Inflammatory myofibroblastic tumor of the kidney: a case report and review of the literature

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KEY WORDS

kidney ◗ anaplastic lymphoma kinase (ALK) ◗ inflammatory myofibroblastic tumor ◗ kidney ◗ plasma cell granuloma

ABSTRACT

Inflammatory myofibroblastic tumor (IMT) is an uncommon tumor believed to be benign and can arise in various organs. We report a case of renal IMT in a 74-year old female with right upper quadrant abdominal pain. Computerized tomography showed a 4.5 cm upper pole renal mass, consistent with a renal cell carcinoma. Subsequently a laparoscopic right radical nephrectomy was performed. Pathology revealed an unusual lesion consisting of spindle cells having prominent nucleoli, with plasma cells and lymphocytes throughout the lesion. Staining was positive for anaplastic lymphoma kinase (ALK), vimentin, CK 18, CK-AE 1/3, and CAM 5.2 consistent with an IMT.

INTRODUCTION

Renal inflammatory myofibroblastic tumor (IMT) is a rare tumor. It was first reported as a plasma cell granuloma of the renal pelvis.[1] In the urogenital tract, the bladder is the most common site for IMT, however an IMT of the prostate has also been reported. [2] Renal IMT is uncommon, with only a few cases reported to date. Some of the synonyms for this entity include plasma cell granuloma, pseudosarcomatous fibromyxoid tumor, and inflammatory pseudotumor. IMT, as it is presently termed, presents a challenge in terms of etiology, clinical diagnosis, and treatment. We report a case of IMT in the right kidney of a 74 year old female.

CASE REPORT

A 74 year old female presented with a four month history of right upper quadrant abdominal pain. Past medical history was non-contributory and physical examination was unremarkable. Urinalysis was negative for hematuria. CT scan showed a 4.5 cm homogenous mass occupying the lateral aspect of the upper pole of the right kidney and was enhanced from 33 to 95 Hounsfield units (HU) following contrast administration. The tumor protruded into the renal sinus fat without invading it. The renal vein and inferior vena cava (IVC) were patent (Fig. 1). The preoperative diagnosis was renal cell carcinoma. The patient underwent an uneventful laparoscopic right radical nephrectomy. At 24 months follow-up the patient is disease free.

PATHOLOGICAL FINDINGS

The excised kidney measured 13.5 x 7.0 x 4.0 cm and weighed 217 grams. Gross examination revealed a well circumscribed, fleshy, gray-yellow mass in the upper pole of the right kidney (Fig. 2). Microscopic examination showed an unusual lesion consisting of a relatively monomorphous proliferation of spindle cells with prominent nucleoli. A rare pleomorphic nucleus was seen, yet no mitotic activity was evident. Plasma cells and lymphocytes were scattered throughout the lesion (Fig 3). On staining with immunohistochemistry, many of the tumor cells were positive for keratin and CK18 as well as diffusely positive for ALK-1. The tumor was also positive for vimentin and CK-AE 1/3 in addition to focally positive for CAM 5.2. The markers SMA, MSA, CD34, desmin, and S-100 protein were all negative. Surgical margins and lymph nodes were also negative. The diagnosis of IMT was made.

DISCUSSION

IMT, also known as inflammatory pseudotumor (IPT), was first described by Davides et al. in 1972 as a plasma cell granuloma of the renal pelvis [1]. Since then, nearly 53 cases have been reported. Bell et al. suggested removal of the kidney as a treatment for IMT. To the best of our knowledge, all, except three cases reported so far, have been treated by partial or total nephrectomy. There are no reports of recurrence following surgical therapy [3, 4]. We found two recent case reports of renal IMT treated conservatively. One of them had a history of recurrent systemic IMT involving the common bile duct, lymph nodes, pancreatic tail, and both kidneys. The other patient had bilateral infiltrating renal tumors. In both cases, successful response to high dose corticosteroid treatment (60, 40, and then 30 mg/d of oral prednisone, each for 1 month) allowed clinical confirmation of IMT. A Renal biopsy was performed in one of the two cases. Further follow-up was not available for neither of the cases. Others however, have not had the same success with conservative management [5]. Nephrectomy still remains the treatment of choice considering the fact that, in most cases, IMTs are indistinguishable from malignant renal tumors.

The age at presentation varies from 3 to 89 years with a higher incidence reported in males [6]. Although the lung was the first reported site, it has been known to arise anywhere from the orbit to the abdomen where it can involve the retroperitoneum, liver, mediastinum, abdominal wall, mesentery, or omentum [6]. Differential diagnosis of a renal IMT includes renal cell carcinoma, urothelial carcinoma, myxoid leiomyosarcoma, malignant fibrous histiocytoma, and rhabdomyosarcoma in adults and Wilms' tumor in children. Clinical presentation can vary from abdominal or flank pain, fever, and hematuria to an asymptomatic mass [4, 6-9].

The histopathology is characterized by differentiated myofibroblastic components which can include histiocytes, lymphocytes, plasma cells, eosinophils, and other cellular elements. Three histological patterns have been described: a myxoid and vascular pattern with inflammatory infiltrate; a compact spindle cell proliferation; and a hypocellular fibrous pattern. These three can coexist with one occasionally predominating over the other [8]. Genetic

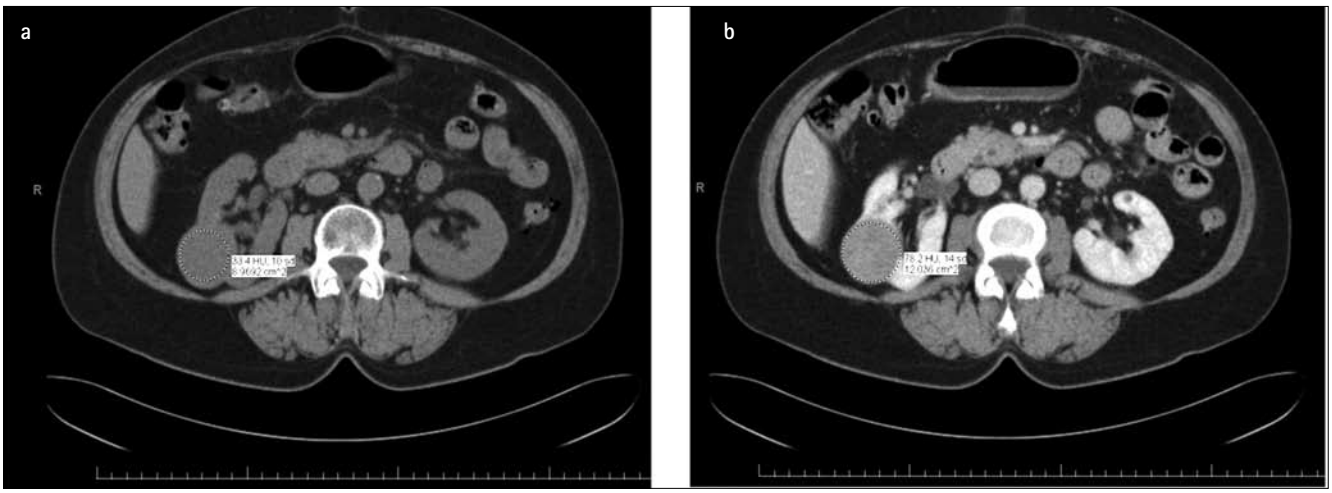

Fig. 1. Preoperative computed tomography scan showing a homogenous 4.5 cm right renal mass: a) pre-contrast b) post-contrast (enhancing by >44 Hounsfield units).

studies have described periodic translocations in IMT involving chromosome 2p23 (the site of the ALK gene), which in turn leads to over-expression of ALK-1 protein [8]. This protein belongs to the insulin receptor family, is associated with a more aggressive course, and is used to support the diagnosis of IMT [8]. Qiu et al. performed a comparative study between low-grade myofibroblastic sarcomas and IMT's and found that the ALK-1 protein, when positive, could be helpful in distinguishing these two entities [10]. In his study of ALK expression, Cook et al. reported that 5.5% showed an "aggressive phenotype". All of which were strongly ALK positive [8].

The diagnosis of IMT is made as a result of the presence of spindle cell proliferation accompanied by a population of plasma cells, lymphocytes, and eosinophils. These spindle cells are monotonous without considerable pleomorphism. Immunohistochemical stains are done to rule out other tumor entities. In our case immunohistochemistry revealed positive staining for vimentin, CK 18, CK-AE 1/3, and CAM 5.2, which are usually present in IMT [6], but negative staining for smooth muscle actin (SMA) and desmin. CD-10 staining, present in renal cell cancer, was also negative.

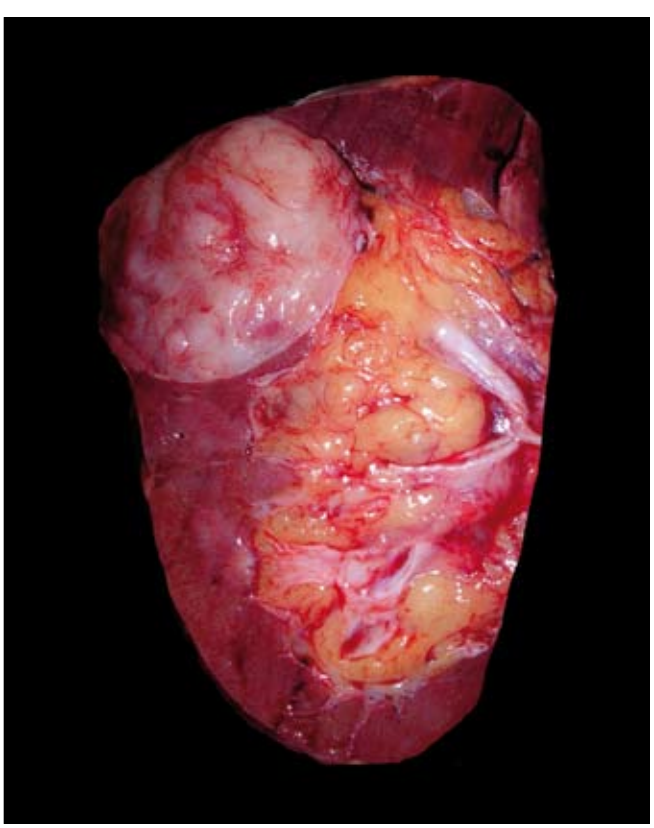

Fig. 2. Nephrectomy specimen showing the upper pole renal mass.

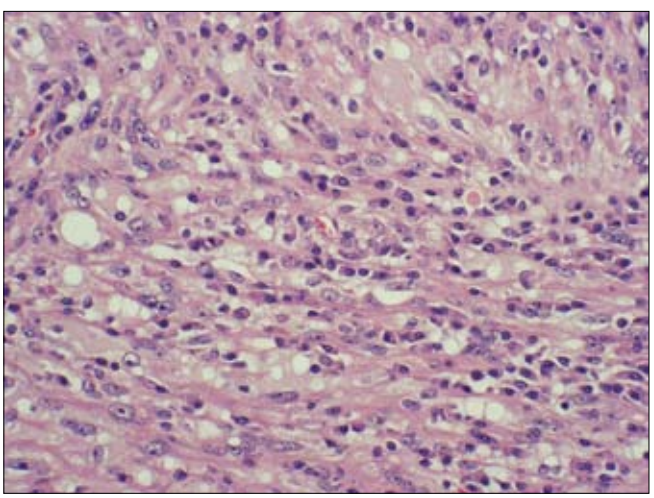

Fig. 3. Histopathological examination showing monomorphous proliferation of spindle cells, a prominent infiltrate consisting of plasma cells and lymphocytes.

Controversy exists regarding the exact nature of the tumor i.e. neoplastic vs. reactive. According to the World Health Organization (WHO) 2002 classification, IMT is under intermediate tumors, rarely metastasizing. The risk of recurrence is approximately 25%. The difficulty lies in determining which subsets of IMT are likely to recur.

Pathologically, when a well-circumscribed spindle cell neoplasm with a dense lymphoplasmacytic infiltrate is encountered a diagnosis of IMT needs to be considered.

In our case the histopathology involved a lesion with monomorphous proliferation of spindle cells with prominent nucleoli and a rare pleomorphic nucleus with no mitotic activity. The ALK-1 protein was diffusely positive in our case, further establishing the diagnosis of IMT.

The need for a cautious approach is emphasized by the reported case of a renal IMT harboring a renal cell carcinoma [7].

The most common radiological appearance of renal IMT is an ill-defined, hypovascular, homogeneous tumor on CT scan. On MRI, variable signal intensity on T1 weighted images and low signal intensity on T2 is observed [11]. However, the radiological appearance is often indistinguishable from other renal tumors making a confident preoperative diagnosis unlikely.

With more cases being reported, additional knowledge of this rare tumor will be obtained.

CONCLUSION

Renal IMT must be considered when evaluating a solitary, solid renal mass. However, it is difficult to make a diagnosis of IMT preoperatively. Nephrectomy was done in our patient with good outcome. Further data on this rare condition can help formulate guidelines for management.

Abbreviations

IMT – Inflammatory myofibroblastic tumor
ALK – anaplastic lymphoma kinase

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REFERENCES

1. Davides KC, Johnson SH, 3rd, Marshall M et al: *Plasma cell granuloma of the renal pelvis*. J Urol 1972; 107: 938-939.
2. Pérez García FJ, Pinto Blázquez J, Gutiérrez García R et al: *Inflammatory prostatic pseudotumor (fibromyxoid pseudosarcomatous tumor)*. Arch Esp Urol 2004; 57: 657-660.
3. Ma Y, Zieske AW, Fenves AZ: *Bilateral infiltrating renal inflammatory pseudotumor responsive to corticosteroid therapy*. Am J Kidney Dis 2008; 51: 116-120.
4. Sugimoto K, Hayashi T, Imanishi M et al: *A case of inflammatory pseudotumor of renal pelvis showing a complete response to antibiotics*. Hinyokika Kiyo 2004; 50: 629-631.
5. Ho PH, Chen SY, Hsueh C et al: *Inflammatory myofibroblastic tumor of renal pelvis presenting with prolonged fever and abdominal pain in children: report of 1 case and review of literature*. J Pediatr Surg 2005; 40: e35-37.
6. Montgomery EA, Shuster DD, Burkart AL et al: *Inflammatory myofibroblastic tumors of the urinary tract: a clinicopathologic study of 46 cases, including a malignant example inflammatory fibrosarcoma and a subset associated with high-grade urothelial carcinoma*. Am J Surg Pathol 2006; 30: 1502-1512.
7. Gwynn ES, Clark PE: *Inflammatory myofibroblastic tumor associated with renal cell carcinoma*. Urology 2005; 66: 880.
8. Cook JR, Dehner LP, Collins MH et al: *Anaplastic lymphoma kinase (ALK) expression in the inflammatory myofibroblastic tumor: a comparative immunohistochemical study*. Am J Surg Pathol 2001; 25: 1364-1371.
9. Sugimoto T, Miwa S, Sugata T, Zen Y: *A case of inflammatory myofibroblastic tumor recognized in the retroperitoneum and the kidney*. Hinyokika Kiyo 2008; 54: 5-7.
10. Qiu X, Montgomery E, Sun B: *Inflammatory myofibroblastic tumor and low-grade myofibroblastic sarcoma: a comparative study of clinicopathologic features and further observations on the immunohistochemical profile of myofibroblasts*. Hum Pathol 2008; 39: 846-856.
11. Nakamura Y, Urashima M, Nishihara R et al: *Inflammatory pseudotumor of the kidney with renal artery penetration*. Radiat Med 2007; 25: 541-547.

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