Inflammatory Myofibroblastic Tumor
Report of a Rare Case in Kidney

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Inflammatory myofibroblastic tumor (IMT) is a rare neoplasm mostly seen in the lungs, but also in extrapulmonary sites. The most common genitourinary site of IMT is the bladder, but it may rarely be seen in the kidneys. We report a case of a 15-year-old girl presented with flank pain and hematuria, in which computed tomography scan revealed a mass in the left kidney. The patient underwent left nephrectomy for a diagnosis of Wilms tumor. Further assessment of the tissue demonstrated a pathologic diagnosis of IMT. Despite improvements in imaging technology, the preoperative diagnosis of IMT remains difficult and surgery is the only way for the diagnosis and treatment. Considering the role of the pathologic examination in making the definite diagnosis of IMT, we should be aware of this entity and it must be considered in the differential diagnoses.

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Case Report

INTRODUCTION

Inflammatory myofibroblastic tumor (IMT) is a rare disease of the kidney which may be seen at any ages and any anatomic location.[1] Although recent studies suggested the neoplastic process of the disease, its origin was doubtful to be a neoplasm or a postinflammatory process.[2] Although the most commonest site of IMT is the lung, as first described in 1937, there are some reports of IMT in the extrapulmonary sites.[3,4] Inflammatory myofibroblastic tumor could also be seen in the genitourinary system, mostly in the bladder,[5] and its occurrence in the kidney is rare. Therefore, it may misdiagnosed as a malignant tumor during clinical and pathological evaluations of a renal mass.[6] First case of the IMT of kidney was published in 1972.[3] Herein, we report a case of a 15-year-old girl with IMT who was misdiagnosed with Wilms tumor based on imaging findings, but further pathologic assessment showed the original process of the disease.

CASE REPORT

A 15-year-old girl was referred to Alzahra Hospital (affiliated to Isfahan University of Medical Sciences, Isfahan, Iran) with a left renal mass. This was discovered incidentally during her evaluation for the complaints of weight loss for 2 months, hematuria, and flank pain. Past medical, social, and family history of the patient was unremarkable. Basic laboratory examinations revealed normal hematological and biochemical parameters of

complete blood count and serum biochemistry. Urinalysis results were normal.

A contrast-enhanced computed tomography scan showed a solid mass with round borders in the upper pole of the left kidney. It was slightly enhanced with contrast, suggesting a malignant neoplasm. The contralateral kidney was normal. Considering the clinical and radiologic findings, malignancy, particularly Wilms tumor, was suspected. A left radical nephrectomy plus adrenalectomy was performed. Grossly, a yellowish well-defined mass measuring 13.5 cm in its greatest dimension was found in the upper pole of the left kidney. Renal capsule was intact.

Histologic examination of the specimen showed proliferation of spindle cells with elongated cytoplasmic processes in a loose edematous and myxoid background. Nucleoli revealed occasional atypia and some contained prominent nucleoli. Diffuse lymphoplasmacytic inflammation with occasional eosinophis and neutrophils infiltration was found. Immunohistochemical staining demonstrated the spindle cells diffusely positive for vimentin, and focally positive for both actin and anaplastic lymphoma kinase (ALK; Figures 1 and 2). Staining for epithelial membrane antigen, cytokeratin 7, CD34, Bcl2, and desmin was negative. A final diagnosis of IMT was made. The remaining renal parenchyma and adrenal gland were unremarkable and surgical margins were free of tumor.

The patients was doing well during the follow-up period with no evidence of disease recurrence for 1 year after the surgery.

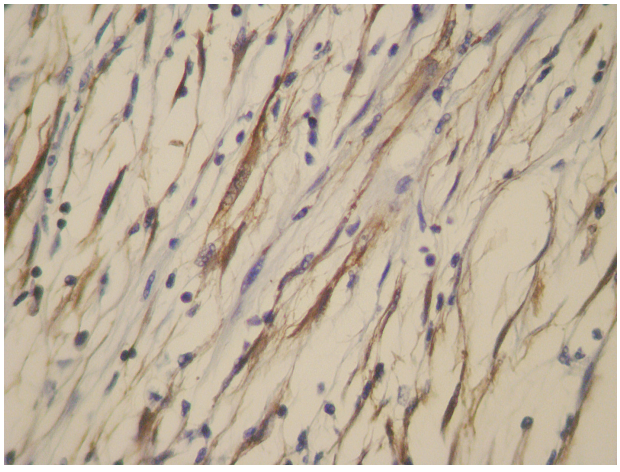

Figure 1. Positive cytoplasmic immunostaining for anaplastic lymphoma kinase (ALK1).

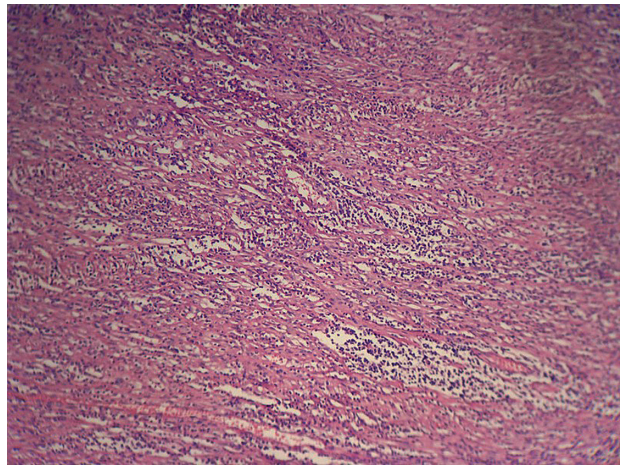

Figure 2. Spindle cell proliferation in an edematous stroma with scattered acute and chronic inflammatory cell infiltration (hematoxylin-eosin, × 100).

DISCUSSION

Inflammatory myofibroblastic tumor, or *inflammatory pseudotumor* (as named before), is a proliferative lesion of spindle cells which mainly affects the lungs, but also may be seen in other extrapulmonary sites.[7,8] Renal IMT is a rare pathologic diagnosis because of its rare occurrence. Also, the histological changes make the diagnosis difficult. It is composed of spindle cells with variable inflammatory component.[2] There are 3 histologic patterns for IMT: a myxoid and vascular pattern with inflammatory infiltrate (same as our case), compact spindle cell proliferation, and hypocellular fibrous pattern.[4] Preoperative diagnosis of IMT is only confirmed by nephrectomy and pathologic assessment.[9] Differential diagnoses of renal IMT are renal cell carcinoma, sarcomatoid urothelial carcinoma, leiomyosarcoma, and rhabdomyosarcoma.[3] Follow-up of patients did not show any recurrences or metastases of IMT. Local recurrences and malignant transformation have been reported in a small subset of patients.[10] Previous studies suggested Epstein-Barr virus and human herpesvirus 8 as a major cause of IMT, Yamamoto et al. evaluated 21 patients with IMT, Epstein-Barr virus and human herpesvirus 8 were both negative among them.[11] Anaplastic lymphoma kinase (ALK), which is used in diagnosis of anaplastic large cell lymphoma, is expressed in IMT cells caused by chromosomal translocation of the *ALK* gene (chromosome 2p23) with a partner gene (nucleophosmin or others), which is useful in confirming the diagnosis.[8,12,13]

Despite recent improvements in imaging technology, preoperative diagnosis of IMT remains a dilemma, and final diagnosis is based on histopathologic evaluation of the involved tissue. In the present case, a left radical nephrectomy was carried out as the disease was presumed to be a Wilms tumor. However, histopathological diagnosis suggested IMT and expression of vimentin, smooth muscle actin, and ALK confirmed IMT as the final diagnosis. Careful histologic examination and immunohistochemical staining will generally determine the appropriate diagnosis. As nearly all the reported cases of IMT had long disease-free survival, we suggest IMT be considered as a differential diagnosis for renal masses. Further studies are required to find an accurate way for preoperative diagnosis of IMT.

CONFLICT OF INTEREST

None declared.

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