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GENERAL UROLOGY

Case Report

Isolated flat desmoid-type terminal ileal mesenteric fibromatosis masquerading as lower ureteric stricture-an intraoperative surprise

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ABSTRACT

Ureteral strictures are usually caused by ureteral calculi, endoscopic instrumentation, infections like tuberculosis, surgical dissection, radiation, malignancy and periureteral fibrosis. A 42-year-old man presented with right loin pain 2 months previously. Contrast-enhanced computed tomography showed right lower ureteral stricture and he was scheduled for right ureteral reimplantation. Intraoperatively, a large hard mass arising from the mesentery of the terminal ileum infiltrating the retroperitoneum and encasing the right external iliac artery, vein and ureter was identified. The mass was resected and psoas hitch was performed over a 5F (26 cm) double J stent. Patient was discharged on tenth postoperative day and is still doing well at 18 months of follow up period. We are reporting this case to highlight the rare possibility of mesenteric fibromatosis and its management.

Keywords: Desmoid; mesenteric fibromatosis; ureteral stricture.

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Introduction

The common causes for ureteral stricture include ureteral calculus, endoscopic instrumentation, infections commonly tuberculosis, surgical dissection, radiation, malignancy and periureteral fibrosis.[1,2] Mesenteric fibromatosis (MF) or desmoids are rare causes of ureteral obstruction and they are usually associated with familial adenomatous polyposis (FAP).[3] Though cross-sectional imaging was performed, this etiology could not be revealed and it was an intraoperative surprise. We report the management of ureteral obstruction due to desmoids.

Case presentation

A 42-year-old man presented with right loin pain starting 2 months ago. He did not have fever, vomiting or lower urinary tract symptoms. His past history was unremarkable. His serum creatinine was 1 mg/dL, results of other blood and urine tests were within normal limits. Contrast-enhanced computed tomogram (CECT) showed dilated right renal pelvis, tortuous dilated ureter with severe narrowing in the lower one-third of the ureter (Figure 1). Left kidney was normal. Tc^{99m} diethylene triamine pentaacetic acid renogram suggested right ureteral obstruction with 36.37% renal function (glomerular filtration rate: 27.42 mL/min/1.73 m^2) of the right kidney. He had undergone colonoscopy for long standing constipation that showed few ulcers and erosions in the rectum. Rectal biopsy revealed nonspecific proctitis.

Patient was taken up for right ureteral reimplantation. Cystoscopy showed bladder with good capacity and normal ureteral orifices. Retrograde pyelography showed stricture at the level of upper border of sacroiliac joint. Right modified Gibson incision was made. Intraoperatively, a large hard mass arising from the mesentery of the terminal ileum infiltrating the retroperitoneum and encasing the right external iliac artery, vein and ureter was identified. Resection of the terminal ileum, cecum along

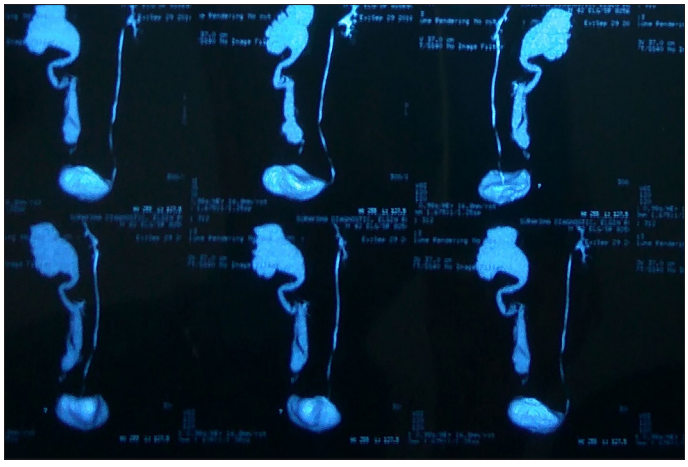

Figure 1. Contrast-enhanced computed tomogram of the abdomen showing dilated right renal pelvis, tortuous dilated ureter with abrupt narrowing in the lower one-third of the ureter

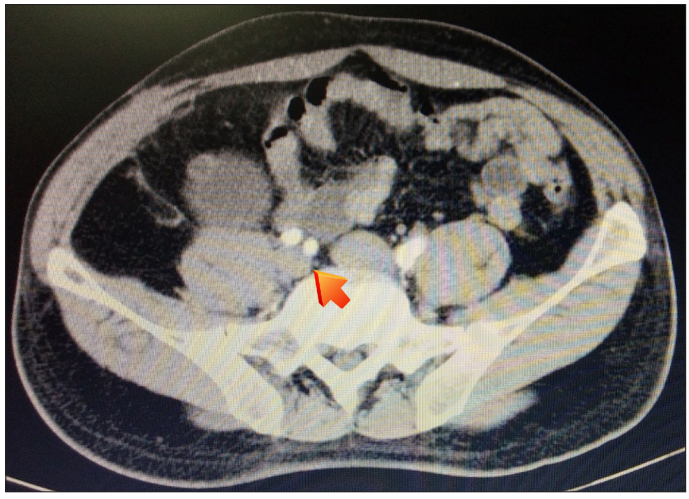

Figure 3. Contrast enhanced computed tomogram of the abdomen (arterial phase) showing no evidence of recurrence of mesenteric fibromatosis at the level of bifurcation of common iliac artery

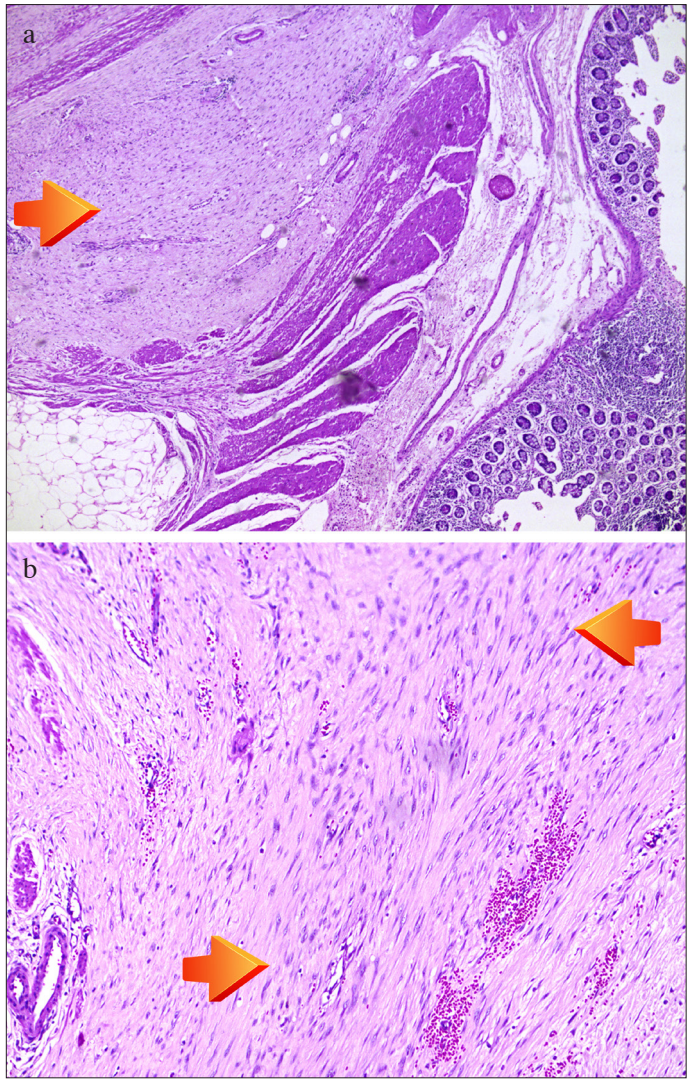

Figure 2. a, b. Photomicrographs. Spindle-shaped tumor cells arranged in long sweeping fascicles with elongate, vesicular nuclei, moderate amounts of eosinophilic cytoplasm and much intercellular collagen and mitotic activity of 1/10 high power field H&E, (a). 40X magnification. (b). 100X magnification

with mesentery was performed and end to side ileocolonic anastomosis was done. Bladder was mobilized and psoas hitch was performed over a 5F (26 cm) double J stent. Bladder was closed with 3-0 polyglactin over perurethral and suprapubic catheters. Catheters were removed on the tenth postoperative day and the patient recovered well.

On gross examination, tumor measured around 7x3.8x2.5 cm, involving mesentery and wall of terminal ileum. No polyps were seen and appendix was normal. On microscopy, tumor was composed of long sweeping fascicles of spindle shaped cells with elongate, vesicular nuclei, moderate amounts of eosinophilic cytoplasm and lot of intercellular collagen. Tumor cells were arranged in sheets and composed of spindle shaped cells with plump nuclei resembling fibroblasts and mitotic activity per 1/10 high power field (Figure 2). On immunohistochemistry, tumor was negative for CD117, CD34, smooth muscle actin, S100 and β-catenin, suggestive of MF. Double J stent was removed after 6 weeks. Patient is still doing well at 18 months of follow-up and repeat CECT showed no recurrence of MF (Figure 3).

Discussion

Mesenteric fibromatosis is a locally invasive tumor and it does not metastasize.[3] The term "fibromatosis" or desmoid tumor includes clonal fibroblastic proliferations characterized by infiltra-

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tive growth with a tendency to local recurrence and an inability to metastasize.[4-6] Desmoids account for 0.03% of all malignancies. The most common site of involvement is the mesentery of the small bowel, but may also occur in the retroperitoneal area and abdominal wall aponeurosis.[3] Desmoids may rarely cause obstructive uropathy due to upper urinary tract obstruction.[7] Mesenteric fibromatosis can occur spontaneously, after surgical trauma or with FAP.[3] Fibromatosis behaves like malignancy in that they are infiltrative and has a tendency to recur. But contrary to the features of malignancy, it does not usually metastasize.[3,4]

Diagnosis of MF is based on clinical suspicion.[3] The differential diagnosis for MF includes gastrointestinal stromal tumors (GIST), retroperitoneal fibrosis, sclerosing mesenteritis, solitary fibrous and inflammatory myofibroblastic tumors.[5] Cross-sectional imaging is mainly performed to assess local extent, tumor relation to neurovascular structures and resectability than for confirmation of diagnosis.[3] On CECT, MF appears as an irregular, infiltrating, and non-enhancing mass. On magnetic resonance imaging (MRI), desmoids appear hypointense to muscle on T1-weighted images and appear as variable signal intensity on T2 weighted images. There are no specific imaging features that can help differentiating desmoids from other solid tumors. Though it is difficult to diagnose isolated desmoids on imaging, MF should be considered in the differential diagnosis of an infiltrating right iliac fossa mass.[3,8]

Desmoids are frequently found in patients with FAP.[5,9] Desmoids associated with FAP are more likely to be multiple and they involve the mesentery and abdominal wall up to 3.5-32% of the cases, whereas isolated desmoids were singular and located in the retroperitoneum and pelvis.[5,8] Only 5% of sporadic desmoids are intraabdominal, while 10% of the patients with FAP will develop desmoids and 80% of FAP-associated desmoids develop intraabdominal disease.[5,8] Solitary occurrence in the retroperitoneal space is rare and associated with FAP.[4] Few cases of ureteral obstruction have been reported secondary to MF in FAP which is rarely associated with isolated desmoid.[3,7,8,10,11]

The primary modality of treatment for localized desmoids is complete surgical resection including a margin of normal tissue.[3-5] Apparent cure is possible in majority of cases with sporadic desmoids after surgical resection and morbidity/mortality is mainly related to small bowel resection.[8,12] Surgery should be reserved for the palliation of complications.[8] As MF lacks a definite capsule and infiltrates deeply, complete resection by identifying margins during surgery is not always possible. There is a high rate of postoperative recurrence and may need adjuvant therapy. The aim is to preserve the quality of life of patients, which is threatened by the loss of function and pain due to proliferative disease.[5] The most important strategy in the management of MF is to prevent direct invasion into adjacent tissues.[4]

The forms of adjuvant therapy include postoperative radiotherapy, pharmacotherapy in the form of non steroidal antiinflammatory drugs (NSAIDs), tamoxifen, interferons and chemotherapy.[4,5] Patients with minor symptoms, especially those with FAP or vascular involvement, the least toxic agents like antiestrogens or suldinac (non steroidal anti inflammatory drug) may be preferable to resection.[8] Preoperative radiotherapy has been advocated as well.[4] Neoadjuvant chemoradiotherapy prior to surgical resection is promising, though only small number of patients in this study received this treatment.[8,13] Only 10% of desmoids progress rapidly, hence watchful waiting appears to be the most appropriate management in select cases where tumors could not be completely resected.[5,9]

Histopathological examination is done to assess margins and the completeness of resection. The most important pathologic differential diagnosis for MF is GIST which is mainly submucosal. GIST can grow to large size, infiltrate adjacent structures similar to MF, but have increased mitotic activity. GIST is positive for CD117 while β-catenin is positive in MF. Also, absence of CD34 and S100 expression favors diagnosis of MF.[3] However our patient was negative for β-catenin. Although nuclear staining for β-catenin is highly specific, the intensity of β-catenin and the percentage of nuclear staining do not influence the diagnosis of desmoids much.[3,9]

Desmoids may cause ureteral obstruction, even if no obvious tumors are observed because flat desmoids associated with FAP do not present as masses. Desmoid precursor lesions reported as mesenteric plaques in FAP patients can cause ureteral obstruction.[9] Our patient had undergone CECT but it failed to demonstrate desmoid hence it was an intraoperative surprise. Desmoids causing ureteral obstruction were mostly in association with FAP and intraabdominal desmoids were treated with unilateral, bilateral or partial resections. Surgical options included ureteral stenting or percutaneous nephrostomy, ureterolysis, ureteral resection, nephroureterectomy, ureteral resection and renal autotransplantation. Autotransplantation done for ureteral stent obstruction has an excellent renal function at 2 years.[7]

In conclusion, isolated desmoid tumor is a rare cause of ureteral obstruction. It should be considered in the differential diagnosis of an infiltrative mass with hydroureteronephrosis. Complete surgical resection is the treatment of choice however it is difficult in all cases. Adjuvant therapy should be individualized and patient should be kept on long-term follow-up.

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Processing – A.K., J.V., V.S.K., M.K.; Analysis and Interpretation – A.K., J.V., V.S.K., M.K.; Literature Search – A.K., J.V., V.S.K., M.K.; Writing Manuscript – A.K., J.V., V.S.K., M.K.; Critical Review – A.K., J.V., V.S.K., M.K.

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