

Pyeloduodenal Fistula in Xanthogranulomatous Pyelonephritis: A Series of Two Cases

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ABSTRACT

Xanthogranulomatous inflammation, characterized by destruction and replacement of tissues with chronic inflammatory cells, including foamy histiocytes and hemosiderin-laden macrophages, is uncommon. In patients with xanthogranulomatous pyelonephritis, inflammation may extend from the kidney to the overlying duodenum, creating a pyeloduodenal fistula that further complicates medical and surgical management. We present two cases with recurrent kidney infections who each ultimately received a nephrectomy and repair of their duodenal fistula.

INTRODUCTION

Pyelointestinal fistulas, or false passages between kidney urothelium and bowel mucosa, are a rare phenomenon first reported in 1893, with only 80 cases reported to date.^{1,2} These fistulas are more commonly associated with malignancies, trauma (eg, ingesting fish bones), renal calculi, and other inflammatory processes. In a setting of xanthogranulomatous pyelonephritis (XGP), which is an unusual variant of chronic pyelonephritis, normal kidney parenchyma is replaced by lipid-laden macrophages.³ The inflammation may lead to development of pelocolonic and rarely pyeloduodenal fistulas that often fail medical management. We present two cases with XGP and pyeloduodenal fistulas that each required surgery for definitive treatment.

CASE PRESENTATIONS

Case 1

A 52-year-old woman presented to the Emergency Department with hyperlipidemia and a history of recurrent urinary tract infections. She reported 3 weeks of persistent and worsening right

flank pain and swelling, with associated hematuria, fever, headache, and nausea. Laboratory work-up revealed leukocytosis at 12,200/ μ L, an elevated platelet count at 490,000/ μ L, and positive urinalysis and urine culture but otherwise normal creatinine at 0.58 mg/dL (glomerular filtration rate [GFR] 106).

With a presumed initial diagnosis of a urinary tract infection, our patient was treated with intravenous antibiotics including clindamycin and piperacillin/tazobactam. This treatment failed to improve her symptoms, prompting further urologic work-up. A computed tomography scan identified what was initially thought to be a large angiomyolipoma with a staghorn calculus filling the right renal collecting system, but the scan failed to visualize a viable right renal parenchyma (Figure 1). No air was seen in the urinary tract and conversely no extravasation of orally administered contrast was noted from the duodenum into the renal pelvic system, making a preoperative diagnosis of pyeloduodenal fistula unlikely. Initial biopsy of the right kidney revealed multiple granulomas with minimal necrosis, but abundant acute

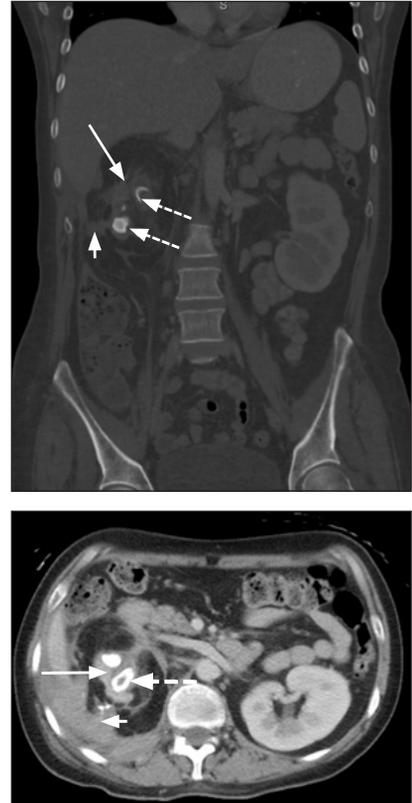


Figure 1. Coronal (top) and axial (bottom) contrast-enhanced computed tomography images of the patient's abdomen from Case 1, showing a sclerotic right kidney (long arrow) and kidney stones (dashed arrows). The psoas abscess (short arrow) can be seen posterior to the kidney stones.

inflammation and fibrosis not typical of XGP. Sampling of the right perinephric fluid revealed no organismal growth, although radiographic findings and

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clinical symptoms were consistent with an intraabdominal infection with associated psoas abscess.

After 2 weeks of antibiotic treatment with no growth from collected cultures and an obvious improvement in clinical symptoms, a right laparoscopic nephrectomy was planned. During the surgery, severe inflammation and tight adherence between the kidney and the duodenum were noted, requiring a conversion to an open procedure. A pinpoint defect with minimal surrounding inflammation was noted in the second part of the duodenum overlying the right kidney. The pyeloduodenal fistula was resected, and the dilated duodenum was primarily repaired in 2 hand-sewn layers without duodenal stricturing. Our patient remained nil per os with nasogastric tube suctioning for 3 days. Her diet was advanced after an upper gastrointestinal swallow study revealed no leak at the repair site. Our patient was discharged 6 days later after an uneventful hospital course. She experienced no further sequelae and was tolerating a regular diet. Creatinine at the time of discharge was 0.45 mg/dL, and at 1-month follow-up creatinine was 0.59 mg/dL (GFR 105).

Pathology of the patient's right kidney using hemotoxylin and eosin staining revealed classic foamy histiocytes consistent with XGP, nephrolithiasis, and perinephric acute and chronic inflammation (Figure 2). Duodenal pathology at the site of the fistula showed muscularis hyperplasia, acute and chronic inflammation, and patchy microabscess formation, but healthy mucosa at the resection margins.

Case 2

A 58-year-old man with hypertension, obesity, nephrolithiasis, and a recent robotic right radical nephrectomy was hospitalized with urosepsis. Physical examination revealed persistent malodorous right flank drainage confirmed on computed tomography scan to be a nephrocutaneous fistula (Figure 3) arising from an incompletely removed right kidney. Imaging further revealed a very large staghorn calculus and no oral contrast extravasation suggestive of pyeloduodenal fistula. Laboratory studies revealed leukocytosis of 23,800/ μ L,

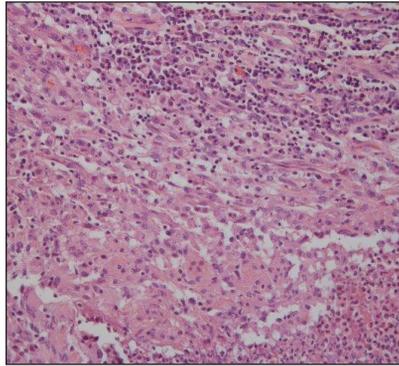


Figure 2. Hemotoxylin and eosin stain (40x magnification) of the biopsy from Case 1 showing pyelonephritis and abscess (lower right corner) with large areas of mononuclear inflammatory cells (center) surrounding areas of necrosis.

anemia with hemoglobin of 11 g/dL, and creatinine which fluctuated between 1.3 mg/dL and 1.9 mg/dL (GFR 41). Review of surgical pathology from a robotic biopsy demonstrated a partly necrotic renal parenchyma with global segmental glomerulosclerosis, chronic tubulointerstitial nephritis, extensive granulation tissue, and nephrolithiasis, none of which were diagnostic of XGP.

Medical management with daptomycin, followed by penicillin and minocycline for culture-proven methicillin-resistant *Staphylococcus aureus* and two different strains of *Streptococcus viridans* was unsuccessful in treating the patient long term because the nephrocutaneous fistula continued to spontaneously recur. Approximately nine months after the patient's initial presentation, we proceeded with a right radical nephrectomy. The operation was extremely difficult. We noted a scarred, dense kidney, tightly adherent to the retroperitoneum posteriorly, to the liver and the adrenal gland cranially, to the inferior vena cava medially, and to the overlying duodenum anteriorly. Interestingly, there were no obvious signs of previous surgical dissection despite operative reports suggesting a radical nephrectomy. During the operation, we noted a dilated second portion of the duodenum with a diverticulum spanning across the upper pole of the kidney. The diverticulum was excised transversely using a linear stapler, and no duodenal fistula was noted at the time.

Within the next 24 hours, our patient developed leukocytosis with white blood cells rising to 18,500/ μ L and obvious bilious drainage (via a surgically placed retroperitoneal Jackson-Pratt drain) requiring exploration. A small, pinpoint duodenal fistula was noted distal to the previously excised duodenal diverticulum. The duodenal defect was excised and primarily repaired in 2 layers without duodenal stricturing. Our patient remained nil per os for 5 days to allow for healing at the duodenal repair site. After an upper gastrointestinal swallow study confirmed no extravasation of oral contrast, his diet was advanced to regular without further sequelae.

Postoperative recovery was complicated by development of a subcutaneous and retroperitoneal abscess requiring drainage and treatment of *Escherichia coli* and *Candida tropicalis* with prolonged course (> 14 days) of intravenous piperacillin/tazobactam and fluconazole. During his 4-week hospitalization, our patient required a short course of hemodialysis but



Figure 3. Axial (top and bottom) computed tomography images of the patient's abdomen from Case 2. The top image shows the nephrocutaneous fistula tract with air (long arrow) and the staghorn calculus (dashed arrow). In the bottom image, the thinning kidney parenchyma (short thick arrow) and fistulous tract (long arrow) can be seen in the retroperitoneum.

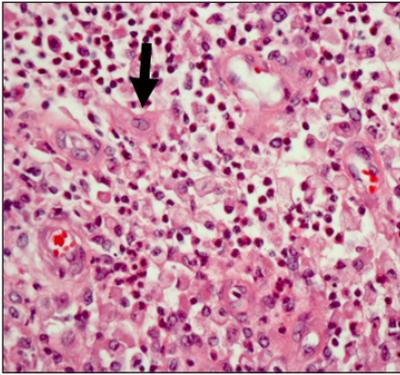


Figure 4. Photomicrograph (Periodic-acid Schiff stain, 400x magnification) of the kidney biopsy from Case 2 showing interstitial infiltrates composed of epithelioid histiocytes/macrophages (black arrow) with positive granular content without Michaelis-Gutman bodies within the cells.

was ultimately discharged with a baseline creatinine of 1.5 mg/dL (GFR 51) and close monitoring by the Nephrology Department.

Pathology of the patient's right kidney using Periodic acid-Schiff staining revealed classic foamy histiocytes consistent with XGP (Figure 4). The kidney had undergone marked tubular atrophy, fibrosis, and interstitial necrosis. There was also severe, near-occlusive atherosclerosis of the renal artery and arterioles, an incidental papillary renal adenoma, and fragmented staghorn calculi embedded within renal parenchyma. No duodenal specimen was analyzed.

DISCUSSION

XGP is an unusual variant of chronic pyelonephritis with an incidence of 1.4 cases/100,000 population per year.⁴ It is almost universally associated with a urinary obstruction, such as congenital ureteropelvic abnormalities or more frequently a calculus. Patients present with vague symptoms of flank pain and tenderness, fever, a palpable mass, or less commonly weight loss, abdominal pain, urinary symptoms, anorexia, and malaise. Laboratory test results may reveal anemia, leukocytosis, elevated alkaline phosphatase, liver enzymes, or creatinine abnormalities.⁵ Low-virulence organisms like *E coli* or *Proteus* species are the most frequently cultured bacteria from urine in XGP. Diagnosis of XGP remains difficult and is most often confused with

renal cell carcinoma in presentation and radiographic appearance, but high suspicion must be maintained. Computed tomography is considered the imaging modality of choice, visualizing staghorn calculi, enlarged or chronically diminishing kidneys that fail to excrete contrast, enhancing rim of tissue suggestive of infections, and occasional extrarenal involvement including pyelointestinal fistulas.³ Biopsy may be inadequate because it can fail to show the classic appearance of granulomatous tissue containing lipid-laden macrophages and small abscesses surrounding renal calculi.⁴ Biopsy may rule out other inflammatory conditions like malakoplakia or megalocytic interstitial nephritis.⁶

The etiology of XGP remains unknown, possibly related to a defect in macrophage processing of bacteria. XGP tends to be unilateral.⁷ Urinary obstruction and chronic kidney infection facilitate complete kidney destruction.⁷ Treatment often requires several courses of antibiotics, which ultimately fail, with surgical removal of the inflammatory nidus, including spleen, pancreas, or bowel, providing definitive treatment.⁸ Laparoscopic removal can be attempted, although given the likelihood of dense adhesions to the surrounding structures and the extensive scarring associated with XGP, conversion to an open procedure may be necessary.⁹

To our knowledge, only 2 previous case reports have described the rare complication of pyeloduodenal fistula in association with XGP.^{5,10} Pyeloduodenal fistulas have historically been divided into 2 groups: Spontaneous and traumatic.¹¹ Nephrolithiasis accounts for the majority of cases of spontaneous nephro-enteric fistulas.¹² In XGP, chronic inflammation, staghorn calculi, and lingering infection probably cause erosion and perforation of the urinary pelvis into the overlying digestive tract.⁵ High suspicion for a pyelointestinal fistula is necessary to identify it preoperatively; radiologic findings of gas bubbles within the bladder, oral contrast extravasation from the duodenum into the renal pelvis, or evaluation with retrograde pyelography may offer rare clues.¹³ Infectious symptoms secondary to a pyeloduodenal fistula may be

present, but they are nonspecific and include persistent flank or abdominal pain (60%); general malaise; weight loss (43%); lower urinary tract symptoms (32%); and physical findings of hyperpyrexia, pyuria, and flank tenderness.¹⁴ Manifestations of repeat infections with enteric bacteria may also offer clues, although most commonly, pyelointestinal (especially pyeloduodenal) fistulas may go unrecognized until surgery.

Successful nonsurgical treatment with high-dose proton pump inhibitors and close follow-up with esophagogastroduodenoscopy has been reported in patients whose pyeloduodenal fistulas are secondary to gastrointestinal causes like chronic nonsteroidal anti-inflammatory drug use.¹⁵ Furthermore, conservative or minimally invasive treatment of obstructing nephrolithiasis and chronic infections should be conducted in an attempt to preserve a partially functioning kidney.⁶

XGP, however, leads to a complete destruction of the renal parenchyma. As such, preservation of the kidney would be futile, especially when the inflammatory process, and its neoplasmlike properties, may invade the surrounding tissues, causing pyelointestinal fistulas, and also may invade the spleen and pancreas.⁷ Only surgical treatment with complete removal of the kidney and closure of the duodenal fistula will result in a permanent cure. This operation is extremely difficult because of the amount of densely adherent, chronically inflamed tissue, and in most cases an open surgical approach is probably necessary. Furthermore, locating pyeloduodenal or pyelointestinal fistulas requires a high level of clinical suspicion because they may not be immediately obvious. ❖

Disclosure Statement

The author(s) have no conflicts of interest to disclose.

How to Cite this Article

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