare they have no conflicts of interest.

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