Primary Ureteral Lymphoma Presenting with Acute Flank Pain

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Key Words
Ureteral lymphoma • Ureteral carcinoma

Abstract
Non-Hodgkin’s lymphoma (NHL) represents 4% of newly diagnosed cancer in 2013 with a 59–82% 5-year survival depending on the tumor location. Primary presentation of lymphoma consists of lymphadenopathy or swelling of the lymph nodes and non-specific systemic symptoms such as fevers, night sweats, and weight loss. Less commonly, NHL arises from non-lymphoid tissue. We report a unique case of NHL arising from the ureteral wall which was visualized via non-contrast CT and direct vision through ureteroscopy.

Case Report

A 28-year-old male with a past medical history of sickle-cell trait and tobacco use, presented to the emergency room with nausea and sharp right-sided flank pain. Laboratory values revealed an elevated serum creatinine of 1.7 mg/ml. Urinalysis and all other laboratory values were unremarkable. Non contrast CT of the abdomen and pelvis revealed an enlarged right kidney with moderate hydronephrosis and blunted calyces. The proximal
right ureter was fluid filled and mildly dilated measuring 4 mm further dilating in its mid portion to 12 mm (fig. 1). At its widest portion the ureter was filled with an ill-defined soft-tissue density mass with peripheral punctuate calcification. The right ureter distal to this lesion was completely decompressed (fig. 2). No renal, ureteral or bladder calculi were present and no other pathologic abnormalities noted on CT scan.

The patient subsequently underwent cystoscopy and diagnostic retrograde pyelogram. A right mid-ureteral obstruction was identified which prevented the passage of any contrast dye. The obstructing lesion was an irregular intraluminal filling defect demonstrating a “goblet” sign, consisting of focal dilatation of the ureter around and slightly below the mass (fig. 3). After failing to pass a standard 0.035 guidewire into the renal pelvis, ureteroscopy

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**Fig. 2.** CT pelvis revealing the dilated ureter and soft tissue mass. Also of note are the small calcifications.

**Fig. 3.** Goblet sign. Retrograde pyelogram demonstrating filling defect in right mid-ureter.

**Fig. 4.** a Ureteroscopic image of the lymphoma; b Ureteroscopic image of the lymphoma.
was performed which identified an obstructing mass in the right mid ureter (fig. 4). A guide wire was then passed into the kidney, and multiple biopsies of the mass were obtained. A retrograde pyelogram was completed and a 6-french ureteral stent was left in place to relieve the obstruction. The remainder of the ureter and renal collecting system were normal. Pathology at that time was positive for proliferation of large cells which stained positive for CD5 suggesting T-cell origin but inconclusive for a specific diagnosis.

One month later, the patient underwent repeat ureteroscopy which again revealed a papillary, bulbous, avascular mass which was approximately 3 cm in length. Holmium laser excision was performed with careful sparing of the ureteral mucosa. Biopsy histology confirmed the mass to be ALK positive anaplastic large-cell lymphoma (fig. 5). PET/CT performed prior to treatment demonstrated the right ureteral mass to be mildly FDG avid with an SUV of 2. Two FDG avid lymph nodes, one 1.6 cm node in the right groin and one 0.9 cm node in the retroperitoneum, were identified and the patient was staged as IVa.

**Discussion**

The estimated incidence of any type of ureteral malignancy in 2013 is less than 0.1% of all cancers, with urothelial carcinoma by far the most common primary ureteral malignancy [2]. Metastatic disease to the ureter is relatively rare with the most common primary sites including: melanoma, breast, prostate, cervix and colon cancer and may be bilateral in up to 30% of cases [4]. Most metastases are discovered at autopsy. Non-Hodgkin lymphoma arising primarily from the ureteral wall is exceedingly uncommon, with fewer than 20 reported cases [1, 3, 5–14]. When lymphoma affects the ureters it is usually as indirect displacement by bulky retroperitoneal adenopathy [15]. Furthermore, anaplastic large-cell lymphoma is a less common variant comprising less than 15% of all cases of non-Hodgkin lymphoma in adults [16]. To our knowledge it has never been documented originating from the ureter.

Anaplastic large-cell lymphoma, most frequently presents with painless lymphadenopathy. On rare occasions, patients can present with symptoms secondary to tumor mass effect [17]. In the reported cases of ureteral lymphoma patients complain of either vague flank pain or abnormal renal function secondary to the indolent obstruction. In the majority of these reports hydronephrosis was caused by encasement of the ureter by lymphomatous deposits or diffuse infiltration of the ureteral wall. Because mucosal involvement is so rare, patients do not usually present with hematuria [3].

The unique features of this case include the presentation, imaging findings and unusual pathology. When...
lymphoma involves the ureters its source is either direct invasion or hematogenous spread to the adventitia. In our case at initial presentation the only site of lymphoma was an intra-luminal ureteral mass. The initial CT demonstrated the classic findings of an acute ureteral obstruction which, uncharacteristically in a young adult, was due to a mass rather than a calculus. The retrograde pyelogram demonstrated an intraluminal ureteral mass more typical of an urothelial malignancy. Lastly, the pathologic type of lymphoma was a less common variant of non-Hodgkin’s lymphoma.

Conclusion

In summary, primary ureteral lymphoma of any variety is extremely rare. To our knowledge, this is the first reported case of anaplastic large-cell lymphoma arising primarily from the ureter in a patient presenting with acute obstructive uropathy.

References