Title
Urologic manifestations of inflammatory pseudotumor: Report of 2 cases and review of the literature.

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Introduction

Inflammatory pseudotumor (IPT) (synonyms: inflammatory myofibroblastic tumor [IMT] or plasma cell granuloma) is an uncommon disease of unknown etiology. IPT exhibits a wide spectrum of biologic behavior, varying from the more frequent benign lesions to rare tumors that are multifocal and prone to recurrence. It most commonly involves the lung and orbit, but it has been reported to occur at several sites in the body, including the urinary system (1-10). IPT often mimics malignant tumors clinically and radiologically; the clinical course and prognosis of the disease often conform to a benign entity. Preoperative diagnosis of IPT in retroperitoneum is usually not possible, due to varied clinical presentations and nonspecific appearance on imaging studies. Image-guided fine-needle aspiration cytology (FNAC) or a biopsy is required to confirm the diagnosis. It is however important to suspect this condition in the

We report two adult patients with varied urologic symptoms who were found to have inflammatory pseudotumor on histopathology. The first patient had a large, solid, enhancing retroperitoneal mass lesion and presented with increased frequency of urination and recurrent urinary tract infections. The second patient had an obstructing left distal ureteric stricture and presented with painless hematuria. Though preoperative radiological diagnosis of this entity is not feasible, the present article illustrates the imaging findings in this unusual disease entity with review of the relevant literature.

Figure 1. Case 1: 44-year-old male with IPT. Noncontrast CT axial image shows a large, mildly hyperdense mass (arrow) with foci of negative attenuation suggestive of intralesional fat (point ROIs with attenuation values shown). The urinary bladder is displaced to the left side. [UB: Urinary bladder].
appropriate clinical setting, as this entity does not usually require radical surgical management.

Case 1
A 44-year-old male presented with increased frequency of urination and recurrent urinary tract infection of six months' duration. Urine analysis following a course of antibiotic performed before imaging workup did not reveal any pus cells or red blood cells in the urine. Ultrasound examination revealed an ill-defined mass lesion in the right supravesical location. Contrast-enhanced CT study showed a large, rounded, solid, peripherally enhancing mass lesion in the pelvis on the right side displacing the urinary bladder to the left side. Both studies were performed at an outside hospital (not shown). On noncontrast CT images, performed as part of the CT-guided biopsy, the lesion showed focal areas of negative attenuation, consistent with intraläsional fat (Fig. 1). The urinary bladder was otherwise normal in capacity, without any focal abnormality. The prostate gland was also normal in size. Ultrasound and CT imaging findings suggested a likely malignant retroperitoneal mesenchymal tumor. The presence of intraläsional fat also raised suspicion for liposarcoma. After CT-guided biopsy, histopathology showed polymorphic cells consisting of mononuclear and polymorphonuclear cells on low-power magnification. Large multinucleated cells were scattered throughout. High-power magnification showed polymorphonuclear cells, plasma cells, and histiocytes, consistent with a diagnosis of inflammatory pseudotumor (Fig. 2). The patient was advised to undergo surgical resection but declined and is presently being followed up by serial imaging.

Case 2
A 47-year-old male presented with painless hematuria of one week's duration. No similar episode was reported in the past. The complete blood-cell count, serum biochemistry, and coagulation profile were within normal limits. Transabdominal ultrasound showed moderate left-sided hydronephrosis and hydroureter up to the mid segment. No definite calculus was identified in the urinary system. CT urography showed left-sided proximal and mid hydroureteronephrosis with decreased enhancement pattern of the left kidney (Fig. 3). The left distal ureter showed circumferential enhancing mural thickening, suggesting a transitional cell carcinoma (Figs. 4, 5). After wide segmental resection of the ureteric mass with uretero-ureteric anastomosis, histopathology showed curled muscle fibers and fibromyxoid stroma, consistent with a diagnosis of inflammatory pseudotumor (Fig. 6). Followup CT urography at 3 months showed near-complete regression of the left-sided hydroureteronephrosis. Urinalysis was within normal limits, and the patient was free of any symptoms.

Discussion
The term inflammatory pseudotumor (IPT) was coined by Umiker et al (11), as the disease often mimics a malignant process clinically as well as radiologically. The entity was first described in lungs, but this description fits virtually all other body regions. IPT is an uncommon spindle-cell proliferative disorder of unknown etiology, but likely an exaggerated response to tissue injury or infection, as suggested by the presence of proliferating spindle histiocytes and/or dendritic cells within the lesion. It is characterized histologically by the presence of acute and chronic inflammatory cells with a variable fibrous response (12). Myofi-
broblastic proliferation in a few of these tumors also led to the term inflammatory myofibroblastic tumour (IMT). This entity has also been described as fibromyxoid pseudotumor, pseudosarcomatous myofibroblastic tumor, pseudosarcomatous fibromyxoid tumor, and nodular fasciitis, reflecting the lack of consensus as to the exact nomenclature and underlying pathophysiology of this disease.

Despite the complexity, variable histologic characteristics, and behavior of IPT, the condition is now widely accepted as a quasineoplastic lesion, which can present as single or multiple masses with polymorphous inflammatory-cell infiltrate and variable amounts of fibrosis, necrosis, granulomatous reaction, and myofibroblastic spindle cells (1, 13). The clinical and histopathological similarity between IPT in lungs and sclerosing pancreatitis also suggests the possibility of an IgG4-related immunopathologic process in the pathophysiology of IPT (14, 15). The locally aggressive nature of the lesion, with its tendency for multifocal involvement and occasional progression to a true malignant tumor, suggests association with lymphoma. Immunohistochemical studies of T- and B-cell subpopulations may be helpful in distinguishing IPT from lymphoma. IPT usually contains both T cells and B cells, unlike lymphoma, where either B- or T-cell population predominates (1, 12). Heterogeneity of the inflammatory cell population in IPT is in fact a useful parameter to exclude lymphoma. An autoimmune mechanism has also been implicated, as reports have described association of IPT with vasculitis and inferior vena caval thrombosis, along with demonstration of anti-C3 and anti-fibrinogen deposits in the vessel wall (3).

The radiologic features of IPT are variable and nonspecific, possibly due to varying degree of fibrosis and cellular infiltration in these lesions (6). On ultrasound, lesions can be hypoechoic or hyperechoic, with either ill-defined or well-defined borders (16). These lesions may show increased vascularity on color- or power-Doppler examinations. CT also shows variable attenuation depending on the precise contents of the lesion and the degree of fibrosis as compared to surrounding tissue (6). On MRI, IPT usually has low signal intensity on both T1- and T2-weighted images, which may reflect the fibrotic nature of these lesions (16). Contrast-enhanced CT and MRI studies may show a mild homogeneous or heterogeneous enhancement. The enhancement is particularly prominent on delayed images due to the presence of intra-lesional fibrosis.

Urinary system involvement by IPT was initially described in the urinary bladder, which is the most frequent...
Malignancy. The presence of smooth stenosis of the ureter on renal angiography, suggested the diagnosis of urothelial malignancy in view of the focal, abrupt, luminal constriction of the resected surgical specimen. There was suspicion of malignancy present in this case either, and was established after histopathology of the resected ureteric segment were found to be normal.

IPT involvement of the kidneys and the adrenal glands is extremely rare. Renal and adrenal IPT is more common in men and usually presents as flank pain and hematuria (6). The imaging characteristics are nonspecific and may present as a hypoechoic or hyperechoic mass with associated vascularity at color Doppler. These lesions may be hypointense on CT (6, 26). Complete or partial nephrectomy is usually performed in these patients because of the inability to differentiate these masses preoperatively from renal-cell carcinoma on imaging studies. A previously reported case of bilateral renal IPT also highlighted the role of corticosteroids to preserve renal function (27). Adrenal IPT is nonspecific, presenting as a solid mass (6). IPT can also at times present as a paratesticular or epididymal mass. Surgical resection without orchietomy is the ideal treatment for IPT, but in cases with large lesions or testicular involvement, the testicle may have to be removed (20). These tumors often behave in an indolent manner and do not recur after complete surgical excision.

The imaging characteristics of IPT are nonspecific and often mimic other entities, including tumors (29). Though it is often difficult to suspect or diagnose IPT on imaging, diagnosis can be established preoperatively with biopsy of the lesion. The possibility of IPT should be included in the differential diagnosis of these lesions when the diagnosis is not certain. Image-guided biopsy should be advised to establish the preoperative diagnosis and to avoid unnecessary radical surgery in these cases.

Figure 6. Case 2: 47-year-old male with IPT. On pathology, curled muscle fibers and fibromyxoid stroma are seen on low-power magnification. High-power magnification shows muscle and clear fibromyxoid stroma within the ureteric wall. The margins of the resected ureteric segment were found to be normal.

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