Ureteropelvic junction obstruction caused by metastatic cholangiocarcinoma

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ABSTRACT

We describe the rare case of a 61-year-old female with right ureteropelvic junction (UPJ) obstruction caused by metastatic cholangiocarcinoma. Her past medical history was notable for cholangiocarcinoma treated with neoadjuvant chemoradiation and two orthotopic liver transplants six years earlier. Urology was consulted when she presented with flank pain and urinary tract infection. Diagnostic workup demonstrated right UPJ obstruction. She was managed acutely with percutaneous nephrostomy. She subsequently underwent robotic pyeloplasty and intrinsic obstruction of the UPJ was discovered. Histological examination revealed adenocarcinoma, consistent with systemic recurrence of the patient’s known cholangiocarcinoma.

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INTRODUCTION

Obstruction of the ureteropelvic junction (UPJ) is a common urological problem that can result in hydronephrosis and deterioration of renal function. Although most commonly observed in pediatric populations, UPJ obstruction in adults can arise from a number of etiologies, including congenital causes, acquired stenosis due to urolithiasis, urothelial malignancy, and other retroperitoneal disease processes (1, 2). Intraluminal malignant obstruction due to metastatic involvement of the UPJ, however, is exceedingly rare, with few case reports existing in the literature (3, 4). Herein we describe the case of a patient with a history of hilar cholangiocarcinoma treated with a liver transplant, whose systemic recurrence presented as an isolated right UPJ obstruction six years later.
CASE REPORT

A 61-year-old female presented to the emergency department with right flank pain, fever, chills, nausea, and vomiting. Urinalysis showed positive nitrates, moderate leukocyte esterase, 4+ bacteria, and >100 white blood cells on microscopy. An abdominal/pelvic CT scan was obtained which revealed moderate-to-severe right hydronephrosis with delayed right nephrogram, concerning for high-grade right UPJ obstruction (Figure-1). No associated mass, lymphadenopathy, calculus, or other cause of obstruction was identified. Her past medical history was notable for hilar cholangiocarcinoma treated with neoadjuvant chemoradiation followed by two orthotopic liver transplants six years prior with no interval evidence of recurrence or metastasis.

The patient was admitted to the hospital and treated with intravenous antibiotics for acute obstructive pyelonephritis. A retrograde pyelogram was performed which revealed a dilated collecting system and features concerning for UPJ configuration (Figure-2), and a right ureteral stent was placed under fluoroscopy prior to discharge.

One month later, the patient presented again to the emergency department with continued right flank pain. CT abdomen/pelvis revealed persistent hydronephrosis, necessitating placement of a right percutaneous nephrostomy tube. During subsequent follow-up, diuretic renal scintigraphy performed with the nephrostomy tube clamped demonstrated persistent high-grade right-sided obstruction. The patient was counseled on treatment options and elected to undergo a right retroperitoneal robotic-assisted laparoscopic pyeloplasty. At this time, there was no suspicion of malignant obstruction based upon preoperative imaging, and the UPJ obstruction was presumed to have been either congenital or due to retroperitoneal fibrosis caused by the patient’s previous liver transplants.

Intraoperatively, dissection of the lower pole of the right kidney revealed a moderate degree of fibrosis. Amidst the fibrosis, the ureter was identified and followed cephalad to the dilated renal pelvis. The UPJ was dissected and the renal pelvis was opened, revealing an obstructive intrinsic papillary lesion at the UPJ, which was biopsied and sent for frozen section. Histopathological analysis of the frozen section revealed findings concerning for adenocarcinoma. The UPJ area was then resected back to what appeared to be uninvolved mucosa. A dismembered pyeloplasty was performed and a right ureteral stent was placed.

The UPJ and periureteral fibrotic tissue were sent for permanent section. Histopathologic sections of all biopsy specimens revealed infiltrating, atypical glands lined by columnar tumor cells with nuclear and cytoplasmic abnormalities (Figure-3). The morphological consistency with the patient’s known cholangiocarcinoma, combined with the patient’s history, were concerning for a metastatic cholangiocarcinoma recurrence at the UPJ.

Figure 1 - Coronal (a) and axial (b) contrast-enhanced computed tomography revealed moderate-to-severe right hydronephrosis with decreased nephrogram of the right kidney, concerning for high-grade right UPJ obstruction.
Upon histopathologic diagnosis of her ureteral metastases, the patient was started on a regimen of gemcitabine and cisplatin, which is currently considered the reference regimen for advanced biliary cancer (5). Both medications were subsequently discontinued after five months due to fatigue and poor patient tolerance. Her right UPJ obstruction has persisted since surgery, presumably due to persistent cholangiocarcinoma within the retroperitoneum observed on surveillance CT scans. This has been managed with percutaneous nephrostomy drainage as she has not tolerated ureteral stents. Interestingly, at 1-year, there remains no radiographic evidence of disease progression.

DISCUSSION

While malignancies of the genitourinary system are common, it is estimated that only 1.6%–3.0% represent secondary tumors (6). Of these, most present either in the kidney or bladder. There have
been many reported cases of metastatic spread from the pancreas, stomach, breast, prostate, lung, and colon presenting initially as ureteral obstruction (7–13). The vast majority of these cases present as extrinsic obstruction secondary to retroperitoneal lymphadenopathy. Secondary malignancies causing intrinsic blockage and presenting as isolated UPJ obstructions, on the other hand, are extremely rare in the literature. Naranji et al. (4) reported a case of right-sided UPJ obstruction in a 78-year-old woman who underwent laparoscopic pyeloplasty that showed mantle cell lymphoma on final histology. The only reported observation of malignant infiltration of the UPJ by a solid organ cancer is a case of recurrent breast cancer reported by Shah et al. (3) in 2016. Importantly, to the best of our knowledge, this case report is the first to describe metastasis of a primary biliary tract malignancy to the UPJ.

CONCLUSION

We describe a rare presentation of metastatic cholangiocarcinoma causing intrinsic UPJ obstruction. In patients with a history of malignancy, physicians should consider metastatic relapse in the differential diagnosis for acquired UPJ obstruction. Endoscopic evaluation may be helpful in both diagnosis and the planning of surgical intervention.

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CONFLICT OF INTEREST

None declared.

REFERENCES


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