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Case Report

Child inflammatory myofibroblastic tumor of the kidney misdiagnosed as Wilms' tumor: case report [☆]

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ABSTRACT

Inflammatory myofibroblastic tumor (IMT) is a rare mesenchymal tumor with recurrent potential, most commonly occurring in the lung but rarely in the kidney with nonspecific clinical symptoms and radiographic features, thus may be misdiagnosed as primary malignant lesions. We described a 6-year-old boy with renal IMT misdiagnosed as Wilms' tumor and then treated with right nephrectomy. It should be emphasized that in addition to the most common renal tumors in children, IMT should also be taken as a differential diagnosis. It is therefore mandatory to carry out clinical interpretation, careful histologic examination, and immunohistochemical studies collectively to make solid diagnosis.

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Introduction

Inflammatory myofibroblastic tumor (IMT) is a rare mesenchymal tumor tends to be locally invasive or recurrent and rarely metastasizes. Originally described in the lung, IMT of the urinary system occurs mostly in the bladder [1]. However, kidney

IMT represents a challenging diagnostic entity due to its non-specific clinical presentation, laboratory findings, and most importantly radiologic findings. It is especially difficult to distinguish renal IMT from malignant tumor before surgery in children. Therefore, the awareness of its existence and deep understanding of its clinical and imaging characteristics are essential to avoid misdiagnosis and to provide better management. Despite the uncertain biological behavior of IMT, most

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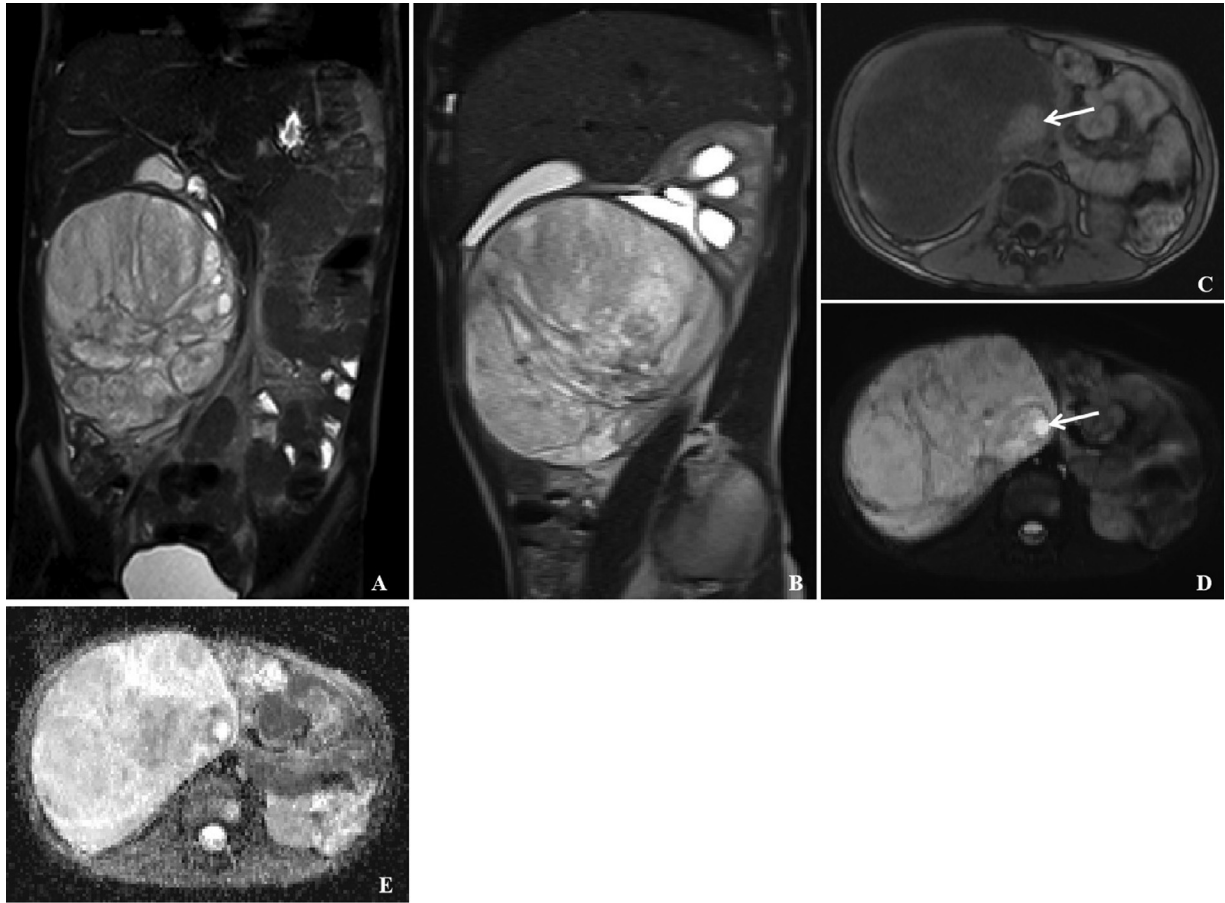


Fig. 1 – The magnetic resonance imaging (MRI) images of our patient. (A) Coronal T2WI demonstrates a rounded mass with mixed signals in the right middle lower abdomen. (B) Sagittal T2WI showed the mass associated with the lower pole of the right kidney and hydronephrosis. (C) Axial T1WI showed low intensity accompanied patchy hyperchromatic areas (arrow in C). (D) Axial DWI showed high signal mixed up with patchy higher signal shadow (arrow in D). (E) ADC presents uneven mixed signals.

patients get a favorable prognosis after surgery [2]. Herein, we report an IMT in a 6-year-old boy that was misdiagnosed as Wilms' tumor.

Case report

A 6-year-old boy was admitted to the hospital due to a 3-day right lower abdominal distension, the child's family complained that the child got a pain and discomfort in the right lower abdomen after getting up in the morning of July 4, 2016. The family found that the right lower abdomen was raised, accompanied with obvious tenderness as well as slight nausea, the body temperature was 37.5°C. They went to the emergency room of a local hospital. Abdominal ultrasound and CT indicated that the right lower abdomen was occupied with a huge mass. Additional imaging with magnetic resonance imaging (MRI) in our hospital demonstrated a 11.7×11.7×9.8cm size heterogeneous mass originating from the lower pole of the right kidney with a clear boundary (Fig. 1). On T2WI, the mass showed mixed signals with multiple low signal separations;

The mass was dominated with low intensity accompanied patchy hyperchromatic areas on T1WI; DWI showed high signal mixed up with patchy higher signal shadow; ADC presented uneven mixed signals. The tumor constricted pelvic canal junction and caused mildly dilated right renal pelvis and calyceal system. Besides, The surrounding structure were compressed. The patient underwent right nephrectomy due to suspicion of Wilms' tumor and the mass was pathologically and immunohistochemically diagnosed as an IMT (Fig. 2), in which spindle cells were admixed with variable amounts of neutrophils, plasma cells, and lymphocytes on a mucinous background. The patient recovered well after operation, and no recurrence or metastasis was noted during follow-up.

Discussion

Inflammatory myofibroblastic tumor (IMT) is a very rare mesenchymal tumor that can behave as a locally benign or aggressive lesion [3]. Histologically, it is composed of spindle cells with an inflammatory infiltrate of plasma cells, lympho-

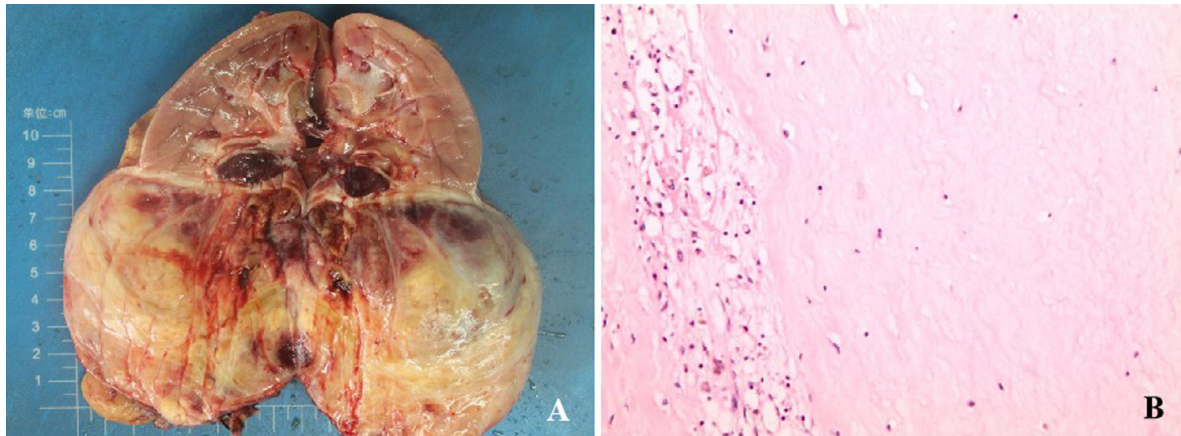


Fig. 2 – Pathological findings of the IMT. The gross specimen (A) and Hematoxylin and Eosin(H&E) stainings of the IMT were demonstrated at 100 (B) magnifications.

cytes, and/or eosinophils [3,4]. Microscopically, 3 histologic patterns have been previously described: Type I was characterized as loosely organized spindle cells admixed with small blood vessels and inflammatory cells in a myxoid background, namely mucous/vascular intensive; Type II is featured with dense clumps of spindle cells, scattered in a small number of inflammatory cells, called spindle cell rich type; Type III is the hypocellular fibrous pattern, usually occurs in soft tissue of limbs, showing large collagen fibers mixed with a few tumor cells and local calcification or ossification [3,5]. Immunohistochemical analyses confirmed that vimentin, muscle-specific actin, smooth muscle actin and CK were positive in IMT.

Etiology has been linked to surgery, vascular causes, chronic inflammation, infection, trauma, immune disorders and autoimmune reaction, however, it remains unclear [6]. The occasionally seen aggressive features and distant metastasis cannot be well explained by the inflammatory etiology alone. The current WHO classification of this lesion is intermediate grade malignant, with the potential of recurrence and rare metastasis. Many studies tried to link the aggressive behavior with certain molecular markers such as P-53 and translocations of ALK-1 gene, yet inconsistent results among these studies make a prediction of prognosis difficult [7]. The fever may indicate a chronic inflammatory process in our case.

IMT frequently involves the lung and now has been reported at multiple extrapulmonary sites. In the urogenital tract, IMT mainly affects the urinary bladder, but rarely the kidney. The clinical manifestations were not typical, mainly manifested as lumbago, gross hematuria, and fever [7,8]. In our case, the patient presented with abdominal pain accompanied with a fever.

The imaging manifestations of renal IMT are highly variable due to pathological types, disease duration and blood supply, it is nonspecific and easy to be misdiagnosed as malignant tumor before surgery, almost all cases were diagnosed at the time of surgical intervention. When it occurs in children, it is difficult to differentiate IMT from the most common Wilms' tumor. Nevertheless, the purpose of imaging examination is to judge the location, size, invasion, relationship with surround-

ing structure and metastasis, so as to guide the patient management.

At present, anti-inflammatory drugs, steroids, surgery, even radiotherapy has been reported as an option to controlling kidney IMT [8]. Nevertheless, total nephrectomy is still advocated due to a subset of rare metastasis and recurrence. In the present case, the preoperative diagnosis was Wilms' tumor, as the mass was large and lead to hydronephrosis, a right nephrectomy was carried out.

Although IMT mimics malignancy, however, these tumors generally behave in an indolent manner and do not recur after complete surgical excision and the outcome is quite favorable [9]. Local recurrence and malignant transformation have been reported in a small subset of patients, particularly in those cases in which complete surgical excision was not possible. Recurrence or metastasis has not yet been discovered from our case.

Conclusion

Renal inflammatory myofibroblastic tumor (IMT) is extremely rare, when it occurs in children, it is difficult to distinguish from nephroblastoma both clinically and radiologically, and the final diagnosis and differential diagnosis rely on pathology and immunohistochemistry. The analysis of their reliable imaging characteristics is of great importance to determine the appropriate diagnosis and patient management to avoid excessive treatment. In addition, IMT should be considered as a differential diagnosis when presented as a large renal mass in children with pain and fever.

Patient consent

Written informed consent was obtained from the children's parents for the publication of patient information in this article.

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