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

Journal
Radiology Case Reports, 7(3)

ISSN
1930-0433

Authors
Ghonge, Nitin P
Mittal, Disha
Jain, Sudhir
et al.

Publication Date
2012

DOI
10.2484/rcr.v7i3.742

Peer reviewed

Radiology Case Reports

Volume 7, Issue 3, 2012

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

Nitin P. Ghonge, MD, DNB; Disha Mittal, MBBS; Sudhir Jain, MD; Bharat Aggarwal, MD, DNB; Mariam Moshiri, MD; Chandana Lall, MD; and Puneet Bhargava, MD

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.

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

Citation: Ghonge NP, Mittal D, Jain S, Aggarwal, B. Moshiri M, Lall C, Bhargava P. Urologic manifestations of inflammatory pseudotumor: Report of 2 cases and review of the literature. *Radiology Case Reports*. (Online) 2012;7:742

Drs. Ghonge and Aggarwal are consulting radiologists, and Dr. Mittal is a resident, at the Diwan Chand Satyapal Aggarwal Imaging Research Centre, New Delhi, India. Dr. Jain is a Senior Consultant Pathologist at Oncquest Laboratories, New Delhi, India. Dr. Lall is Section Chief of Abdominal Imaging in Radiological Sciences and Professor of Clinical Radiology, University of California at Irvine, Orange CA. Drs. Bhargava and Moshiri are Assistant Professors in the Department of Radiology, University of Washington, Seattle WA. Contact Dr. Ghonge at drnitinghonge@rediffmail.com.

Competing Interests: The authors have declared that no competing interests exist.

DOI: 10.2484/rcr.v7i3.742

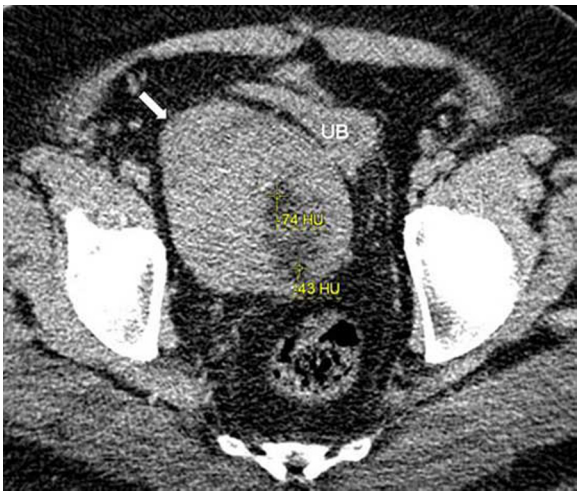

Figure 1. Case 1: 44-year-old male with IPT. Noncontrast CT axial image shows a large, mildly hyperdense mass (arrow) with foci of negative attenuation suggestive of intralesional fat (point ROIs with attenuation values shown). The urinary bladder is displaced to the left side. [UB: Urinary bladder].

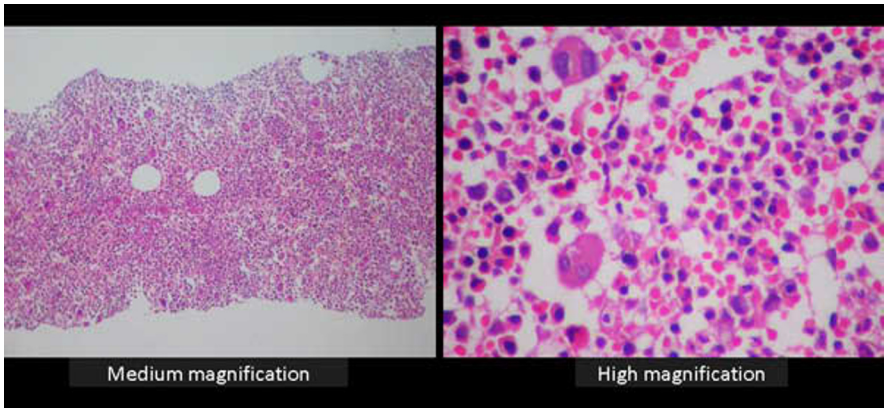

Figure 2. Patient 1: 44-year-old male with IPT. Photomicrographs show mononuclear and polymorphonuclear cells on low-power magnification. Large multinucleated cells are scattered throughout. High-power magnification view shows polymorphonuclear cells, plasma cells, and histiocytes, consistent with IPT.

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 intralesional 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 intralesional 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.

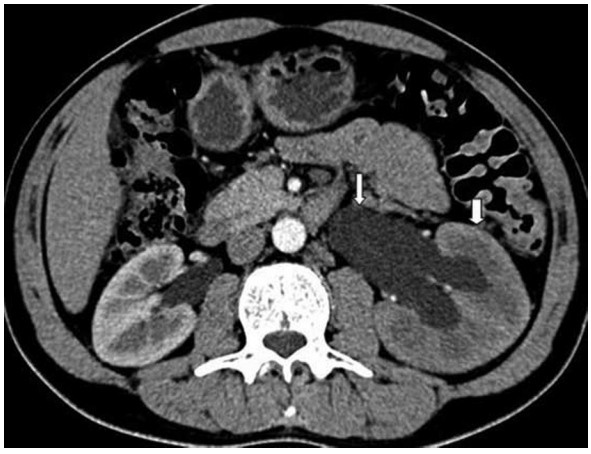

Figure 3. Case 2: 47-year-old male with IPT. Contrast-enhanced CT axial image shows the left-sided hydroureteronephrosis (thin arrow) with asymmetric decreased renal parenchymal enhancement (thick arrow). The right kidney is normal.

Case 2

A 47-year-old male presented with painless hematuria of one weeks' 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 spindled 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-

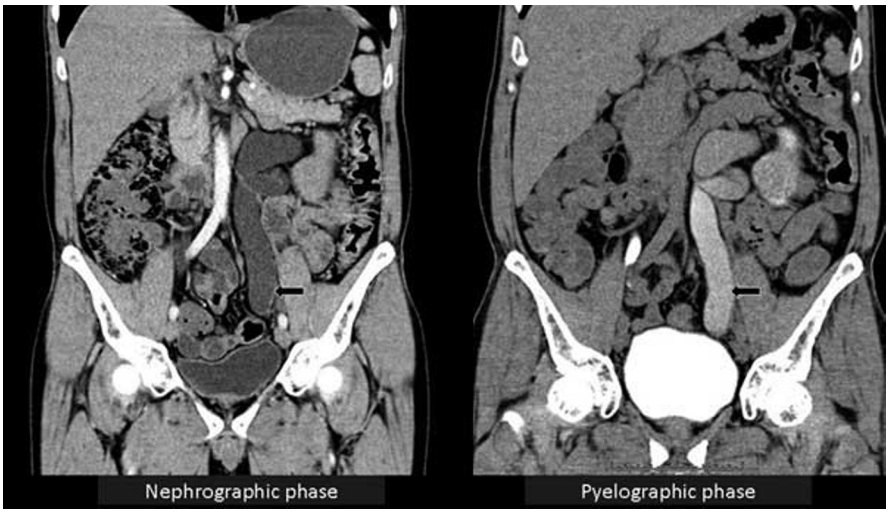

Figure 4. Case 2: 47-year-old male with IPT. Contrast-enhanced CT coronal images in nephrographic and pyelographic phases show a dilated tortuous left ureter (arrow) in the proximal and mid segments.

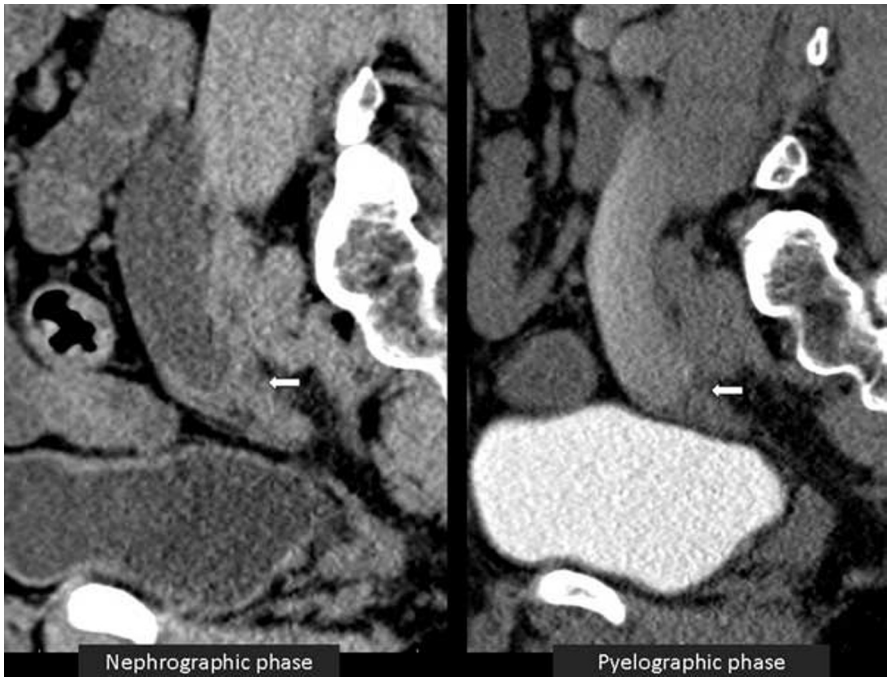

Figure 5. Case 2: 47-year-old male with IPT. Contrast-enhanced CT sagittal images in nephrographic and pyelographic phases show a distal ureteric stricture caused by a circumferential mass (arrow). The mass also shows mild contrast enhancement in the nephrographic phase.

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

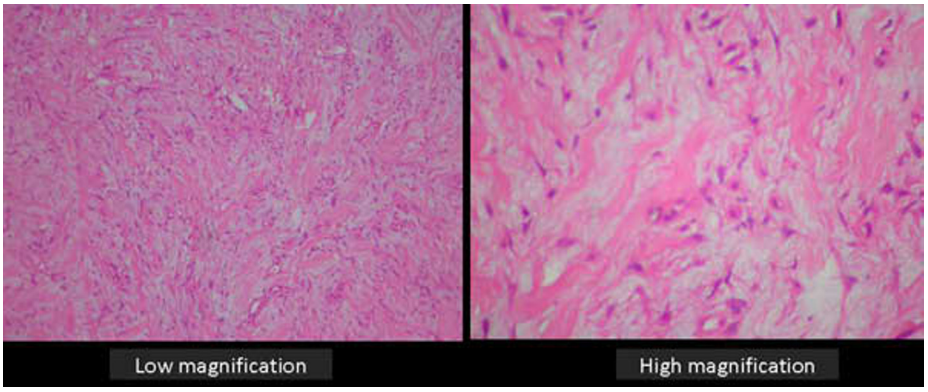

Figure 6. Case 2: 47-year-old male with IPT. On pathology, curled muscle fibers and fibromyxoid stroma are seen on low-power magnification. High-power magnification shows muscle and clear fibromyxoid stroma within the ureteric wall. The margins of the resected ureteric segment were found to be normal.

site for this disease in the urinary system (18). Painless gross hematuria from exophytic and ulcerated lesions is the most common initial manifestation and may progress to anemia (19). Increased frequency of urination, dysuria, and signs of urinary-tract obstruction can also occur. IPT should be considered when a clot, particularly in young adults, surrounds an enhancing tumor. Bladder IPT is rare in children, typically occurs in young adults, and is more common in women than in men (6). In childhood, rhabdomyosarcoma is an important differential diagnosis. Bladder IPT can have various presentations, including a polypoid enhancing intraluminal or submucosal mass lesion, with or without perivesicular fat involvement (20). Conservative surgery is often the standard of care. In patients with extensive bladder-neck involvement and ureteric involvement, preservation of bladder functions is often difficult. Preoperative treatment with a cyclo-oxygenase 2 inhibitor may be helpful to shrink the tumor before resection in these select cases (21). Spontaneous regression of IPT of the urinary bladder has also been described (22).

IPT in the urogenital tract has also been reported in an extravesical location (23), as in our first case. The reported patient presented with nonspecific urinary symptoms, possibly related to the extrinsic bladder compression. IPT was not suspected on imaging and was diagnosed only on biopsy. IPT of the ureters presenting as an ill-defined mass causing ureteric stenosis was described on CT in two previous case reports (24, 25). In our second case, the patient presented with gross hematuria and a distal ureteric stricture. The diagnosis of IPT was not suspected on imaging in this case either, and was established after histopathology of the resected surgical specimen. There was suspicion of malignancy in view of the focal, abrupt, luminal constriction with enhancing circumferential soft-tissue thickening in the left distal ureter. In the case reported by Hosokawa et al (26), the irregular margin of the mass on CT and MRI images, along with neovascularization and tumor staining on renal angiography, suggested the diagnosis of urothelial malignancy. The presence of smooth stenosis of the ureter on retrograde pyelography, however, suggested a benign process. Even MR urography revealed that the mass was engulfing the ureter rather than invading its walls and lumen on retrospective review.

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 hypoattenuating 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 orchiectomy is the ideal treatment for IPT, but in cases with large lesions or testicular involvement, the testicle may have to be removed (28). 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.

References

1. Narla LD, Newman B, Spottswood SS, Narla S, Kolli R. Inflammatory pseudotumor. *RadioGraphics* 2003;23:719-729. [PubMed]
2. Agrons GA, Rosado-de-Christenson ML, Kirejczyk WM, Conran RM, Stocker JT. Pulmonary inflammatory pseudotumor: Radiologic features. *Radiology* 1998;206:511-518. [PubMed]
3. Stark P, Sandbank JC, Rudnicki C, Zahavi I. Inflammatory pseudotumor of the heart with vasculitis and venous thrombosis. *Chest* 1992:102:1884-1885. [PubMed]
4. Torzilli G, Inoue K, Midorikawa Y, Hui A, Takayama T, Makuuchi M. Inflammatory pseudotumors of the liver: prevalence and clinical impact in surgical patients. *Hepatogastroenterology* 2001;48:1118-1123. [PubMed]

5. Sanders BM, West KW, Gingalewski C, Engum S, Davis M, Grosfeld JL. Inflammatory pseudotumor of the alimentary tract: clinical and surgical experience. *J Pediatr Surg* 2001;36:169-173. [PubMed]
6. Park SB, Cho KS, Kim JK, et al. Inflammatory pseudotumor (myoblastic tumor) of the genitourinary tract. *AJR* 2008;191:1255-1262. [PubMed]
7. Bhargava P, Dighe MK, Lee JH, Wang CL. Multimodality imaging of ureteric disease. *Radiologic Clinics of North America* 2012;50:271-299. [PubMed]
8. Taneja R, Bhargava P, Cuevas C, Dighe MK. Common and less-common renal masses and masslike conditions. *Radiologic Clinics of North America* 2012;50:245-257. [PubMed]
9. Mirochnik BD, Bhargava P, Dighe MK, Kanth N. Ultrasound evaluation of scrotal pathology. *Radiologic Clinics of North America* 2012;50:317-332. [PubMed]
10. Dighe M, Bhargava P, Wright JL. Urinary bladder masses: Review of techniques, imaging spectrum, and staging. *Journal of Computer Assisted Tomography* 2011;35:411-424. [PubMed]
11. Umiker WO, Iverson LC. Post-inflammatory tumor of the lung: report of four cases simulating xanthoma, fibroma or plasma cell granuloma. *J Thorac Surg* 1954;28:55-62.
12. Dehner LP. The enigmatic inflammatory pseudotumors: the current state of our understanding, or misunderstanding (editorial). *J Pathol* 2000;192:277-279. [PubMed]
13. Scott L, Blair G, Taylor G, Dimmick J, Fraser G. Inflammatory pseudotumors in children. *J Pediatr Surg* 1988;23:755–758. [PubMed]
14. Zen Y, Kitagawa S, Minato H, et al. IgG4-positive plasma cells in inflammatory pseudotumor (plasma cell granuloma) of the lung. *Human Pathology* 2005;36;710-17. [PubMed]
15. Zen Y, Fujii T, Sato Y, Masuda S, Nakanuma Y. Pathological classification of hepatic inflammatory pseudotumor with respect to IgG4-related disease. *Modern Pathology* 2007;20:884–894. [PubMed]
16. Lim JH, Lee JH. Inflammatory pseudotumor of the liver: Ultrasound and CT features. *Clin Imaging* 1995;19:43–46. [PubMed]
17. Kelekis NL, Warshauer DM, Semelka RC, Eisenberg LB, Woosley JT. Inflammatory pseudoturmo of the liver: Appearance on contrast enhanced helical CT and dynamic MR images. *J Magn Reson Imaging.* 1995;5:510-553. [PubMed]
18. Roth JA. Reactive pseudosarcomatous response in urinary bladder. *Urology* 1980;16:633-637. [PubMed]
19. Inoue H, Iwabuchi K, Kuwao S, Kasai K, Furuhata M, Kameya T. A case report of inflammatory pseudosarcoma of the urinary bladder. *Acta Pathol Jpn* 1992;42:760-765. [PubMed]
20. Fuiwara T, Sugimura K, Imaoka I, Igawa M. Inflammatory pseudotumor of the bladder: MR findings. *J Comput Assist Tomogr* 1999;23:558–561. [PubMed]
21. Berger A, Kim C, Hagstrom N, Ferrer F. Successful preoperative treatment of pediatric bladder inflammatory myofibroblastic tumor with anti-inflammatory therapy. *Urology* 2007;70:e313–e375. [PubMed]
22. Mochizuki Y, Kanda S, Nomata K, et al. Spontaneous regression of inflammatory pseudotumor of the urinary bladder. *Urol Int* 1999;63:255-257. [PubMed]
23. Jimenez JM, Poustchi-Amin M, Leonidas JC, Pena A. Extraperitoneal abdominopelvic inflammatory pseudotumor: report of four cases. *Pediatr Radiol* 1997;27:170–174. [PubMed]
24. Nozawa M, Namba Y, Nishimura K, Sugao H. Inflammatory pseudotumor of the ureter. *J Urol* 1997;157:94. [PubMed]
25. Hosokawa C, Tsubakimoto M, Inoue Y, Goto T, Senju M, Yoshimura R. Radiologic features of inflammatory pseudotumor of the ureter. *AJR* 2007;188:W202–W205. [PubMed]
26. Tarhan F, Gul AE, Karadayi N, Kuyumcuoglu U. Inflammatory pseudotumor of the kidney: A case report. *Int Urol Nephrol* 2004:36:137-140. [PubMed]
27. Ma Y, Zieske AW, Fenves AZ. Bilateral infiltrating renal inflammatory pseudotumor responsive to corticosteroid therapy. *Am J Kidney Dis* 2008;51:116–120. [PubMed]
28. Tunuguntla H, Mishra A, Jorda M, Gosalbez R. Inflammatory myofibroblastic tumor of the epididymis: case report and review of the literature. *Urology* 2011;78:183–185. [PubMed]
29. Patnana M, Sevrukov AB, Elsayes KM, Viswanathan C, Lubner M, Menias CO. Inflammatory pseudotumor: The great mimicker. *AJR* 2012;198:W217-W227. [PubMed]