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Inflammatory Myofibroblastic Tumor of the Kidney Misdiagnosed as Renal Cell Carcinoma

The inflammatory myofibroblastic tumor (IMT), also knowns as inflammatory pseudotumor, is a soft tissue lesion of unknown etiology. In the urogenital tract, IMT mainly affects the urinary bladder or prostate, but rarely the kidney. It has been considered as a nonneoplastic reactive inflammatory lesion, but nowadays, it is regarded as a neoplasm due to its high recurrence rate and metastasis. We describe a case of a 61-yr-old woman that had originally been misdiagnosed as renal cell carcinoma, which was pathologically revealed to be an IMT.

Key Words : *Granuloma, Plasma Cell; Inflammation; Kidney Neoplasms*

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INTRODUCTION

Inflammatory myofibroblastic tumor (IMT) is a rare pathologic entity composed of myofibroblasts and an accompanying inflammatory infiltrates. It affects individuals of both sexes and is seen in a wide range of age groups. First described in the lung which is the most common site of involvement, IMT has been described as a benign lesion that mimics malignancy (1). But, this lesion is often mistaken for malignant process in diagnostic procedures and at the time of surgical intervention. In the urogenital tract, the urinary bladder is the most common site of occurrence (2). It rarely originates from the kidney (3). Because of its rare occurrence, it is possible that it is not considered by the physician, and is potentially overdiagnosed as a malignancy both clinically and pathologically. We describe a case of renal IMT in a 61-yr-old woman that clinically diagnosed as renal cell carcinoma.

CASE REPORT

A 61-yr-old woman was referred to the Chonnam National University Hwasun Hospital with a left renal mass which was discovered incidentally. She had been evaluated for a chief complaint of weigh loss with associated gastrointestinal symptoms. Medical, social and family histories of the patient were unremarkable. Basic laboratory examinations of complete blood count, serum biochemistry, and urinalysis were normal. A contrast-enhanced computed tomography (CT) scan demonstrated a round, and solid mass (3.0 × 2.0 cm) originating from the lower pole of the left kidney. It was slightly enhanced with contrast, suggesting a malignant tumor such as renal cell carcinoma. Additional imaging with magnetic resonance imaging (MRI) demonstrated a 3.0 × 2.5 × 2.0 cm heterogeneous mass with a central necrotic portion. T2-weighted MRI demonstrated a mildly hyperintense lesion, possibly renal cell carcinoma (Fig. 1). Neither renal vein invasion nor inferior vena cava extension was evident on MRI. The contralateral kidney was normal. On the basis of the clinical and radiologic findings, a malignancy such as renal cell carcinoma was suspected. A laparoscopic left radical nephrectomy and adrenalectomy were performed under the diagnosis of renal cell carcinoma. The tumors were well encapsulated with dense collagenous fibrous tissue (Fig. 2). Histologic examination of the tumor revealed spindle cells with myxoid change admixed with lymphocytes, and plasma cells (Fig. 3A, B). The spindle cells were diffusely positive for vimentin, and focally positive for both actin and epithelial membrane antigen (EMA) (Fig. 3C, D). Immunohistochemistry demonstrat-

ed no staining for cytokeratin, HMB45, cyclin D1, S-100, C-kit, CD31, CD34, desmin. The pathologic diagnosis was renal inflammatory myofibroblastic tumor. The remaining renal parenchyma and adrenal gland were unremarkable. Surgical margins were clear and no histologic evidence of extension into the renal vein, artery, or ureter was found. This patient was doing well without evidence of recurrence 3 months after surgery.

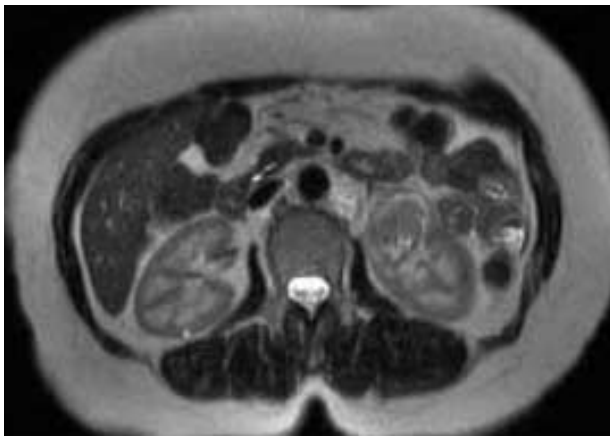

Fig. 1. Magnetic resonance image demonstrates a 3.0×2.5 cm size solid mass with mildly enhancement on left kidney lower pole with a central necrotic portion.

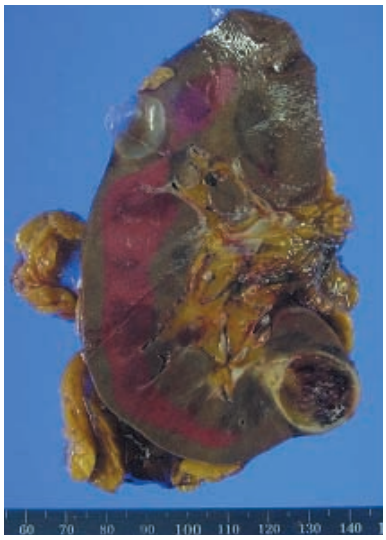

Fig. 2. Left kidney coronal opening specimen shows a well-circumscribed encapsulated mass measuring 2.7×2.8 cm size, involving the lower pole. The mass revealed areas of myxoid change necrosis, and cystic change.

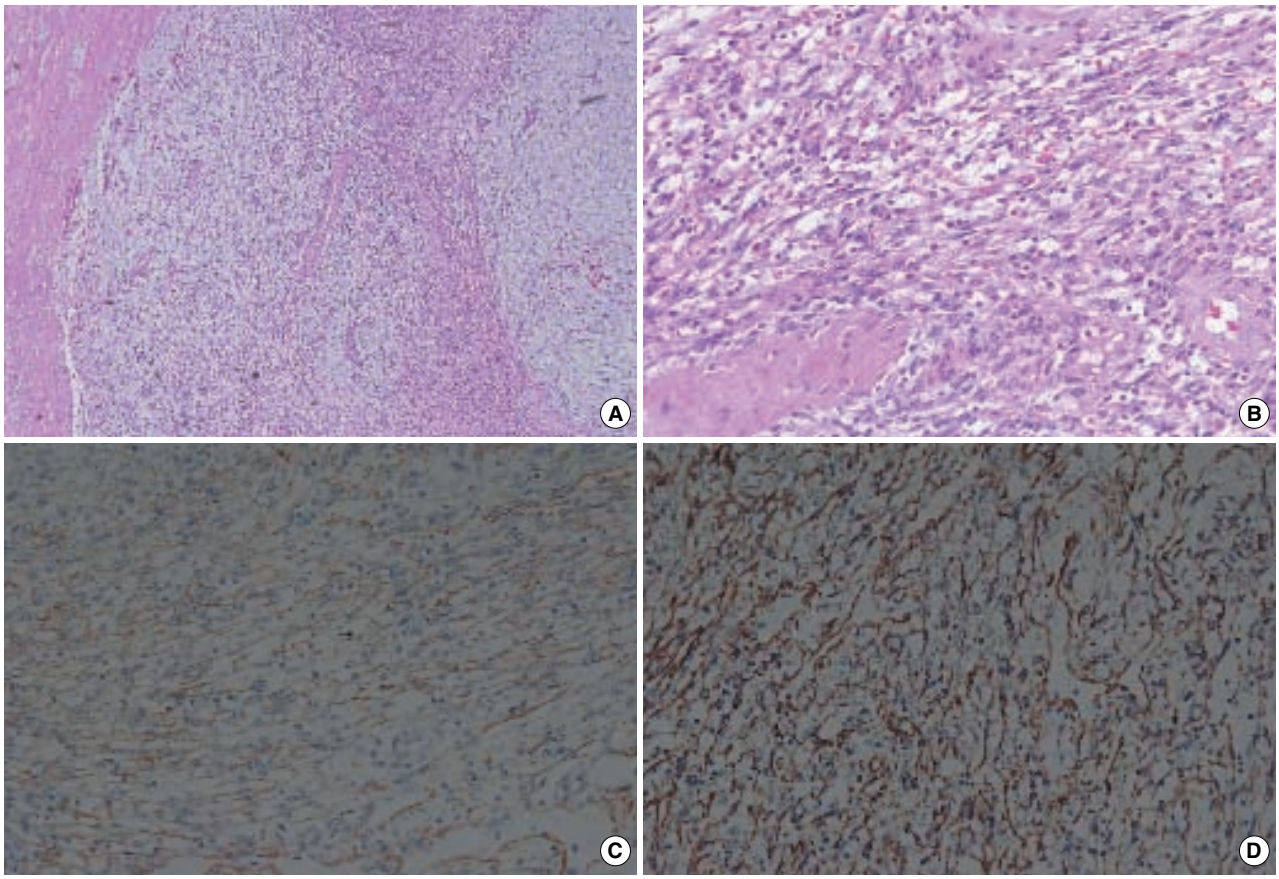

Fig. 3. Microscopic findings. (A) The low power appearance demonstrating a capsule of dense collagenous fibrous tissue and myxoid zone and inflammation with cellular zone consisting of spindle cells arranged in fascicles. (B) The area of myofibroblastic proliferation showing densely cellular fascicles. The tumor cells were potive for smooth muscle actin (C) and vimentin (D).

DISCUSSION

IMT is a rare entity with distinctive histologic features. Originally described in the lung, IMT has now been reported at multiple extrapulmonary sites (4). However, in the urogenital tract, IMT of the kidney is extremely rare. Patients usually present with hematuria and/or abdominal pain. Kapusta et al. (5) reported a series of twelve IMT of the kidney and the renal mass, in many patients, was discovered incidentally. This tumor has been described as a benign lesion that mimics malignancy. Clinical examination and radiological investigations are often inconclusive. Almost diagnoses have been made at the time of surgical intervention. Eric et al. (6) demonstrate the first reported association of malignancy with IMT in the urogenital tract and the necessity for surgical excision. Therefore, preoperative exclusion of malignancy is very difficult and there is a risk that this lesion could be misdiagnosed such as renal cell carcinoma. Most reports noted no local recurrence, no malignant transformation, and no recurring clinical course after complete surgical excision (4). Although these generally tend to be where complete resection has been impossible, a subset of cases may be locally aggressive, recurrent, or rarely metastatic. Additional reports by Ludvik et al. (7) noted sarcomatous transformation and multiple metastases of IMT. The radiological findings in renal IMT are not well described. Also, there are no published reports or guidelines regarding the optimal imaging technique for followup. The differential diagnoses of the renal IMT include malignant tumors such as renal cell carcinoma, sarcomatoid renal cell carcinoma, inflammatory fibrosarcoma, myxoid leimyosarcoma and nonmalignant tumors such as angiomyolipoma, xanthogranuloma and pyelonephritis. Histologically, IMT is characterized by a proliferation of spindle cells admixed with variable amounts of a lymphoplasmacytic infiltrate. Because IMT can be confused with both reactive processes as well as potentially malignant neoplasm, distinguishing them from their histologic mimics plays an important role in assuring appropriate patient management. Three histologic patterns (myxoid-vascular pattern, compact spindle cell pattern, and hypocellular fibrous pattern) of IMT have been described by Coffin et al. (4). Immunohistochemical studies support the myofibroblastic nature of this lesion with consistent expression of vimentin and smooth muscle actin. Variable positivity has been identified for HHF-35, cytokeratins, and CD68. The pathogenesis of IMT remains uncertain. Initially, IMT was thought to represent a reactive inflammatory process, or "pseudotumor". This inflammatory reaction may be secondary to surgery, trauma, or infection. No single etiology or pathogenesis has yet been established for IMT. Some cases may be related to an infectious or autoimmune process. A subset of inflammatory pseudotumors appears to be associated with a variety of infectious agents including Actinomyces, Pseudomonas species, and mycoplasma. Another suggested etiologic agent is Epstein-Barr virus, as some cases of IMT have been positive for Epstein-Barr virus latent membrane protein, especially in the liver and spleen (8, 9).

In the present case, a laparoscopic radical nephrectomy was carried out as the disease was presumed to be renal cell carcinoma. However, histopathological diagnosis was IMT. Clinical presentation, careful histologic examination, and immunohistochemical studies will generally determine the appropriate diagnosis.

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