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Inflammation and infection

A rare case of spontaneous kidney rupture from xanthogranulomatous pyelonephritis presenting with neurological symptoms and septic peritonitis

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ABSTRACT

Xanthogranulomatous pyelonephritis (XGP) is a rare, chronic inflammatory kidney disorder. It typically arises in patients with a history of nephrolithiasis, urinary tract obstruction, or recurrent infections. Additional risk factors include diabetes and immunocompromised states. We present an unusual instance of XGP in a patient without known risk factors, who developed spontaneous kidney rupture complicated by life-threatening sepsis. This case underscores the importance of maintaining XGP in the differential diagnosis even for atypical presentations, particularly when sepsis is the dominant clinical feature. Given the risk of serious complications, prompt diagnosis and intervention are crucial. Nephrectomy is the mainstay treatment of XGP.

1. Introduction

Xanthogranulomatous pyelonephritis (XGP) is a rare, chronic inflammatory kidney disorder characterized by the destruction of renal and perirenal tissue, replaced by granulomatous infiltrates containing lipid-laden foamy macrophages. It typically arises in patients with a history of nephrolithiasis, urinary tract obstruction, or recurrent infections. Additional risk factors include diabetes and immunocompromised states.[1,2] In this case report, we present an unusual instance of XGP in a patient with no known risk factors, who developed spontaneous kidney rupture complicated by life-threatening sepsis.

2. Case presentation

A 67-year-old man presented with a one-week history of fever and right lower limb numbness. He denied abdominal pain, hematuria, stone passage, or urinary symptoms. On examination, tenderness was noted over the right loin and right upper quadrant, but neurological assessment of the lower limb was unremarkable. At admission, he was febrile and tachycardic, with leukocytosis (21.3×10^9/L) on complete blood count. Renal function was preserved (eGFR 70 mL/min/1.73 m^2), and urinalysis revealed no red blood cells, leukocyte esterase, or nitrites. Urine culture was negative.

Given his unstable vitals, an urgent contrast-enhanced CT abdomen/pelvis was performed, revealing a large right renal mass ($15.2 \times 13.5 \times 18.5$ cm) with a cystic cavity and internal fat locules (Fig. 1). The lesion extended into the right psoas muscle ($6.4 \times 4.2 \times 7.2$ cm collection) (Fig. 2) and hepatic segment 6 ($2.8 \times 2.3 \times 3.3$ cm collection) (Fig. 3). Broad-spectrum intravenous antibiotics and fluid resuscitation were initiated, but his condition deteriorated rapidly, progressing to septic shock and respiratory distress.

An emergency exploratory laparotomy and radical nephrectomy were performed. Upon entering the peritoneal cavity, approximately 1000 mL of purulent fluid was drained, and a ruptured right renal/perinephric abscess with involvement of hepatic segment 6 was identified (Fig. 4). The abscess was debrided, and the psoas collection was drained. Postoperatively, he was managed in the ICU, with eventual resolution of sepsis and right lower limb numbness. The culture grew Klebsiella pneumoniae. The patient received a two-week course of amoxicillin-clavulanate therapy after right nephrectomy. His renal function remained stable (eGFR 72 mL/min/1.73 m^2 at discharge), and follow-up CT confirmed resolution of the collections.

Histopathological examination of the kidney demonstrated dense infiltrates of foamy histiocytes, lymphocytes, plasma cells, and

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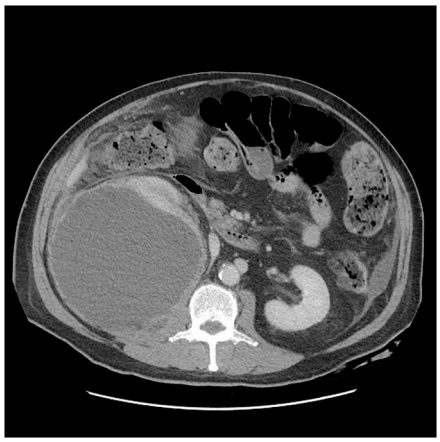

Fig. 1. Contrast-enhanced abdominal CT scan (axial view) showing a large, hypodense collection involving the right renal parenchyma, with surrounding inflammatory stranding.

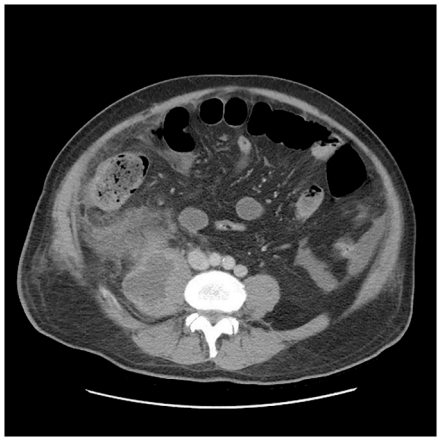

Fig. 2. Contrast-enhanced axial CT image demonstrates right psoas muscle involvement with hypodense fluid collection consistent with abscess formation.

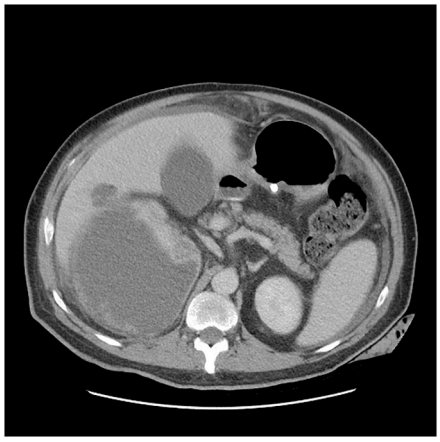

Fig. 3. Contrast-enhanced abdominal CT scan demonstrating a hypodense lesion in hepatic segment VI, consistent with liver abscess.

neutrophils with occasional multinucleated giant cells. Immunohistochemistry revealed CD68-positive but S100-negative histiocytes, confirming the diagnosis of xanthogranulomatous pyelonephritis.

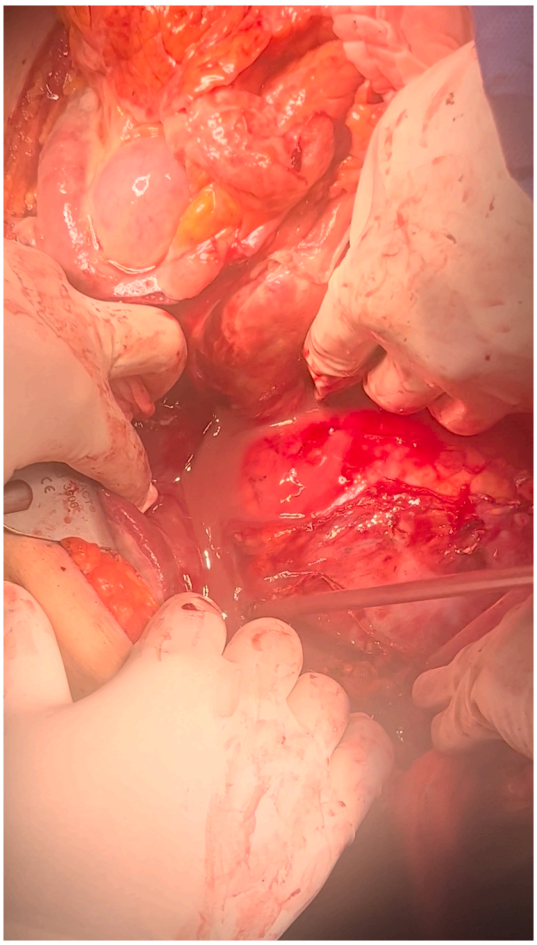

Fig. 4. Surgical exploration revealed frank rupture of the right kidney with active spillage of purulent material into the retroperitoneal space.

3. Discussion

Xanthogranulomatous pyelonephritis (XGP) is a rare inflammatory condition of the kidney. Classical risk factors include female sex, nephrolithiasis, recurrent urinary tract infections, and diabetes mellitus.[3] Typical clinical manifestations encompass flank/abdominal pain, lower urinary tract symptoms, fever, palpable mass, gross hematuria, and weight loss. Notably, our patient presented atypically—lacking diabetes, immunocompromise, or urolithiasis—which initially lowered clinical suspicion for XGP.

Histopathological examination reveals granulomatous inflammation with neutrophils, lymphocytes, plasma cells, lipid-laden xanthomatous histiocytes, and multinucleated giant cells.

Computed tomography (CT) is the diagnostic imaging modality of choice for XGP, which may manifest in diffuse or focal forms. Despite CT's central role, XGP often mimics other renal pathologies, such as complicated renal cysts, renal cell carcinoma, or renal tuberculosis, necessitating histopathological confirmation as shown in (Table 1).

Xanthogranulomatous pyelonephritis (XGP) necessitates antibiotic therapy, often accompanied by percutaneous drainage or surgical intervention. Radical nephrectomy serving as the definitive treatment for most cases. While partial nephrectomy may be considered in select circumstances, it remains uncommon. Although adjunctive antibiotics are essential in all patients, medical therapy alone is rarely curative. In cases of persistent sepsis despite antibiotic treatment or percutaneous drainage, immediate nephrectomy becomes critical.[8]

Table 1
Reported Cases of Xanthogranulomatous Pyelonephritis mimicking Other Renal Pathologies.

No.	Author	Year	Presentation	Initial imaging findings	Initial diagnosis	Final Diagnosis	Definitive treatment
1	Hoi-Lung Wong (current paper)	2025	Fever, lower limb numbness	Large renal collection, psoas abscess	Infected renal tumor, renal abscess	XGP	Radical nephrectomy
2	Shantanu Chandrashekhar[4]	2024	Right loin pain	Heterogenous renal lesion with hyperdense solid components	Complicated renal cyst	XGP	Partial nephrectomy
3	Yoomee Kang[5]	2024	Right upper quadrant pain	well-enhancing large septated cystic mass in the right kidney	Renal cell carcinoma	XGP	Radical nephrectomy
4	Saad Bkiri[6]	2023	Right abdominal pain	Right hydronephrosis, PUJ stone and liver abscess	Right pyonephrosis, liver abscess	XGP	Radical nephrectomy
5	Bülent Altınoluk[7]	2012	Right loin pain with fever	Retroperitoneal bleeding of renal origin	Urinary tract infection	XGP	Radical nephrectomy

4. Conclusion

Xanthogranulomatous pyelonephritis classically presents in patients with nephrolithiasis, urinary tract obstruction, or recurrent infections. However, atypical cases may occur without these risk factors. The diagnostic challenge is compounded by imaging features that often mimic malignancy or other infections, potentially delaying diagnosis. This case underscores the importance of maintaining XGP in the differential diagnosis even for atypical presentations, particularly when sepsis is the dominant clinical feature. Given the risk of serious complications, prompt diagnosis and intervention are crucial. Treatment of XGP includes antibiotics, percutaneous drainage or surgical interventon. For diffuse or advanced-stage disease, nephrectomy remains the treatment of choice.

CRediT authorship contribution statement

Hoi- Lung Wong: Writing – review & editing, Writing – original draft, Software, Resources, Project administration, Methodology, Conceptualization. **Henry Hong-Yin Lie:** Writing – review & editing, Project administration. **Ting-Fung Wong:** Writing – review & editing, Visualization, Project administration. **Wilson Pui-Long Hung:** Writing – review & editing, Software. **Rong Na:** Writing – review & editing. **Chiu-Fung Tsang:** Supervision. **Terence Chun-Ting Lai:** Supervision. **Brian Sze-Ho Ho:** Supervision. **Ada Tsui-Lin Ng:** Supervision.

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