Inflammatory Myofibroblastic Tumor: A Rare Case in Kidney

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Abstract

Inflammatory Myofibroblastic Tumour (IMT) or ‘pseudotumour’ of the kidney is a uncommon benign tumour, common in young adults with unknown etiology. Clinical feature and radiological investigations are usually inconclusive. Often, biopsy is inconclusive required a management similar to that of Renal Cell Cancer (RCC). Diagnosis is based on immunohistochemistry. We are reporting a case of IMT in a 32-year-old female patient who presented with left flank pain and haematuria which on evaluation was suggestive of left renal cell carcinoma. IMT was confirmed by Surgery, histopathological examination and Immunohistochemistry.

Keywords: Inflammatory myofibroblastic tumor; Inflamatory pseudotumor.

Introduction

Inflammatory myofibroblastic tumor (IMT) is a unique myofibroblastic neoplasm with more common in the lung.¹ In the urological tract, these tumors can occur in the bladder and prostate, but kidney involvement is rare.² Aetiopathogenesis is not yet clear and difficult to differentiate from malignancy. The preoperative diagnosis of renal IMT remains difficult due to its nonspecific manifestations and the mimicry of malignancy by imaging, such as renal cell carcinoma, which possibly leads to nephrectomy.³⁻⁷ Histologically, there is evidence of marked proliferation of myofibroblasts, fibroblasts, histiocytes, and plasma cells. There were few reports of recurrence of IMT.⁶

Case report

A previously healthy 32-year-old presented with pain in the left flank, hematuria and recurrent fevers for 5 days 3 months back. She underwent a course of antibiotics, prescribed by her doctors, for a documented urinary tract infection. On physical examination no abnormality was found. Routine laboratory blood examination was normal. Urinalysis revealed an elevated number of red blood cells. The renal function results were as follows: serum creatinine of 0.93 mg/dl (reference range, 0.6-1.2 mg/dl). Renal sonography was performed. Examination of the left kidney revealed a heterogeneous irregularly outlined isoechoic soft tissue mass with internal cystic space measuring about 5.9cm and 4.2 cm occupying anterior lateral cortex including medulla and sinus; internal flow in seen fewer on Doppler application (Figure 1). The renal artery, veins and inferior vena cava were patent. The right kidney was normal. A contrast-enhanced computed tomography (CT) scan revealed a mass (4.9×4.5 cm) arising from the lower part of the left kidney with presence of perinephric fat stranding. It was slightly enhanced with contrast, suggesting a malignant tumor such as renal cell carcinoma (Figure 2). Enlarged lymph node is noted in left para-aortic region. Sonography guided FNA of left renal mass was done microscopic examination suggestive of renal cell carcinoma.

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Radical nephrectomy was done with a diagnosis of Renal Cell Carcinoma (RCC). On cut section, cortical surface is distorted by a small timorous lesion measuring about 3.0 cm. The cut surface the lesion was solid and fibrotic. Histologically, the tumour consisted of spindle myofibroblast, fibroblasts and infiltration of chronic inflammatory cells including many lymphocytes and few plasma cells. On immunohistochemistry (IHC) the tumour was positive for vimentin and smooth muscle actin (SMA) and negative for ALK 1, CD34 and S-100 protein. The diagnosis of inflammatory myofibroblastic tumor (IMT) was confirmed. There was no recurrence at 1 year follow-up.

**Fig.-1:** Initial longitudinal gray scale sonograms show a 5.9 × 4.2cm mass (arrows) with variable echogenicity in anterior lateral of the left kidney with some areas of cystic components.

**Fig.-2:** A and B, Precontrast (A) and postcontrast (B) axial CT images through the left lower part of the kidney show the heterogeneously enhancing renal mass with perinephric infiltration (C)Coronal reformatted CT image in the excretory phase better delineates the mass arising from the lower part of the left kidney.
Discussion

Inflammatory myofibroblastic tumor, or inflammatory pseudotumor (as named before), is a very rare tumor affecting young people, males (58%) more than females, the occurrence in elderly patients are extremely rare. Inflammatory pseudotumours are common in lungs and orbit. However, in the urogenital tract, IMT of the kidney is extremely rare. In the kidney, so far 40 cases have been reported. Inflammatory myofibroblastic tumor of the kidney was first reported in 1972 as plasma cell granuloma. Aetiopathogenesis is not clear yet, which is likely to be multifactorial, including infection, vascular causes and autoimmune disorders. Initially, IMT was thought to result of a reactive inflammatory process, or “pseudotumor”. This inflammatory reaction may be secondary to surgery, trauma, or infection. A portion of inflammatory pseudotumors may be due to a variety of infectious agents including Actinomyces, Pseudomonas species, and mycoplasma. Infections due to Epstein-Barr virus and herpes virus are also considered causative factors. Rearrangements of ALK (anaplastic lymphoma kinase) gene locus on chromosome 2p23 has been present in both pulmonary and extrapulmonary IMTs, providing further support for the neoplastic nature and distinction from other “inflammatory pseudotumours”.

Although there are no specific clinical symptoms of IMT, patients commonly presented as incidental finding (36%) to pain (38%), microscopic or gross hematuria (28%), and constitutional symptoms (23%).

It is extremely difficult to exclusion of malignancy preoperatively. Renal cell carcinoma, transitional cell carcinoma, malignant lymphoma, xanthogranulomatous pyelonephritis, angiomyolipoma with minimal fat, rhabdomyosarcoma in adults and Wilm’s tumor in children are the differential diagnosis of a renal IMT.

Most diagnoses have been made at the time of surgical intervention, as because it is difficult to obtain enough tissue by CT-guided fine needle aspiration to make a definitive histopathological diagnosis.

Histologically, IMT is consisting of proliferation of spindle cells associated with various amounts of lymphocytoplasma-cytic infiltrate. There are three histologic patterns of IMT as myxoid-vascular pattern, compact spindle cell pattern, and hypocellular fibrous pattern. Immuno histochemical studies suggest the myofibroblastic nature of this lesion with expression of vimentin and smooth muscle actin. These tumors are strongly positive for CD 34 reactivity. ALK is only positive in 50% and it generally tends to be positive in younger patients. There are different architectural appearances and have been described as a Pattern less pattern.

Local recurrences and malignant transformation have been reported in a small number of patients, but usually these occur where tumor is of large size, located in abdominopelvic area, and occurring in elderly patient and complete resection has not been possible.

Although most renal IMTs (95%) were treated with nephrectomy, in some cases that were treated conservatively, corticosteroid therapy was somewhat beneficial. In our case, because the preoperative imaging and FNAC were confirmed as renal cell carcinoma, surgical resection was implemented, and there was no recurrence observed during follow-up.

Conclusion

In conclusion, patients presenting with renal mass, haematuria and pain in the loin need detailed evaluation with various imaging modalities like Ultrasonography, CT scan and MRI. Rare tumour like IMT has no specific imaging features provided a definitive diagnosis. Careful histological examination and immunohistochemical study will generally determine the appropriate diagnosis. As most of the reported cases of IMT had long disease free survival, we suggest IMT may be considered as a differential diagnosis for renal masses. Further studies are required to find an accurate way for preoperative diagnosis of IMT.

References
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