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Oncology Renal leiomyoma: An uncommon differential diagnosis of renal masses in pediatric age

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 A R T I C L E I N F O
 A B S T R A C T

 Keywords:
 Leiomyoma is a mesenchymal tumor which arise from any structure or organ containing smooth muscle, but is frequently seen in the female genital tract. Renal leiomyoma is extremely rare benign lesion with low incidence (1:1000), has been reported mostly in adults and very few cases have been described in the pediatric age-group. It is often asymptomatic and can be diagnosed when reaches large sizes. Hereby, we present a case of renal leiomyoma in a smaller size, in the pediatric age group.

Introduction

Renal leiomyoma is a benign mesenchymal tumor originating from smooth muscle cells, is extremely rare with low incidence (1:1000).¹ It has been reported mostly in adults and very few cases have been described in the pediatric age-group.^{1,2}

We present a case of renal leiomyoma in a smaller size, in the pediatric age.

Case presentation

A 17-year-old girl was referred to our department for further investigation due to a renal mass lesion that was incidentally detected by ultrasonography examination performed for mild abdominal pain in the outer center. On sonographic examination, a hypoechoic, wellcircumscribed solid mass lesion with a diameter of 3×2 cm was observed in the dorsolateral of the middle pole of the right kidney, and no significant vascularization was detected on color Doppler imaging (Fig. 1). On magnetic resonance imaging (MRI), the slightly lobule contoured and partially exophytic mass lesion measuring $29 \times 18 \times 30$ mm in size, showed mild high signal intensities on T1-weighted (T1W) images and low signal intensities on T2W images. Subtle heterogeneous enhancement of the tumor was revealed on the fat-suppressed dynamic contrast-enhanced images. On diffusion-weighted MRI, mild diffusion restriction was revealed (Fig. 2). No retroperitoneal lymphadenopathy was detected. Laboratory findings were normal. Histopathological diagnosis after partial nephrectomy was reported as renal leiomyoma. Microscopic findings revealed circumscribed lesion composed of spindle shaped cells with bland cytologic atypia in the kidney. There was no necrosis and low mitotic activity was observed. Immunohistochemical evaluation revealed that tumor cells were strongly positive for smooth muscle actin, desmin and negative for EMA, Melan-A, AMACR, CK7, CK19, Keratin, S100, CD34 (Fig. 3).

Discussion

Leiomyoma which is the most common benign tumor of the genital tract in women of reproductive age, may rarely arise from almost any part of the genitourinary system.^{1–5} Renal leiomyoma is extremely rare benign lesion originating from the tunica media layer of renal cortical vascular structures or smooth muscle cells in the renal capsule or renal pelvis, which can be usually classified as subcapsular, capsular, or subpelvic type, respectively.^{1–5}

Renal leiomyomas are usually detected incidentally in adults with a female predilection (2:1) and the average age has been reported as 47.^{1–5} These tumors constitute 1.5% of benign renal tumors and 0.3% of overall renal tumors.⁵ It has been reported that it is seen in the rate of 4.2–5.2% in autopsy series.^{1–5} To date, few cases have been described in pediatric age.^{1–3}

The tumor can be located equally in both kidneys and is more frequently seen (75%) in the lower pole of the kidney.^{1,3,4} Unlike the literature, the mass lesion was detected in the middle pole of the kidney

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Fig. 1. US examination revealed a hypoechogenic mass lesion in the middle pole of the right kidney (a), and significant flow was not encoded by Color Doppler imaging (b).



Fig. 2. On magnetic resonance imaging, the slightly lobule contoured and partially exophytic renal mass lesion showed mild high signal intensities on T1WI (a) and low signal intensities on T2WI (b). Subtle heterogeneous enhancement of the tumor (arrow) was shown on the pre (c) and postcontrast (d) enhanced fat-suppressed axial T1-weighted images. Diffusion-weighted MR imaging revealed mild diffusion restriction in the mass lesion (e–f).



Fig. 3. Benign spindle shaped smooth muscle cell proliferation beside the normal renal parenchyma (arrow) (H&E x200) (a). Actin positivity on the tumor cells (IHC x 100) (b). High power view of the tumor cells with regular oval nuclei and bland chromatin oval (H&E x400) (c).

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Renal leiomyoma usually remains asymptomatic until producing a mass effect. Symptomatic cases are presented with palpable mass (57%), abdominal/flank pain (53%) and 20% with microscopic or gross hematuria.^{1–5} In the literature, the average lesion size is reported as 12.3 cm.^{3,4} Unlike the literature, our case presented with abdominal pain, although the mass was small in size.

The differential diagnosis of renal leiomyoma includes fat-poor, smooth muscle predominant angiomyolipoma, fibroma. 1,4

Leiomyoma has a well-defined margin, without any invasion into the surrounding parenchyma. On ultrasound, a first-line diagnostic tool, it usually presents as a solid mass, but cystic changes can also be detected. On MRI, generally low signal intensity on T1-and T2-weighted images and lower enhancement than renal cortex at the corticomedullary phase, is detected. The contrast pattern may show heterogeneity due to hemorrhage and cystic or myxoid degeneration in large tumors. The capsular location of the mass lesion may clue the diagnosis.^{1–5} In addition, restricted diffusion may be detected in MRI due to different cellular and fibrous component contents.⁵ Definitive diagnosis of extrauterine leiomyomas requires histological examination of resected specimen, and confirmation of the immunohistochemical stain.^{1,2,4,5}

Renal leiomyoma is benign, nonmetastasizing tumor in pediatric age and complete excision of the lesion is the only therapeutic option. Partial nephrectomy can be performed for smaller lesions or moderate sized exophytic capsular or subcapsular lesions. The prognosis is good generally without recurrence after complete excision.¹

Conclusion

Renal leiomyoma is an extremely rare, benign nonmetastasizing tumor in children, that should be considered in the differential diagnosis of radiologically benign-looking solid renal mass lesions in guiding the surgical treatment.

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Declaration of competing interest

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