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Renal leiomyoma in pediatric age: a rare case report with review of the literature

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ABSTRACT

Renal leiomyoma (RL) is a rare benign tumor originating from muscle cells and has been rarely described in the pediatric age-group. A case of a large RL in an 11-year-old girl is reported with a literature review. The girl was admitted for abdominal pain due to mass effect. Contrast-enhanced CT showed a circumscribed low density mass (22 × 19 × 12 cm) arising from the right kidney. A right nephrectomy was performed. The mass was well defined and encapsulated. Only the histopathology and immunohistochemistry confirmed the diagnosis of RL. Even though partial nephrectomy may be possible, radical nephrectomy remains the recommended treatment for large leiomyoma.

1. Introduction

Renal leiomyomas are exceptionally rare benign tumors of the kidney, with smooth muscle differentiation, that occur with low incidence (1:1000) [1].

These tumors account for 1.5% of benign lesions and 0.29% of all treated renal tumors, with autopsy evidence in 4.2%–5.2% of cases [2,3].

Renal leiomyomas are found equally in both kidneys and are located more frequently in the lower pole (75%). There are less than 100 cases in the literature of leiomyoma confined to the kidney. Most cases are reported in adults, with a mean patient age of 47 year-old and a female predilection (2:1) [3–5]. Very few cases have been described in the pediatric age-group [6–9].

A case of a large renal leiomyoma in an 11-year- old girl is discussed along with a literature review relative to this type of the tumor in children.

2. Case report

An 11-year-old female patient was admitted to the Pediatric Surgery Unit, with abdominal pain and an abdominal mass of 3 months duration, following ongoing progressive growth. Family history was negative for familial cancer syndromes. No symptoms suggesting renal abnormality were present.

On physical examination, a huge hard mass occupying most of the abdomen, mainly from the right side, was palpable. The remainder of the exam was normal. General physical condition were stable (Heart Rate 76 BPM, Respiratory Rate 20 RPM, Saturation 98%, Temperature 36.7°, blood pressure 120/73 mmHg).

Laboratory studies showed the hemoglobin level, blood counts, blood urea nitrogen, and serum creatinine were in the reference range. Urinalysis was also normal. Given the size of the tumor (22 cm) occupying the whole abdominal cavity and pelvis raised suspicion regarding its origin. Tumor markers such as beta human chorionic gonadotropin and alpha fetoprotein were requested to exclude a germ cell tumor. These were negative.

A contrast enhanced computed tomography (CT)-scan of her abdomen showed a well circumscribed, low density mass, with areas of nodular high density demonstrating enhancement on contrast, ranging between 20 and 80 HU, filling the whole abdomen, and measuring 22 × 19 × 12 cm. The mass displaced the right kidney superolaterally, and appeared to be arising from its anterior parenchyma. No calcification or any hemorrhagic component was seen. The right pelvicalyceal system was distorted with

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hydronephrosis. The right ureter was compressed against the psoas muscle and inferior vena cava (IVC). The mass extended to the left, crossing the midline and ventral to the aorta and IVC, compressing this and displacing the small and large bowels to the left lumbar region and caudally to the pelvis. The left kidney appeared normal (Fig. 1).

A true-cut biopsy was performed; this showed benign skeletal muscle fibers and fibrillary connective tissue. Histopathologic abnormalities were not identified and the result was considered as non-diagnostic as no renal tissue was identified. This was followed by an open biopsy that showed, on frozen section, fibroconnective tissue only with no obvious histological abnormalities, and negative for S100 immunohistochemistry. Again no clear diagnosis was possible.

The patient was prepared for surgery. A median incision was made. The mass was found arising from the right kidney and extending from the 11th rib margin down to the pelvis. Transversely it was crossing the midline and pushing all the intestinal loops laterally. Careful dissection of the tumor followed, keeping the capsule intact. The dissection continued caudally where the right ureter was identified. This was sectioned and ligated at its distal part behind the bladder. Dissection was also continued at the upper pole of the kidney and the adrenal gland was spared. Dissection of the kidney hilum was difficult due to presence of oedema and enlarged lymph nodes, so the whole hilum was ligated with a double ligature and the mass was totally excised. The only lymph nodes found at the hilum were sent to pathology.

On gross examination, the mass was well circumscribed and encapsulated (weight 4 kg), Fig. 2. On microscopic examination, the mass was found to be composed of a relatively bland proliferation of smooth muscle cells. There were no features of any concern such as nuclear atypia, necrosis, increased cellularity or increased mitoses and the sampled lymph nodes were reactive with no evidence of malignancy (Fig. 3 Panels A–B). Immunohistochemical stains of the tumor cells were positive for desmin, smooth muscle actin, caldesmon and negative for WT-1, myogenin, and MyoD1 (Fig. 3, Panel C). The final diagnosis was a renal leiomyoma. No adjunctive treatment was given.

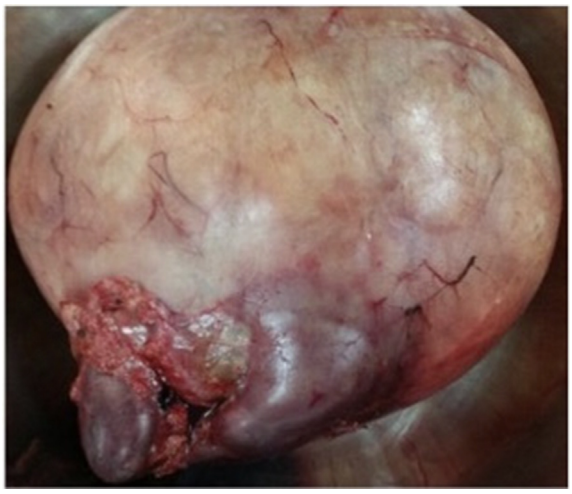

Fig. 2. Macroscopic aspect of the excided mass, included total nephrectomy.

3. Discussion

A case of primary renal leiomyoma of the right kidney, in an 11-year-old girl, presenting with a right flank mass and abdominal pain, is reported.

Benign renal tumors have been reported infrequently in children, and renal leiomyoma is one such rare lesion. Renal leiomyoma 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 [3].

Primary renal leiomyoma was first described by Busse in 1899 [10] and the first pediatric case of leiomyoma in a newborn infant was recorded by Zuckerman et al. [6]. To date, few cases have been described in children. Gupta et al. [7] described a case of a right renal leiomyoma in a 6-year-old boy which was suspected to be a Wilms tumor. Dionne et al. [8] reported a pediatric renal transplant patient who developed an Epstein-Barr virus-associated renal

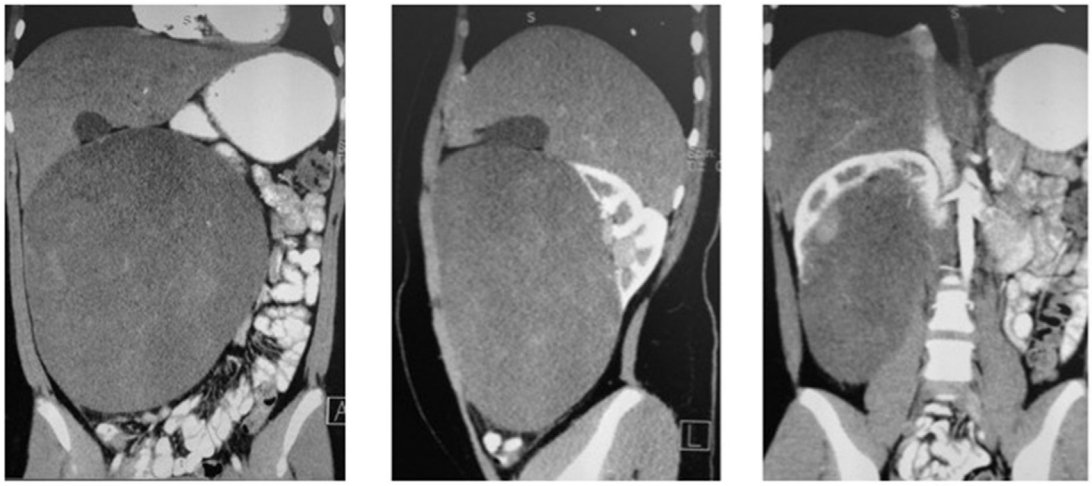

Fig. 1. Contrast-enhanced computerized tomography scan. The mass displaces bowel loops to the left side of the abdomen (Panel A) and the right kidney superolaterally, arises from its anterior parenchyma (Panel B, C).

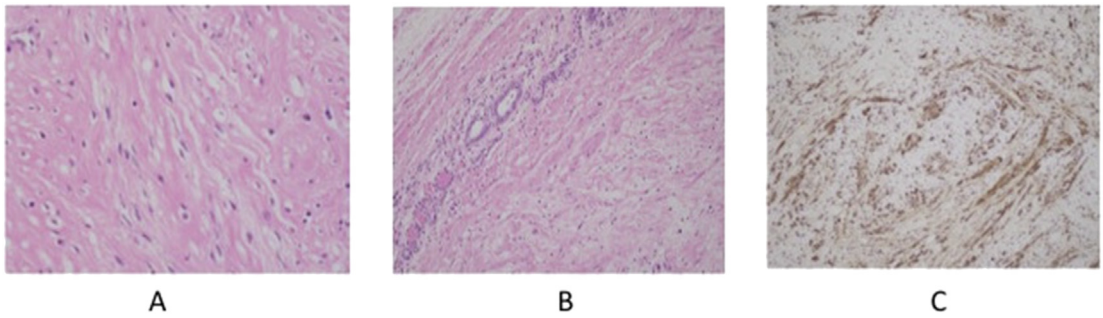

Fig. 3. Histological and immunohistochemical evaluation. Panel A, B: hematoxylin/eosin staining; in B presence of renal tubules. Panel C: Desmin stain, positive reaction.

leiomyoma in his transplanted kidney 5 years post-transplantation. Tawfik et al. [9] showed a unique case of multicentric bilateral renal cell carcinomas and a simultaneous large renal vascular leiomyoma in an 11-year-old child with sickle cell anemia.

Renal leiomyoma usually remain asymptomatic until they produce a mass effect. The most common presenting symptoms, in clinically evident cases, are a palpable mass (57%) and abdominal/flank pain (53%), with a combined occurrence in approximately 33% of cases. Only 20% present with gross hematuria [2,7,11,12].

Given their exceptional incidence, they are rarely included in the differential diagnosis of renal masses, despite their capacity to grow to a considerable size. The imaging procedures such as ultrasonography and CT scan may allow early recognition of renal tumors. The leiomyoma has a well-defined margin, without signs of invasion into the surrounding parenchyma [13,14]. Morphologically renal leiomyomas are peripherally located with well-defined margins and associated buckling of the renal cortex [14]. On ultrasound they present as a solid mass, but cystic changes are not uncommon. On non-contrast CT, leiomyoma is hyperdense compared to the renal parenchyma, with a density similar to muscle. After contrast material administration, leiomyoma shows a lower enhancement than the surrounding parenchyma at the corticomedullary phase [14]. On MRI, they are commonly of low signal intensity on T1- and T2-weighted images [15]. The capsular location of the lesion may provide a clue to the diagnosis. The definitive diagnosis of a leiomyoma is only possible after histopathological examination.

Macroscopically the renal leiomyoma is described as a white or red peripheral lesion, well-defined, with a solid aspect and elastic consistency [12]. Colour is related to vascularization [3]. Focal calcification or cystic degeneration may be present [3]. In our girl, the mass was 22 cm with a weight of 4 kg. In the literature, the average size and weight have been found to be 12.3 cm and 1.84 kg, respectively. The largest reported leiomyoma measured 57.5 cm in maximum diameter and weighed 37.2 kg in an adult patient [2,16].

Histologically, renal leiomyomas shows the presence of fusiform cells distributed in bundles within a lax hyalinized stromal component. Of note are the absence of mitotic figures and other signs of malignancy [3,16]. The differential diagnosis includes leiomyosarcoma, fibroma, angiomyolipoma, congenital mesoblastic nephroma, sarcomatoid renal carcinoma, and, in cases of leiomyoma of the renal pelvis, differentiation with respect to schwannoma is required [17].

Immunohistologically, the tumor cells show positive immunostaining for vimentin and smooth muscle markers. Moreover, the capsular leiomyoma frequently contains a population of cells strongly positive for melanoma markers [3].

Radical nephrectomy remains the recommended treatment for large leiomyoma [7,12,18] because of the difficulty in achieving a diagnosis preoperatively and the size of the tumor; nevertheless kidney-sparing surgery can be performed for smaller or moderate-sized exophytic masses of apparent capsular or sub-capsular origin. Nephrectomy can be performed by a laparoscopic approach, for small size tumors, and seems to be both safe and effective. These tumors generally behave in an indolent manner and generally do not recur after complete excision. However, given the documented potential for local recurrence, a careful evaluation of the patient to rule out recurrence as well as long-term follow-up is recomended [3].

4. Conclusion

Renal leiomyoma is an extremely rare, benign non-metastasizing tumor in children, that must be included in the differential diagnosis of renal masses. Considering the difficulty of clinical diagnosis of this tumor, strong suspicion is indicated when a patient presents with voluminous, well circumscribed renal lesions. Radical nephrectomy remains the recommended treatment for large leiomyoma.

Conflict of interest

The authors Ghassan Nakib, Nidal Mahgoub, Valeria Calcaterra, Gloria Pelizzo declare that they have no competing interests.

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