



Case Reports

A Path Less Traveled: Xanthogranulomatous Pyelonephritis Presenting with Thoracic Empyema and Complicated By Rare Pyeloduodenal Fistula

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Abstract

Xanthogranulomatous pyelonephritis (XGP) is a rare and diffusely destructive variant of pyelonephritis. Diagnosis is made using the characteristic radiological findings of renal enlargement with pelvicalyceal dilatation (the ‘bear paw’ sign) with histological analysis demonstrating replacement of renal parenchyma with foamy lipid-laden histiocytes. Treatment of XGP invariably requires nephrectomy. Two particularly rare complications of XGP are thoracic empyema and pyeloduodenal fistula; this case, to our knowledge, is the first such instance of XGP complicated by both. The patient underwent chest drain placement for treatment of empyema, and, following the identification of the pyeloduodenal fistula on antegrade pyelography, underwent surgical treatment with partial right-sided colectomy, radical nephrectomy, and primary duodenal fistula closure. XGP was subsequently identified on pathological analysis of the removed kidney.

BACKGROUND

Xanthogranulomatous pyelonephritis (XGP) is an uncommon and diffusely destructive variant of pyelonephritis with an incidence rate of 1.4 per 100,000 per annum,¹ which occurs almost exclusively in the setting of obstructive uropathy.^{1,2} A rare complication of this rare entity is pyeloduodenal fistula, with approximately 100 case reports (from any cause) extant in the literature in one 2013 analysis.³ In the largest systematic review of XGP to date, just two cases of pyeloduodenal fistula complicating XGP were identified from 1139 aggregated cases (representing 0.18% of cases).¹ Thoracic empyema is an uncommon complication of subdiaphragmatic infections in general – one analysis of 84 cases of thoracic empyema found subdiaphragmatic infections responsible in just 5 cases.⁴ XGP specifically is a vanishingly rare cause: to our knowledge, just five such cases of XGP causing thoracic empyema have ever been described in the literature.⁵⁻⁹ The case described here represents the first documented instance of XGP remarkably complicated by both spontaneous pyeloduodenal fistula and thoracic empyema.

CASE PRESENTATION

A female in her 60s with a past medical history of asthma and chronic right-sided hydronephrosis secondary to a benign ureteral stricture presented to the emergency department with a 2-day history of dyspnea and fever. Elective nephrectomy for her hydronephrotic and nonfunc-

tional kidney had previously been planned by urology as an outpatient approximately ten months prior to her presentation but had been postponed by the patient. She denied any flank or abdominal pain and had no gastrointestinal symptoms. Admission vitals showed a blood pressure of 154/99, heart rate of 139, respiratory rate of 36, oxygen saturation of 90% on ambient room air, and a temperature of 38.4C (101.1F). High-flow nasal cannula oxygen therapy was started with a flow rate of 40L/min and FiO₂ of 50% to achieve a targeted oxygen saturation of ≥94%. Initial laboratory studies revealed a significant leukocytosis of 43.8K/uL (reference range, 4.5-11.0 K/uL) and a new acute kidney injury with a creatinine of 2.36mg/dL (reference range, 0.5-1.10 mg/dL) and urea of 41mg/dL (reference range, 6-23 mg/dL). The patient’s baseline creatinine was 0.9 mg/dL. An initial chest radiograph demonstrated a large right-sided pleural effusion, with contrast-enhanced computed tomography (CECT) additionally demonstrating the presence of partial loculations and a near-complete collapse of the right lung. The CT images also identified inflammatory changes of the right kidney that had progressed since last seen ([Figure 1](#)). Initial broad-spectrum antimicrobial coverage was empirically started with intravenous piperacillin-tazobactam, vancomycin, and oral azithromycin. Thoracentesis was performed with the return of pus; subsequent fluid analysis was consistent with empyema (total nucleated cells 38.350/uL, 95% neutrophils, pH 6.68, LDH 6,801U/L). A chest drain was placed for drainage, and in-

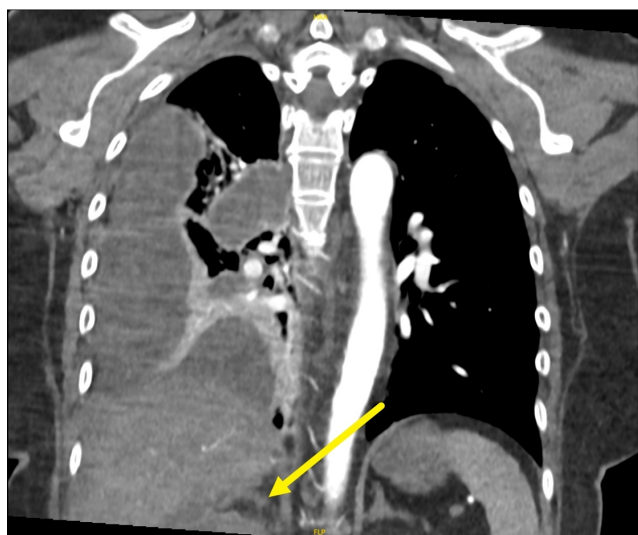


Figure 1. Coronal view of CT scan showing large right-sided empyema with limited visualization of inflammatory renal changes below the right hemidiaphragm (yellow arrow)

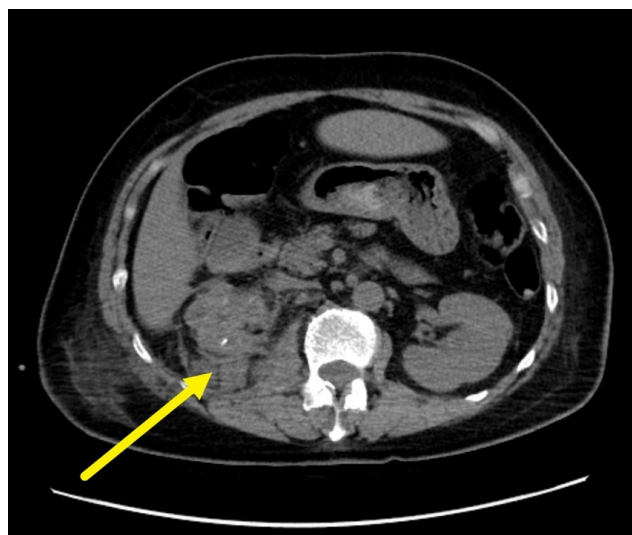


Figure 2. Axial view of CT scan of abdomen and pelvis (without contrast) showing right-sided perinephric fat stranding and a posterior pararenal complex fluid collection (yellow arrow)

tripleural fibrinolytics (alteplase and dornase alfase) were instilled.

The patient responded favorably to initial management. Within hours of stabilization with supplemental oxygen via a flow nasal cannula, the presenting tachypnea resolved, and the tachycardia improved to 100 (resolving completely by day 2). Three days after chest drain placement, the patient was able to be weaned to a low-flow nasal cannula at a flow rate of 6L/min; at ten days, she was able to maintain her oxygen saturation on room air. The chest drain was removed after 14 days in total. Renal function improved in response to the initial treatment with intravenous fluids, and normalized by day 5. Azithromycin was discontinued after three days and vancomycin after two days, following negative microbiology results for atypical organisms and methicillin-resistant staphylococcus aureus (MRSA). Intravenous piperacillin-tazobactam was continued for eight days, during which time the leukocytosis more than halved from 43.8K/uL to 16.2K/uL. On day 9, antibiotics were switched to intravenous ampicillin-sulbactam monotherapy following the identification of susceptible *Proteus mirabilis* and *Streptococcus anginosus* on empyema culture.

A CT of the abdomen and pelvis of the chest without contrast was obtained on day 4 of the admission to further characterize the inflammatory changes seen below the right hemidiaphragm on the CECT of the chest. The study demonstrated right perinephric stranding and the presence of a posterior pararenal fluid collection extending to the right psoas muscle concerning for abscess ([Figure 2](#)). The patient was subsequently scheduled to undergo an abscessogram with fluoroscopically-guided drainage of the infrarenal abscess concomitantly with antegrade pyelography. During the injection of the dye for the antegrade pyelography, the small bowel was seen un-

expectedly to opacify, revealing the existence of a fistulous connection between the kidney and duodenum ([Figure 3](#)). CT abdomen with IV and oral contrast was subsequently obtained, which confirmed the presence of a pyeloduodenal fistula extending from below the renal pelvis to the second portion of the duodenum ([Figure 4](#)). Surgery and urology were consulted and following nutritional optimization with total parenteral nutrition (TPN), the patient was taken to the operating room for exploratory laparotomy, primary fistula takedown, partial right-sided colectomy, and radical right-sided nephrectomy. Intraoperatively, it was appreciated that the infectious process almost completely obliterated the retroperitoneal planes. The patient made a good recovery from surgery and remained in the hospital to complete a further two weeks of intravenous ampicillin-sulbactam (for a total of 21 days of intravenous ampicillin-sulbactam). She was switched to a week-long course of oral amoxicillin-clavulanic acid shortly before discharge, which was completed at home. In total, she was hospitalized for 31 days. Analysis of the removed kidney demonstrated capsular fibrosis and a dilated pelvicalyceal system with necrotic debris, diffuse interstitial areas of hemorrhage and necrosis, and granulation tissue surrounded by foamy lipid-laden macrophages (xanthoma cells) – all findings consistent with a diagnosis of XGP. The long-term outcome in our case was favorable. At outpatient follow-up five weeks after discharge, the patient described being able to tolerate solid food without difficulty, had been able to resume normal activities, including light exercise, and complained only of mild pain associated with the incisional site which was responsive to acetaminophen.

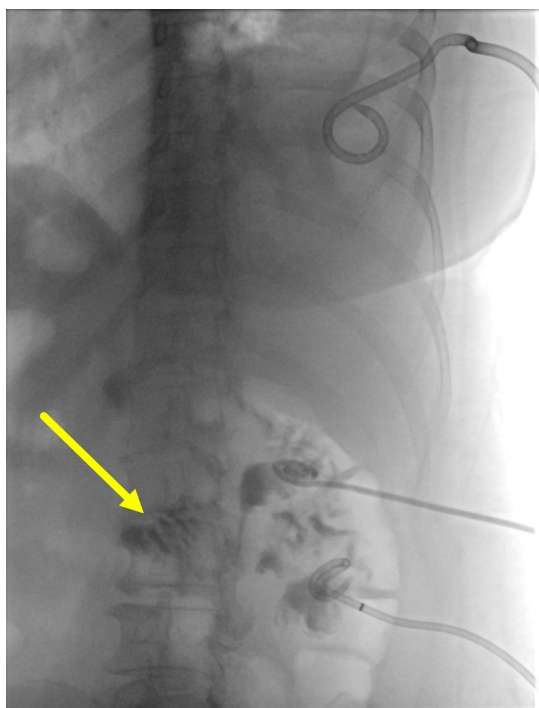


Figure 3. Anterograde pyelography with abscessogram, demonstrating opacification of a segment of small bowel with contrast (yellow arrow) during the procedure



Figure 4. Axial view of contrast-enhanced CT of the abdomen and pelvis, demonstrating presence of the pyeloduodenal fistula (yellow arrow)

DISCUSSION

XGP is a rare variant of pyelonephritis which typically arises in association with chronic obstructive uropathy.¹⁰ It has been described as having a female preponderance with a peak age of incidence in the sixth decade; a history of nephrolithiasis is also often present.¹ Our patient fits broadly within this demographic context, with a benign ureteral stricture being the source of her chronic obstructive uropathy. Risk factors that have been identified for XGP include anatomical variants that predispose toward

obstruction (such as ureteropelvic junction syndrome), renal diseases such as chronic interstitial nephritis, renal malignancies, and certain systemic diseases associated with chronic inflammatory states, including diabetes mellitus, rheumatoid arthritis, and hepatitis C.² Typical symptoms of XGP are flank or abdominal pain, lower urinary tract symptoms, and fever.¹⁰ *E. coli* and *Proteus* are frequently implicated pathogens.¹ Diagnosis of XGP is made using characteristic radiological findings on CT of renal enlargement with pelvicalyceal dilatation (the ‘bear paw’ sign), with histological analysis demonstrating replacement of renal parenchyma with foamy lipid-laden histiocytes.¹⁰ XGP has been described as a notable diagnostic challenge for its tendency to simulate other conditions radiographically, particularly renal cell carcinoma.^{9, 11} Treatment of XGP almost invariably requires radical nephrectomy, with the use of antibiotics in the preoperative and postoperative periods an important factor in achieving successful outcomes.¹⁰ A definitive surgical approach is usually essential because of the disease’s propensity to obliterate tissue planes.¹ Fistulation, as seen in our case, may portend poorer outcomes. In a large systemic analysis by Harley et al., 27% of patients who developed fistulas (from a small subgroup of 15 for whom mortality data was available) died; contrasting with a mortality rate in XGP estimated by the authors to be 1436 per 100,000 patients.¹

Pyeloduodenal fistulas are categorized as either spontaneous or traumatic types. Spontaneous pyeloduodenal fistulas are more common, comprising 82% of cases in one review,¹² and characteristically arising in the presence of chronic renal inflammation. Traumatic pyeloduodenal fistulas are caused by the direct perforation of a foreign object, including iatrogenically, via instrumentation.³ Common symptoms of pyeloduodenal fistula include persistent flank or abdominal pain, malaise, and weight loss, upper gastrointestinal symptoms, and lower urinary tract symptoms.³ Pyeloduodenal fistulas are rare entities; approximately 100 cases are described in the medical literature as of 2013.³ Inflammation secondary to obstructive calculi has been described as the single most common cause of spontaneous pyeloduodenal formation; tuberculosis had historically been responsible for most cases prior to the advent of effective antimycobacterial therapies.¹³ XGP itself is an infrequent cause of pyeloduodenal fistulation, with the most extensive systemic review of XGP to date (comprising 1139 cases and excluding studies of fewer than 10 patients) finding that any kind of fistula development occurred in only 8% of cases of XGP, and that of these just two were pyeloduodenal.¹ Notably, case reports of XGP causing pyeloduodenal fistula appear to be relatively recent: of the ten such cases we identified in the literature, eight had been described since 2016.¹⁴⁻²² Diagnosis of pyeloduodenal fistula is typically made with either CT or antegrade pyelography, both of which are

effective techniques.³ In our case, antegrade pyelography raised the initial suspicion of a fistulous connection, with CECT subsequently confirming presence of pyeloduodenal fistula. Treatment is typically with radical nephrectomy and duodenal fistula closure.^{12,13} In a case review of spontaneous pyeloduodenal fistulas by Rodney et al., all patients treated with nephrectomy and fistula closure had good outcomes; by contrast, all five cases where alternative approaches were documented (conservative management, nephrectomy alone, fistula closure alone, or pyelolithotomy) had poor outcomes: one patient died of sepsis and the remaining four suffered complications including persistently draining cutaneous sinuses, recurrent infections, and pain.¹³ Recent advances in percutaneous interventions have, however, led to reports of patients being treated to good effect with more conservative approaches: these have included nephrostomy, intraureteral stent placement, and endoscopic ligation.^{13, 23-25}

While characteristically a complication of pneumonia, Thoracic empyema can also be caused by non-pneumonic processes such as hematogenous spread of systemic infection, abdominal infection, or as a complication from thoracic surgery.²⁶ One analysis of 82 episodes of empyema identified bronchopulmonary infection as the cause in 73% of cases; subdiaphragmatic sepsis was identified in 6% of cases.⁴ XGP specifically is an infrequent cause of thoracic empyema, with only five cases to our knowledge being previously reported in the literature.⁵⁻⁹ Chest radiograph is the preferred initial imaging modality, which should be followed by visualization on ultrasound to better characterize any identified effusions and plan for diagnostic thoracentesis.²⁷ Chest CT is particularly useful in delineating pathology where empyema (or a complex parapneumonic effusion) is suspected.²⁷ Although aspirated pleural fluid should be sent for analysis, empyema may be diagnosed upon the visualization of frank pus on drainage.²⁷ First-line treatment is with systemic antibiotics and prompt drainage of the pleural space – use of fibrinolytic therapy has additionally been associated with a decreased rate of surgical interventions and reduced length of hospital stay.^{26,28}

The complications of XGP that our patient developed, while rare, highlight the markedly destructive character of this clinical entity. That our patient developed this rare condition after having deferred an outpatient procedure for treatment of a benign ureteral stricture

stresses the importance of timely urological management of chronic obstructive uropathy. Our patient's presentation is particularly instructive with regard to the importance of considering subdiaphragmatic causes of unilateral thoracic empyema, especially where there is a history of ipsilateral chronic renal inflammation. We also believe this case demonstrates that the potential for fistulation must be considered where there is evidence of complicated abscess formation. Prompt referral to surgery when this is found is essential.

Disclosures/Conflicts of Interest

The authors declare they have no conflicts of interest.

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- Substantial contributions to the conception or design of the work; or the acquisition, analysis, or interpretation of data for the work; AND
- Drafting the work or revising it critically for important intellectual content; AND
- Final approval of the version to be published; AND
- Agreement to be accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved.

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