Case Report

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Xanthogranulomatous Pyelonephritis with Staghorn Calculus, Acute Gangrenous Appendicitis and Enterocolitis: A Multidisciplinary Challenge of Kidney-Preserving Conservative Therapy

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Key Words

Xanthogranulomatous pyelonephritis • Staghorn calculi • Kidney preserving therapy

Abstract

Xanthogranulomatous pyelonephritis (XP) is a rare form of pyelonephritis and without treatment destructive to the kidney. We describe a 74-year-old Caucasian immunocompetent female patient with XP and multiple abscesses on the upper pole of the right kidney and several impacted obstructing renal calculi in the middle calyx that developed severe colitis and gangrenous appendicitis during therapy. *Proteus mirabilis* was detected as the major pathogen in the urine culture. Kidney preserving therapy was carried out by intensive parenteral bacterial eradication, CT-guided abscess drainage and stone destruction by 3 sessions of extracorporeal shock wave lithotripsy under ureteral stenting. Large tumor masses in XP are often daunting and may lead to a nephrectomy. However, kidney-preserving therapy is possible and should be considered in non-septic patients or in case of a solitary kidney.

Introduction

Xanthogranulomatous pyelonephritis (XP), which accounts for less than 1% of all cases of chronic pyelonephritis, is a rare form of inflammatory/abscess-forming nephropathy and leads to progressive loss of renal function [1]. Obstructive urolithiasis along the urinary tract and ascending urogenital infections are frequently observed. Clinical presentation and imaging of XP are difficult to differentiate from renal cell carcinoma; therefore the diagnosis is usually made postoperatively/histologically. The yellowish tumor masses hold lipid-containing foam cells (xanthoma cells). In literature there are many studies which describe a trend towards surgical procedures [2] due to suspected malignancy and kidney preserving therapies are foregone. We present a complex case of XP with kidney preserving therapy for multiple upper pole abscesses, obstructive nephrolithiasis, and subsequent gangrenous appendicitis followed by enterocolitis. This case report has been approved by the appropriate ethics committee and has therefore been performed in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki and its later amendments.

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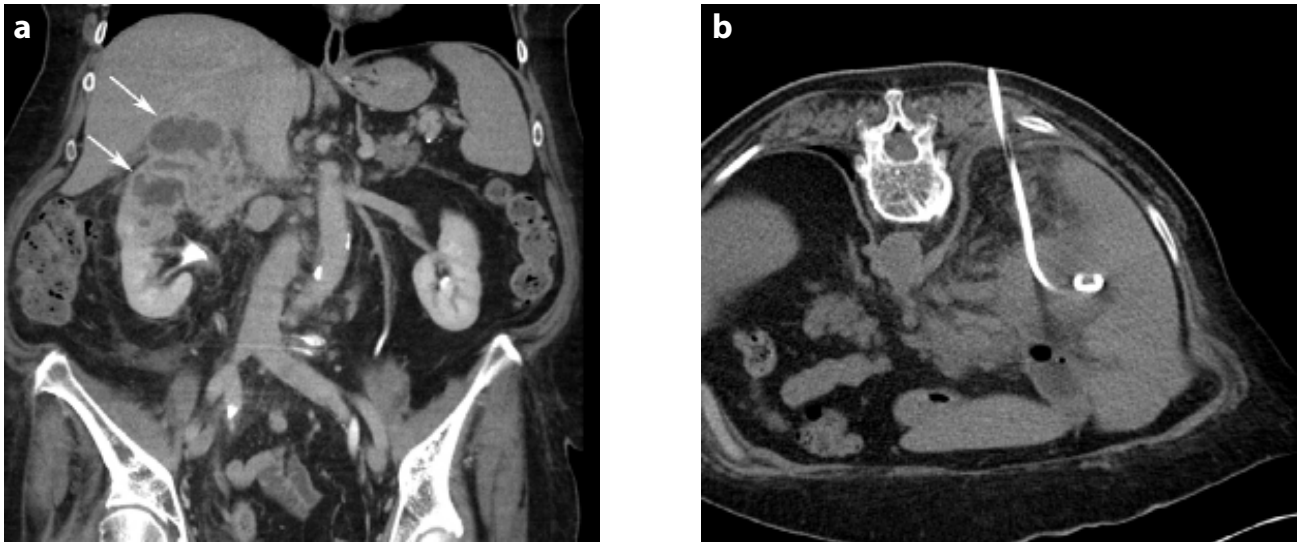

Fig. 1. a MDCT in the coronal plane demonstrates a multiloculated abscess in the upper pole of the right kidney (arrows). There is contrast material present in renal pelvis. **b** CT-guided abscess drainage was performed in prone position.

Case Report

A 74-year-old, immunocompetent Caucasian female patient was admitted to hospital for detailed investigation of a CT-verified diffuse mass in the upper pole of the right kidney with concomitant retroperitoneal lymphadenopathy (diameter > 2 cm) and a calculus in the upper calyx. There was no hydronephrosis, however, there was a high degree of suspicion of a malignant neoplasm (fig. 1a). The patient, a highly mobile woman (Charlson Comorbidity Index: i6), presented subfebrile (37.5°C) with raised inflammatory markers (C-reactive protein 58.4 mg/l), iron deficiency anemia (Hb 8.4 mg/dl, MCV 75 fl, MCH 23.5 pg), reactive thrombocytosis and normal renal function parameters (serum creatinine 0.85 mg/dl , eGFR > 60 ml/min/l using modification of diet in renal disease). Urine culture showed mixed bacterial growth without signs of infection (leukocytes < 10^4).

Recent contrast-enhanced multidetector CT (MDCT) showed a mass in the upper pole of the right kidney with a multi-located abscess and several stones in the renal calyces. After 1 week of parenteral antibiotic therapy (meropenem 500 mg, 3 times daily) repeat MDCT showed slight improvement with a trend towards abscess regression. The subhepatic abscess formation appeared consolidated and encapsulated, the renal calculus with a diameter of 2.5 cm in the middle calyx was unchanged.

CT-guided abscess drainage of the largest fluid collection was carried out introducing a 10F drain and approximately 20 ml of putrid fluid and histological tissue were obtained (fig. 1b). This drain was flushed with 3–5 ml sodium chloride 3 times daily under aseptic conditions, to prevent drainage obstruction and was removed after 7 days. Abundant blood and detritus with a degenerative cell picture and no evidence of malignancy were detected in the aspirate. Additionally *Proteus mirabilis* was grown from the aspirate. The antibiotic therapy was changed to twice daily oral cefuroxime 500 mg. After 40 days of continuous antibiotic cover only minor abscess residues and declining retroperitoneal lymphadenopathy (25–18 mm) could be demonstrated. Blood count (Hb 10.5 mg/dl) and C-reactive protein (7.0 mg/l) were back in the normal range, the urine was sterile. A long-term urinary catheter was never required.

The patient was readmitted with massive right-sided abdominal pain and guarding. CT presented a perforated appendix and an emergency appendectomy was carried out. She received 10 days of antibiotic cover with amoxicillin/clavulanic acid, which led to an improvement of laboratory values (WBC fell from 19.09 to 7.04×10^9/l). Postoperative peritonitis was suspected, but MDCT was unremarkable and showed resolution of XP (fig. 2a).

Stone removal was initiated 3 weeks later by inserting a double-J ureteral stent followed by 3 cycles of extracorporeal shock wave lithotripsy (ESWL). Partial disintegration was achieved after the second ESWL, larger fragments were treated during the third ESWL under ultrasound guidance. Subsequently almost all fragments passed alongside the stent. Stone analysis was performed by X-ray diffraction and demonstrated pure calcium-phosphate.

Meanwhile the patient developed severe *Clostridium* colitis, which was verified by colonoscopy and treated with a 14-day course of parenteral metronidazole 0.5%. The double-J ureteral stent was removed 2 months after insertion, and a renal X-ray showed minimal residual fragments in the upper calyx (fig. 2b), the XP could no longer be detected sonographically. Further follow-up examinations were carried out by the patient's urologist. After 6 months there had been no recurrence; the left kidney was consistently problem-free.

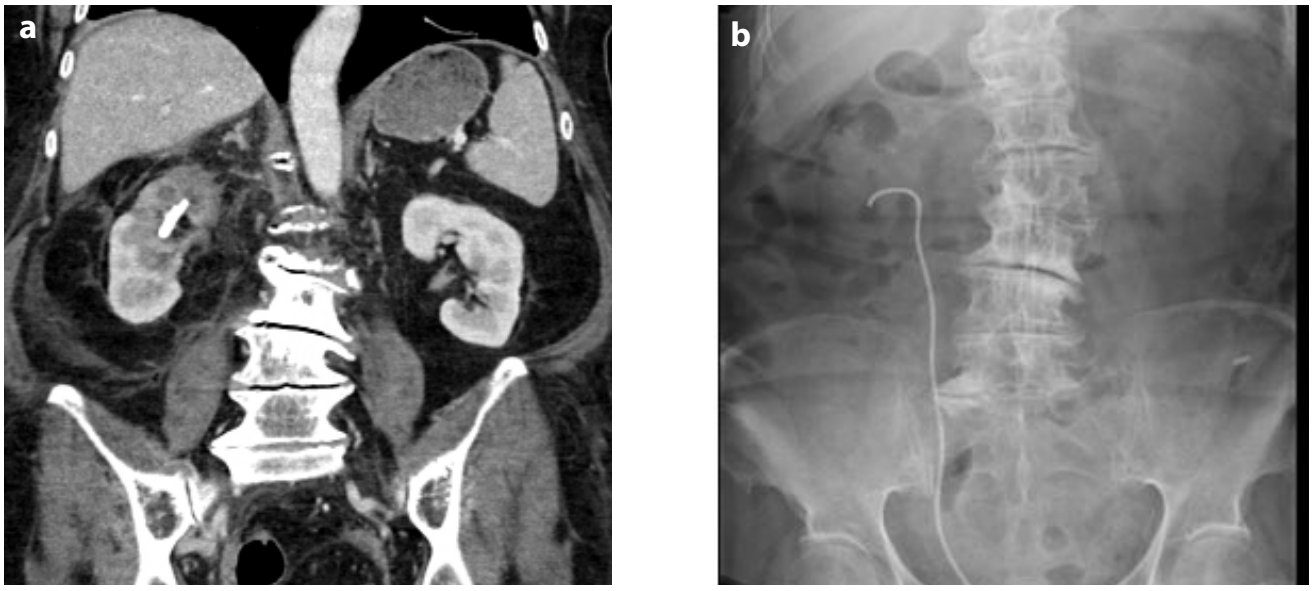

Fig. 2. a Further follow-up MDCT shows resolution of abscesses and scarring of the upper pole. The staghorn calculus is still present. **b** Double J ureteral stent with disintegrated stones after third ESWL.

Discussion

XP is a histologically benign disease, with an annual incidence of 1.4/100,000; it is a rare form of pyelonephritis. A staghorn calculus is present in 34.1% of cases [3]. The inflammation starts in the renal pelvis or calyces, and can consequently segmentally infiltrate the kidney parenchyma and connected organs (psoas muscle, spleen and liver capsule, retroperitoneum). Women are more frequently affected (female-male ratio: 2.5:1) and the mean age of onset is approximately 52–55 years [4]. A New Zealand study observed an increased incidence in Pacific Islanders and Maoris [5].The radiological differentiation from renal cell carcinoma is difficult, however, MDCT is still the diagnostic tool of choice. The incidence in children [6] as well as the coexistence of renal cell carcinoma is rare, and the pathogenetic correlation is unclear [7].

The most common symptoms are subfebrile temperature, flank pain, weight loss and weakness. Pyuria is present in 60–90% and kidney-skin fistulas may arise in 5% of cases [8]. 63% of XP patients are diabetics and in 55% of cases *Proteus mirabilis* is the causative organism for the bacteriuria (which occurs in 90%). Previous urinary tract infections are known in 73% of patients [9]. Clinical features, imaging, urine cytology and renal biopsy may be helpful in diagnosis, but study data show that in case of malignant disease, percutaneous dissemination of tumor cells is possible [9].

Differentiation from renal neoplasia is difficult, therefore in most studies minimally invasive nephrectomy or partial nephrectomy are the primary therapy goal [2]. In our case we were able to show that with sufficient radiologic expertise and correlating clinical course (tumor regression after antibiotic therapy), a conservative therapeutic management can be strived for and that kidney preservation is possible. CT-guided abscess drainage can decrease large perirenal abscess formations immediately. But daily care of the drainage and maintenance by flushing is important to prevent obstructions and non-functioning [10]. The advantage of kidney preservation must be weighed against the possible additional morbidity compared to standard procedure with nephrectomy [4]. In septic patients and treatment-resistant cases, the emergency nephrectomy is the treatment of choice.

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