International Journal of Surgery Case Reports 85 (2021) 106287

Contents lists available at ScienceDirect

International Journal of Surgery Case Reports

journal homepage: www.elsevier.com/locate/ijscr

Case report

Xanthogranulomatous pyelonephritis: A rare case report of a 54 year old female (a potentially fatal infection)

Jacqueline Gri [a,*], Mohamad Ammar Hatahet [b,c,d], Siddharth Chopra [d]

[a] Ross University School of Medicine, Miramar, FL, USA
[b] Michigan State University, East Lansing, MI, USA
[c] McLaren Oakland, Pontiac, MI, USA
[d] Department of Internal Medicine, St. Joseph Mercy Oakland, Pontiac, MI, USA

ARTICLE INFO

Keywords:
Xanthogranulomatous
Pyelonephritis
Proteus
Calculus
Nephrectomy

ABSTRACT

Introduction and importance: Xanthogranulomatous pyelonephritis (XPGN) is a rare pathology of the kidneys occurring in 0.6 to 1% of all cases of renal infections, in both men and women. It is characterized by severe inflammation of the renal parenchyma leading to formation of granulomatous tissue containing lipid-laden macrophages. This condition may mimic less aggressive or benign conditions but may worsen or be fatal if not treated aggressively.

Case presentation: Our patient is a 54 year old Caucasian female who presented with five days of left flank pain, hematuria, chills, nausea and vomiting. Imaging and biopsy results showed that the patient had XPGN.

Clinical discussion: XPGN is a difficult condition to diagnose as the symptoms are non-specific relative to renal cell carcinoma or other common renal infections. Definitive diagnosis is made with a biopsy; however, clues in various imaging modalities are used to make a tentative diagnosis. It is unclear whether earlier surgical intervention would have improved overall patient outcomes. Currently, a partial or complete nephrectomy is the only effective treatment.

Conclusion: Aggressive management including early diagnosis, antibiotics and nephrectomy appears to be critical in preventing progression and complications of XPNG.

1. Introduction

XPGN is a pathology of the kidneys characterized by severe inflammation of the renal parenchyma leading to formation of granulomatous tissue containing lipid-laden macrophages [1]. It most commonly occurs in middle-age diabetic women with a history of recurrent urinary tract infections [2]. Causes include urolithiases and ascending infection of the urinary tract from organisms like *Proteus mirabilis, Escherichia coli, Pseudomonas, Klebsiella* and *Enterococcus faecalis* [3] which secrete urease that makes the urine alkaline. Urease further precipitates phosphate, calcium, and magnesium leading to the formation of a large irregularly shaped staghorn caliculi [4]. XPGN occurs in roughly 1% of all cases of pyelonephritis with a mortality rate of 10% [5]. Complications include renal perforation and sepsis, which are fatal if not treated rapidly and aggressively with surgery.

2. Case presentation

2.1. Admission and history

We present a case of a 54 year old Caucasian female with no significant past medical history who presented with left lower back pain and dysuria for the past five days. She rated her pain as 8/10 in intensity, non-radiating and had five days of hematuria, chills, nausea, vomiting and syncopal episodes. Review of systems was insignificant. Fifteen days prior, our patient presented to another hospital ED with severe left CVA tenderness but was afebrile, non-tachycardic/tachypneic, normotensive and was saturating 91% on room air.

She had leukocytosis with a left shift on the CBC. Urinalysis showed evidence of a urinary tract infection and later cultures were positive for E.coli. CT scan with contrast (Fig. 2) confirmed a staghorn calculus with dilatation of the calyces, cortical thinning, perinephric abscess, perinephric stranding, and collection along the anterior lower pole of the left

Abbreviations: XGPN, Xanthogranulomatous pyelonephritis.

* Corresponding author at: Ross University School of Medicine, 2300 SW 145th Ave #200, Miramar, FL 33027, USA.

https://doi.org/10.1016/j.ijscr.2021.106287
Received 29 June 2021; Received in revised form 3 August 2021; Accepted 3 August 2021
Available online 5 August 2021

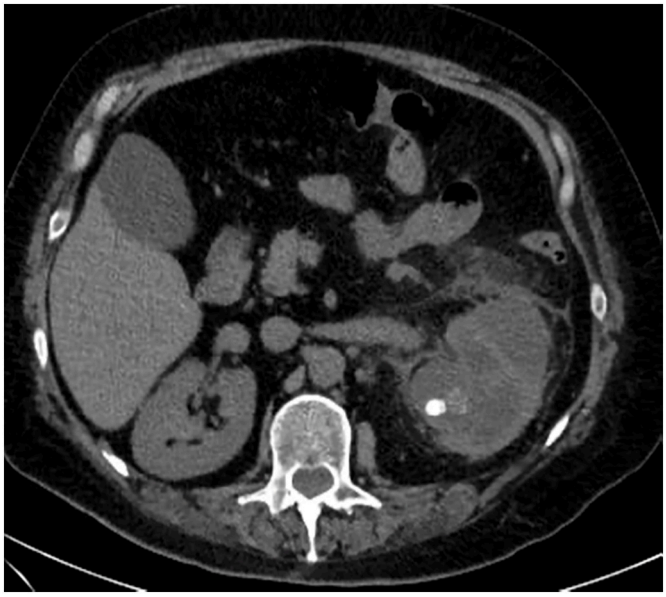

Fig. 2. CT scan of abdomen showing XPGN: used to make a presumptive diagnosis of XGPN.

kidney consistent with XGPN. Another calculus at the proximal left ureter resulted in severe left hydronephrosis (Fig. 1). Reactive lymphadenopathy was also seen adjacent to left renal vein.

The patient became septic on admission and was treated with fluids and antibiotics. She was immediately taken to surgery by urology for cystoscopy with retrograde pyelogram and left-sided ureteral stent placement. She was then transferred to another hospital where Urology placed a left-sided pigtail catheter in the perinephric abscess. Culture of the drainage grew proteus mirabilis with no anaerobes. Despite several days of antibiotic therapy, purulent draining from the pigtail catheter did not subside; therefore, Urology proceeded with left radical nephrectomy. During surgery, the patient was found to have a severely hydronephrotic left kidney with multiple spilling abscesses but no renal perforation. There were dense hypervascular perirenal inflammatory rings and adhesions. There was also a splenic capsule laceration which

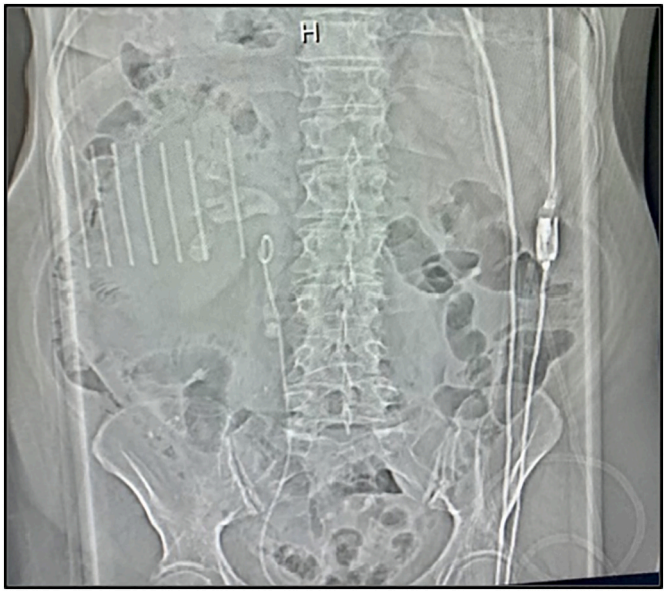

Fig. 1. Plain radiograph of left-sided struvite stone: obtained upon arrival to the ED to identify the struvite stone.

was repaired with surgical bolsters. After the procedure, the patient's hemoglobin dropped from 11.5 to 6.5 g/dl which required blood transfusions and IV iron. Infectious disease initially started the patient on ceftriaxone and metronidazole which were eventually changed to 14 days of piperacillin-tazobactam for coverage of Pseudomonas. Eight days following the nephrectomy, there was stool in the patient's JP drain. Exploratory laparotomy confirmed necrosis and perforation of the bowel at the splenic flexure, a late complication of surgery that required left colectomy with mobilization of the splenic flexure. Linezolid was added to the patient's 14-day antibiotic regimen due to isolation of Enterococcus from the JP drain (Table 1). She had no further complications [6].

2.2. Follow up

Our patient's follow-up was roughly two weeks post-discharge. She reported eating, drinking, urinating, and stooling without difficulty and denied any new symptoms or complications.

3. Discussion

XGPN is a rare condition with an incidence of 0.6 to 1% of all cases of renal infections [7]. Our patient's presentation, diagnosis and management plan will be discussed to provide teaching points for improvement of future cases and prevention of similar complications.

Risk factors of XGPN include female gender, middle-age (40-50th decade) [2], diabetics and those with a history of urinary tract infections or renal stones [3]. Chronic inflammatory conditions such as systemic amyloidosis were reported in two cases of XGPN [8,11]; other risk factors include renal allografts [9], immunocompromised patients, abnormal lipid metabolism, and hypertension [7]. Our patient's only risk factors were gender and age.

In an analysis of a case series of 40 patients with XGPN, close to all patients reported flank pain, 48% of patients reported classic UTI symptoms and 58% of patients had a palpable renal mass. Other findings included anemia, pyuria and elevated blood sedimentation rate [8]. In a

Table 1
Timeline of events.

Day 0	• Patient presented with left flank pain and sepsis • CT scan showed evidence of a staghorn calculi, perirenal abscess and XGPN. • Ceftriaxone and gentamicin started. • Cystoscopy and left stent placement.
Day 3	• Pigtail catheter placed • Kept on ceftriaxone. • Infectious disease consulted.
Day 9	• Open left nephrectomy with complicated spillage of pus into the abdomen and splenic capsule laceration. • Patient became hypotensive and required two blood transfusions with temporary use of vasopressors. • JP drain and foley catheter placed. • Kept on ceftriaxone.
Day 12	• Patient developed abdominal distension, nausea and vomiting; no bowel movements since before the nephrectomy. • CT scan showed evidence of a small bowel obstruction.
Day 13	• NG tube yielded 5 L of bilious material. Patient felt better with some moderate distension.
Day 16	• Patient still had NG tube and started PT/OT due to inability to ambulate and severe weakness.
Day 17	• Stool noted in her JP drain • Exploratory laparotomy showed necrosis of the bowel at the splenic flexure with perforation • Left colectomy performed with transverse sigmoid anastomosis. • Patient antibiotic regimen changed to piperacillin-tazobactam for pseudomonal coverage
Day 22	• Patient transferred out of ICU; no new complaint and felt better overall. • Linezolid added for isolation of Enterococcus from JP drain.
Day 29	• Patient went home with a picc line for completion of 14 days of antibiotics

case series of 90 patients, 77.5% were preceded by a calculus and 72% of those cases had urine *E. coli* or Proteus cultures; Bowel fistula was a rare complication [8]. Our patient's case of XGPN seems to have been caused by an obstructive struvite stone. She reported hematuria, flank pain and UTI symptoms consistent with common literature reports. There does not seem to be a key element in the patient's history, or in studied cases, that would differentiate this condition from uncomplicated pyelonephritis secondary to a ureteral obstruction; all the listed symptoms appear non-specific. In addition, our patient's duration of symptom onset was relatively short at five days.

The diagnosis of XGPN is ultimately confirmed through a pathological sample of lipid laden macrophages. Prior to surgery, a combination of historical finding and imaging are used to make a presumptive diagnosis. XGPN can sometimes be confused with renal cell carcinoma but biopsy with a PAS stain helps to differentiate them. Alternatively, testing for urine foam cells as part of pyelonephritis may be helpful to make an early diagnosis. In a case series of 40 patients, plain radiograph findings consistent with XGPN include outline enlargement of the involved kidney (75%), ipsilateral absence of the psoas shadow (63%) and urinary tract calculi (60%) [8]. CT scan with contrast is the most used diagnostic tool and the findings show hypoechoic areas of dilated collecting system with a multiloculated enhanced rim that resembles a bear paw; the contrast lining is indicative of the necrotic tissue [7]. Our patient's CT scan findings were consistent with typical imaging findings in the literature and allowed a presumptive diagnosis to be made. Contrast must be used to visualize the rim of necrotic tissue and if a patient presents with an acute kidney injury or a contrast allergy, an MRI instead of a CT scan is superior. The use of MRI may pose unnecessary financial burden on the healthcare system if it does not add more diagnostic value; when a CT with contrast is insufficient to make the correct diagnosis, an MRI may be useful as a second line imaging choice.

This condition is unilateral and a nephrectomy is ultimately the only treatment. It remains uncertain whether delaying nephrectomy ultimately leads to fewer complications and better outcomes in patients with XGPN. In our patient, symptoms of colonic injury presented late (eight days later) likely due to septic sequalae of fecal spillage. The injury may have occurred during management of the splenic laceration. Our patient's splenic and colonic injury may have been prevented if the nephrectomy had been performed sooner. Delayed nephrectomy by six months has been studied in a population of 14,677 patients with cT1a renal cell carcinoma and there was no effect on overall survival [13]. The aggressive nature and rapid expansion of XGPN compared to RCC may indicate a need for a more urgent nephrectomy. Some patients can have a partial nephrectomy depending on the risks and benefits [12]. Preoperative presumptive diagnosis allows for early management with antibiotic therapy and nephrostomy to reduce complications like bleeding, fistulas, and infections/sepsis. In our case, presumptive diagnosis was made, and culture of the drainage allowed for targeted antibiotic therapy and a partial nephrectomy was performed. This condition can also be treated with a laparoscopic nephrectomy; however, this may be difficult as not every hospital has the surgical expertise to perform this procedure. It can be performed in highly selected cases and has the advantage of less blood loss, shorter hospital stays, quicker resumption to oral intake and lower transfusion rates.

4. Conclusion

Xanthogranulomatous pyelonephritis (XPGN) is a rare condition that may rapidly worsen or be fatal if not treated aggressively with surgery. CT scan with contrast has been the most used diagnostic tool but ultimate diagnosis is made via pathologic sample; a presumptive diagnosis is made until a pathologic sample of granulomatous tissue containing lipid-laden macrophages is obtained. It remains uncertain whether delaying nephrectomy ultimately leads to fewer complications. Targeted antibiotic therapy and surgical nephrectomy are currently the optimal treatment. The type of nephrectomy (laparoscopic vs open) is dependent on the expertise and resources of the hospital site.

Provenance and peer review

Not commissioned, externally peer-reviewed.

Sources of funding

N/a

Ethical approval

N/a

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Research registration

N/a

Guarantor

Jacqueline Gri, BSc.

CRediT authorship contribution statement

Jacqueline Gri, BSc: writing the paper
Dr. Mohamad Ammar Hatahet, M.D., MPH, FACP: review & editing
Dr. Siddharth Chopra, MD: review & editing

Declaration of competing interest

N/a

References

[1] F. Afgan, S. Mumtaz, M.H. Ather, Preoperative diagnosis of xanthogranulomatous pyelonephritis, Urol. J. 4 (3) (2007) 169–173.
[2] A.U. Chuang CK, M.K. Lai, P.L. Chang, M.H. Huang, S.H. Chu, C.J. Wu, H.R. Wu, Xanthogranulomatous pyelonephritis: experience in 36 cases, SOJ Urol. 147 (2) (1992) 333.
[3] A.U. Goodman, T. Curry, T. Russell, Xanthogranulomatous pyelonephritis (XGP): a local disease with systemic manifestations. Report of 23 patients and review of the literature, Medicine (Baltimore) 58 (2) (1979) 171.
[4] H.L. Mobley, M.D. Island, G.S.O. Massad, Virulence determinants of uropathogenic Escherichia coli and Proteus mirabilis, Kidney Int. Suppl. 47 (S129) (1994).
[5] S. Siddappa, K. Ramprasad, M.K. Muddegowda, Xanthogranulomatous pyelonephritis: a retrospective review of 16 cases, Korean J Urol. 52 (6) (2011) 421–424, https://doi.org/10.4111/kju.2011.52.6.421.
[6] R.A. Agha, T. Franchi, C. Sohrabi, G. Mathew, for the SCARE Group, The SCARE 2020 guideline: updating consensus Surgical CAse REport (SCARE) guidelines, Int. J. Surg. 84 (2020) 226–230.
[7] S.K. Jha, N.R. Aeddula, Pyelonephritis Xanthogranulomatous. [Updated 2020 Dec 1], in: StatPearls [Internet], StatPearls Publishing, Treasure Island (FL), Jan 2021.
[8] A.U. Parsons, M.A. Harris, S.C. Longstaff, A.J. Grainger, Xanthogranulomatous pyelonephritis: a pathological, clinical and aetiological analysis of 87 cases, Diagn. Histopathol. 6 (3–4) (1983) 203.
[9] W. Hitti, C. Drachenberg, M. Cooper, E. Schweitzer, C. Cangro, D. Klassen, A. Haririan, Xanthogranulomatous pyelonephritis in a renal allograft associated with xanthogranulomatous diverticulitis: report of the first case and review of the literature, Nephrol. Dial. Transplant. 22 (11) (2007) 3344.
[11] A.U. Lauzurica R, A. Felip, A. Serra, J.M. Saladie, E. Montserrat, B. Encabo, A. Caralps, Xanthogranulomatous pyelonephritis and systemic amyloidosis: report

of 2 new cases and the natural history of this association, J. Urol. 146 (6) (1991) 1603.
[12] L.M. Peréz, J.B. Thrasher, E.E. Anderson, Successful management of bilateral xanthogranulomatous pyelonephritis by bilateral partial nephrectomy, J. Urol. 149 (1) (1993) 100.
[13] Delayed nephrectomy has comparable long-term overall survival to immediate nephrectomy for cT1a renal cell carcinoma: a population-based analysis, Urol. Oncol. 38 (3) (2020), https://doi.org/10.1016/j.urolonc.2019.11.008. Pages 74.e13-74.e20, ISSN 1078-1439.