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Case report: Xanthogranulomatous pyelonephritis masquerading as cystic renal cell carcinoma

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ABSTRACT

Background: Xanthogranulomatous pyelonephritis (XGP) is a rare chronic bacterial inflammation of the renal parenchyma and is often a diagnostic dilemma.
Case Presentation: We present a challenging case of a patient with XGP. Initially thought to have had renal cell cancer she was treated accordingly with a partial nephrectomy. However, on the final pathology, she was found to have XGP and required further antibiotic therapy and referral to the infectious disease service.
Discussion: Management of XGP and diagnostic pitfalls are discussed.
Conclusion: XGP is a diagnostic and therapeutic dilemma. Partial Nephrectomy may be appropriate in management of XGP in select cases.

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Xanthogranulomatous; pyelonephritis; partial nephrectomy; urology; radiology; infectious disease, infiltrative renal disorder

Background and introduction

Xanthogranulomatous pyelonephritis (XGP) is a rare chronic bacterial inflammation of the renal parenchyma [1,2]. Urine cultures often show the growth of *Escherichia coli* and *Proteus mirabilis* [3]. This disease has historically been a clinical enigma and is often referred to as the 'great imitator'. XGP is more prevalent in women with the mean occurrence age of 45.2 years [4]. It classically presents with malaise, fever, flank pain and weight loss. Radiographic investigations may reveal a diffusely infiltrative renal lesion with concomitant ureteric calculi [5]. Once diagnosed, this malady is classified into three stages depending on the extent of renal involvement: XGP localized to renal parenchyma, XGP involving the renal parenchyma and Gerota's fat, and XGP involving the parenchyma, perinephric and paranephric tissue [6]. Antibiotic therapy and open or laparoscopic radical nephrectomy with removal of all involved tissues is the current curative treatment for XGP [7]. If left untreated, XGP can progress and lead to infection and inflammation of surrounding organs, sepsis and cause significant morbidity and mortality. We presented a case of XGP initially though to have been renal cell carcinoma.

Case presentation

A 55-year-old woman presented to her urologist for overactive bladder symptoms, right upper quadrant pain and management of kidney stones in June of 2017. She was found to have a non-obstructing calculi in her right kidney and subsequently underwent extracorporeal shock wave lithotripsy in September of 2017. Following the procedure, the patient continued to complain about pain. She denied gross haematuria or constitutional symptoms. The physical exam was benign. The patient had a history of several parapelvic cysts in the left kidney found on abdominal ultrasound in March of 2017, non-obstructive renal stones, chronic urinary tract infections (UTI), urgency and stress incontinence. An uninfused computed tomography (CT) in November of 2017 for further investigation of her symptoms showed a 3.9 cm right renal mass not previously reported.

To further characterize the lesion, the patient underwent a contrast infused CT in December of 2017 (Figure 1) which favoured the diagnosis of renal cell carcinoma (RCC). A subsequent MRI was reported to be concerning for cystic RCC (Figure 2). There were no metastatic lesions seen in the chest, abdomen or pelvis at the time. Urinalysis showed growth of pain-sensitive *E. coli* in December 2017, which was subsequently managed with sulfamethoxazole and trimethoprim (TMP/SMX) for 2 weeks. The remainder of her preoperative work up were all unremarkable.

Given the high suspicion of malignancy based on imaging, the patient was prepared for surgery. A right-sided open partial nephrectomy was performed in March of 2018 with complete excision of 4 cm mass. The patient had an unremarkable post-operative course. She was discharged home 4 days after her surgery. Surgical pathology revealed a 3.8 × 3.5 × 3.5 cm partly solid and partly cystic mass. The solid component was yellow-tan and the cystic centre contained green purulent material and red brown solid material. Negative

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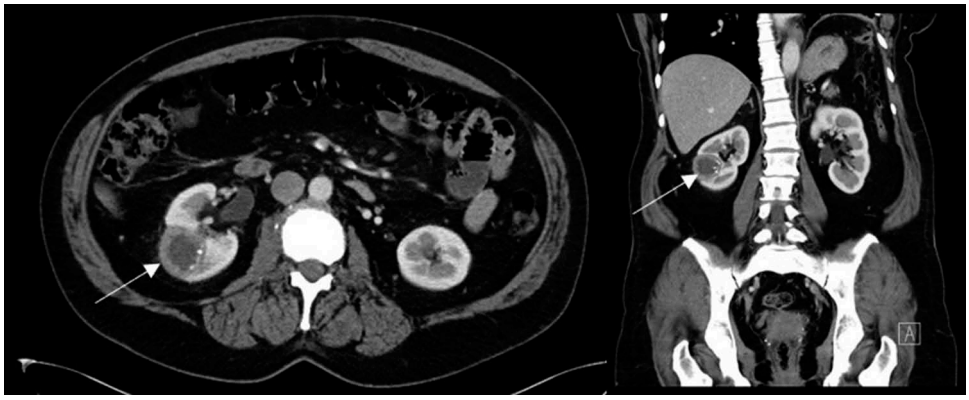

Figure 1. The right renal lesion is indicated by the white arrows (contrast infused CT).

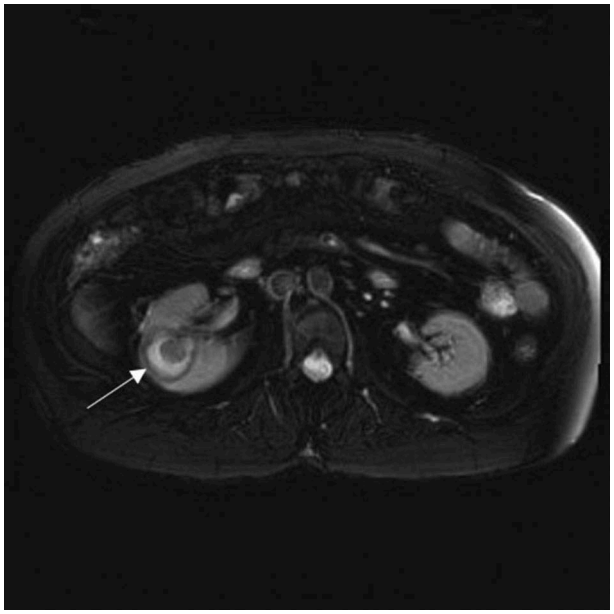

Figure 2. The right renal lesion is indicated by the white arrow.

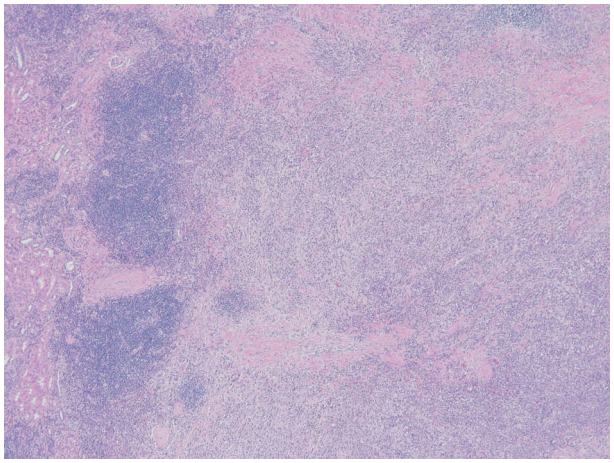

Figure 3. Replacement of renal parenchyma with sheets of foamy lipid-laden macrophages and chronic inflammation with fibrosis at periphery, right side of the picture. Remnants of renal tissue is shown in the left. (H&E; 4X).

margins were achieved. The histopathology report indicated there were changes in the kidney consistent with Xanthogranulomatous Pyelonephritis (Figures 3–6).

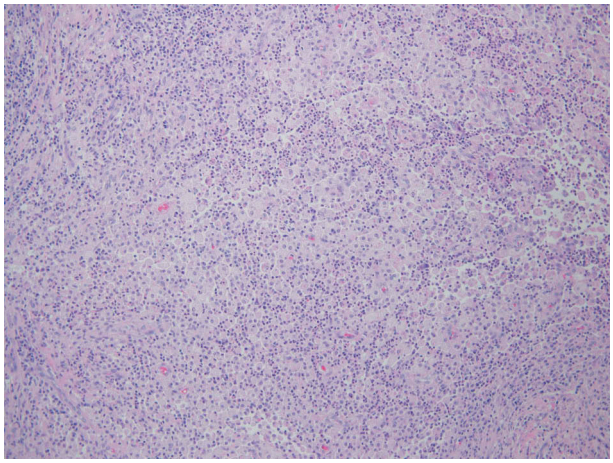

Figure 4. Sheets of lipid-laden foamy histiocytes admixed with acute and chronic inflammatory cells including lymphocytes, plasma cells and neutrophils in necrotic areas shown in right upper corner of the picture. (H&E; 10X).

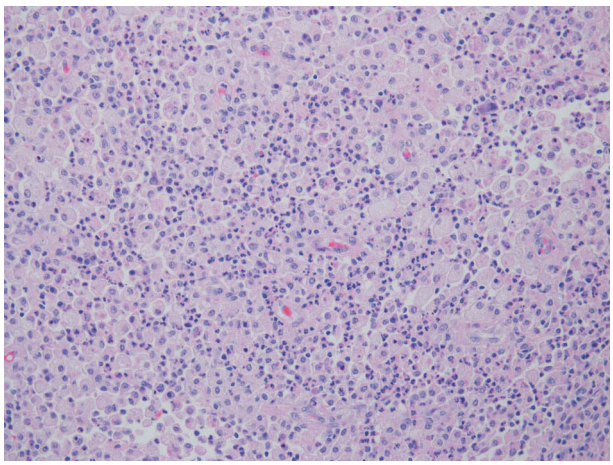

Figure 5. Sheets of lipid-laden foamy histiocytes admixed with acute and chronic inflammatory cells. (H&E; 20X).

A summary of patient's perioperative blood work is shown in Tables 1 and 2. Full blood counts are included from pre-operative (December 2017), early post-operative (April 2018) and late post-operative (November 2018) periods. Electrolytes and liver function tests were only

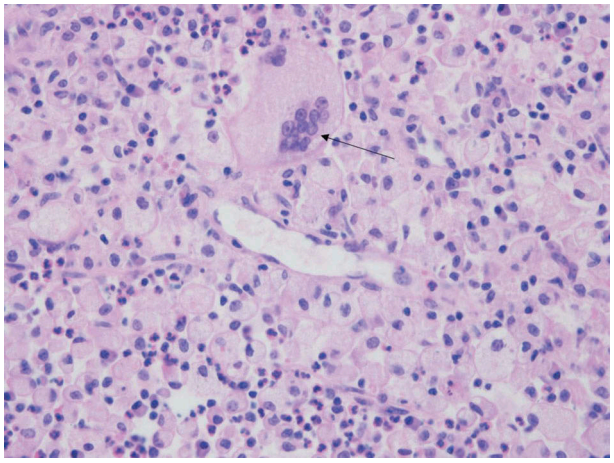

Figure 6. A view of sheets of lipid-laden foamy histiocytes including a multinucleated giant cell shown by the black arrow. (Haematoxylin and eosin staining; 40X).

Table 1. Full blood counts.

Test	December 2017	April 2018	November 2018
Leukocytes	$8.8x10^9$/L	*$14.8x10^9$/L	$7.9x\ 10^9$/L
Erythrocytes	$4.65x10^{12}$/L	$4.28x10^{12}$/L	$4.85x10^{12}$/L
Haemoglobin	136 g/L	126 g/L	145 g/L
Haematocrit	0.417	0.387	0.443
MCV	89.7fL	90.4fL	91.3fL
MCH	29.2pg	29.4pg	29.9pg
MCHC	326 g/L	326 g/L	327 g/L
RDW	13.5%	*14.7%	14%
Platelets	$270x10^9$/L	$246x10^9$/L	$259x10^9$/L
MPV	10.1fL	10.2fL	11fL
Neutrophils	56.3%	*71.4%	58.9%
Lymphocytes	30.2%	*14.6%	30.7%
Monocytes	11.3%	*12.6%	8.9%
Eosinophils	1.7%	0.7%	0.9%
Basophils	0.3%	0.4%	0.3%
Granulocytes immature	0.2%	0.3%	0.3%

Abnormal values are marked*. MCV: mean corpuscular volume, MCH: mean cell haemoglobin, MCHC: mean corpuscular haemoglobin concentration, RDW: Red blood cell distribution width, MPV: mean platelet volume.

Table 2. Pre-operative electrolytes and LFTs.

Test	Result
Sodium	143 mmol/L
Potassium	4.9 mmol/L
Chloride	101 mmol/L
Urea	6.2 mmol/L
Creatinine	81 umol/L
Albumin	*46 g/L
Total Bilirubin	10 umol
Direct Bilirubin	4 umol/L
Aspartate Aminotransferase (AST)	34 U/L
Alanine Aminotransferase (ALT)	*50 U/L
Lactate Dehydrogenase (LDH)	448 U/L
Gamma Glutamyl Transferase (GGT)	26 U/L
Alkaline Phosphatase (ALP)	83 U/L

Abnormal values are marked*. LFTs: Liver function tests.

available from a pre-operative visit. Patient's creatinine remained stable during the post-operative course and was 61 µmol/L and 68 µmol/L in April and November 2018. She was found to have a UTI during the post-operative course in April of 2018. The urine culture at this time showed pan-sensitive E. Coli. This was treated with a two-weeks course of TMP/SMX. She subsequently was sent to the infectious disease service, and was given the diagnosis of XGP, with further assessment and follow up. In November of 2018, she had urine cultures that were negative for bacterial growth. A CT scan in November 2018 showed significant improvement in comparison to her pre-operative imaging (Figure 7). Should she have a recurrence of her XGP she will likely require removal of the remainder of her right kidney.

Discussion and review of the literature

The pathophysiology of XGP is thought to be due to chronic inflammation secondary to destructive process of infection in association with obstructive features [2]. Our patient presented with a history of flank pain, chronic UTI's as well as a recent kidney stone treated with lithotripsy. She did not present with the other symptoms of XGP which can include generalized malaise, flank pain, fever, chills, dysuria and weight loss [2] Her laboratory investigations were unremarkable unlike some other cases where anaemia, leukocytosis and pyuria, can be found[8]. This is in keeping with the variable presentation of XGP. Radiologically diagnosing focal forms of XGP solely on imaging remains to be a challenge as pertinent features on imaging to help guide providers are mainly directed towards the diffuse forms. This includes the radiologic 'bear paw sign', a unilateral enlarged kidney, a renal pelvis stone and a non-functioning kidney, which can be found on a CT scan.

In recent decades, nephron-sparing surgery has become mainstream in treatment of malignant diseases of the kidney. The accepted curative treatment for XGP is thought to be antibiotic therapy and a radical nephrectomy. Given the rarity of XGP, most of literature on it is limited to case reports or series including multiple reports in the literature of focal unilateral XGP being treated with partial nephrectomy. This treatment in some cases has been intentional and occasionally incidental due to the challenging nature of the XGP diagnosis as partial nephrectomy is not considered to be standard of care for this disease. Kim et al. reported the case of a patient suspected of having XGP treated with partial nephrectomy after intraoperative frozen section confirmed their suspicion. Patient is reported to have done well and benefited from nephron-sparing surgery instead of a radical procedure [10]. Ballentine et al. reported a case of a 58-year-old woman who presented with flank pain and peri-renal haemorrhage involving a fat-containing renal mass who was successfully treated with partial nephrectomy [11]. She was found to have XGP on pathological analysis, although preoperatively she was thought to have haemorrhage from a cancerous tumour. Chlif et al. reported a series where they treated 3 patients with localized XGP with partial nephrectomy [11]. Even though these patients were found to have XGP on pathology, they were all treated with presumed preoperative diagnosis of RCC. In one case reported by Al-Darrab et al., a 44-year-old female patient was treated for RCC by partial

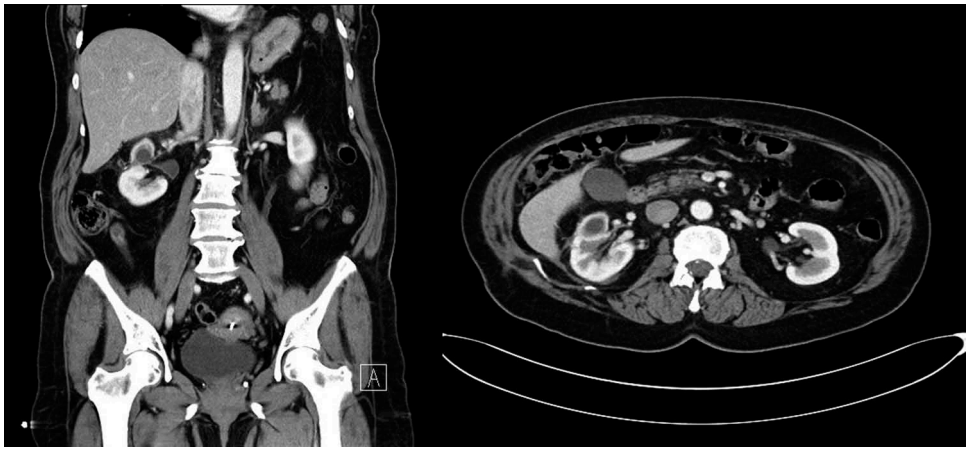

Figure 7. Resolution of right renal mass is shown.

nephrectomy only to return several months later with a presumed local recurrence [12]. Once patient underwent radical nephrectomy, pathology showed she had XGP. A review of her initial pathology, concurred to have had XGP. This case demonstrates that XGP is not only a difficult clinical diagnosis but also can be challenging to diagnose pathologically [13]. This could be due to the fact that despite the classic presence of lipid-laded macrophages, it is easy to misinterpret foam cells as clear cells consistent with RCC [11]. All the above cases have one factor in common: the patient suffering from unilateral focal XGP. This can be easily mistaken for RCC given its radiographic appearance and clinical presentation.

Conclusion

We report a case of a patient with xanthogranulamtous pyelonephritis, as shown on pathology, who was initially suspected to have cystic RCC and was treated with partial nephrectomy. This is an important reminder that rare conditions such as XGP should be on the differential diagnosis for renal masses even when imaging is suggestive of a malignancy. Given the radiographic ambiguity of XGP, it can certainly present a pitfall to even the experienced radiologist. XGP is a rare condition and remains a diagnostic dilemma. When diagnosis is suspected a preoperative biopsy or intraoperative frozen section may be beneficial to help guide decision-making and help expedite appropriate antibiotic therapy if indicated [13]. Evidence in this area remains limited. Review of cases and series published in the literature suggests nephron-sparing surgery may be appropriate for treatment of XGP in select cases.

Disclosure statement

No potential conflict of interest was reported by the authors.

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