Xanthogranulomatous Pyelonephritis with Pyonephrosis and Renal Abscess in a Young Adult: A Consequence of Neglected Urinary Tract Infection Leading to Nephrectomy

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ABSTRACT

Xanthogranulomatous pyelonephritis (XGP) is a rare form of chronic pyelonephritis, which is challenging to diagnose because its clinical presentation mimics other entities and is commonly associated with a history of urinary tract obstruction. We report a case of XGP in a young adult without nephrolithiasis and urinary tract obstruction. A 23-year-old woman presented with intermittent abdominal pain in the right upper quadrant persisting for the last ten months. The pain was dull, poorly localized, and started spreading to the right back, right shoulder, and right thigh in the last three months. Other complaints included fever, chills, pain during urination, and nausea. The patient had a history of infrequent urination, recurrent urinary tract infections (UTIs), and a low fluid intake. A physical examination revealed that the patient had right upper quadrant abdominal tenderness and right costovertebral angle tenderness. Laboratory findings showed leukocytosis and neutrophilia. The radiological examination revealed a round mass in the superior pole of the right kidney with mixed cystic and solid components, and a well-defined margin. It further enlarged from 4.5 cm to 10.6 cm in diameter in three months. The urologist performed a total right nephrectomy. The histopathological examination showed XGP with renal abscess. Proteus mirabilis was identified from the pus specimen culture. XGP should be considered in the diagnosis of patients having chronic UTI presented with or without the findings of urinary tract obstruction.

Keywords: *Xanthogranulomatous pyelonephritis, pyonephrosis, renal abscess, urinary tract infection, nephrectomy.*

INTRODUCTION

Xanthogranulomatous pyelonephritis (XGP) is a rare form of chronic pyelonephritis, which has a challenging diagnosis because of its resemblance to other renal pathologies. XGP is a rare entity that accounts for only 0.6% of histologically documented cases of chronic pyelonephritis.[1] Women are predominantly affected (the male-to-female ratio ranges between 0.5 and 0.6), usually in mid-life years (40–50 years).[2,3] This chronic destructive granulomatous inflammation of renal parenchyma often results in a complete loss of function in the affected kidney.

The exact etiology of XGP is still yet to be discovered, but it appears to be multifactorial. Two of the most common risk factors are urinary tract obstruction and infection, with obstruction playing a major role in pathogenesis.[4] The most common cause of obstruction is nephrolithiasis, which is present in 90% of XGP cases.[2,3] Other causes include ureterolithiasis, ureteral stenosis, pyelo-ureteral junction abnormalities, and pyelo-ureteral duplicities. XGP is also associated with recurrent urinary infections, with common pathogens such as *Escherichia coli* and *Proteus mirabilis*.

The difficulty of diagnosing XGP arises from the fact that it may closely mimic other diseases of the kidney. This nature has earned XGP the title of "the great imitator".[5] The findings in XGP are often difficult to distinguish from pyonephrosis, renal abscess, renal tuberculosis, and renal cell carcinoma by the clinical symptoms, physical examination, and radiological findings.[6] The diagnosis is only confirmed by pathological examination after nephrectomy.

We present an unusual case of XGP in a young adult without urinary obstruction and nephrolithiasis. The case was complicated by pyonephrosis and renal abscess caused by *Proteus mirabilis*, which further led to nephrectomy. A pathological examination confirmed XGP after total nephrectomy.

CASE ILLUSTRATION

A 23-year-old woman presented with intermittent abdominal pain in the right upper quadrant persisting for the last ten months. The pain was dull and poorly localized, with a score of 8 out of 10 on the numerical rating scale. The abdominal pain started spreading to the right back, right shoulder, and right thigh from the past three months. The patient also complained of fever, chills, pain during urination, and nausea. There were no complaints of darker or turbid urine. The medical history was significant for untreated recurrent urinary tract infections (UTI) for the last three years. She said that she rarely drank water in the last five years, only consuming around 400 mL a day. She denied any history of drug use, sexual activity, and urine catheter usage. She usually changed underwear twice a day and kept the genital area clean. The physical examination revealed that the patient had normal vital signs, right upper abdominal tenderness, and right costovertebral angle tenderness.

Complete blood counts showed leukocytosis ($12.05 \times 10^3/\mu L$) and neutrophilia (76.8% neutrophils). Urea and creatinine blood levels were within normal limit, 18 mg/dL and 0.52 mg/dL respectively. The estimated glomerular filtration rate (eGFR) was 133 ml/min/1.73 m^2. Other laboratory tests were also within the normal range. An ultrasound examination revealed a round mass, 4.5 cm in diameter, in the superior pole of the right kidney with cystic and solid components, and well-defined margins (**Figure 1**). Three months later, a computed tomography scan of the abdomen showed a cystic mass measuring $10.6 \times 8 \times 7.72$ cm in the superior pole of the right kidney suppressing the pelvicocalyceal system, accompanied by perirenal fat obliteration and lymphadenopathy (**Figure 2**).

The patient was diagnosed with a kidney mass. Therefore, a urologist performed a total nephrectomy because of the non-functional kidney. Surprisingly, the kidney was found to be enlarged and contained approximately 200

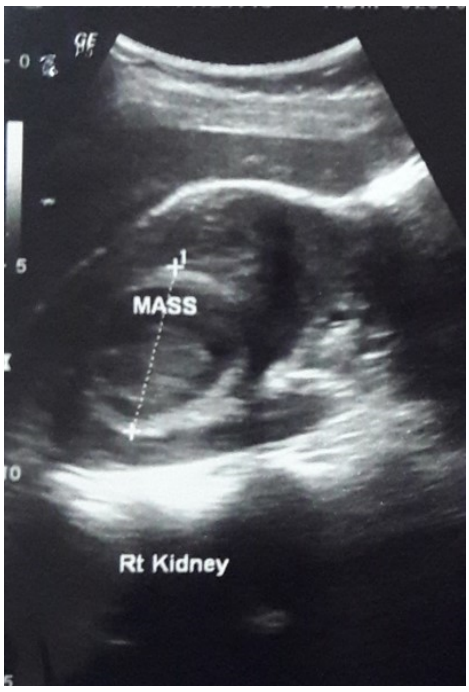

Figure 1. Ultrasonography of the right kidney showed a round, well-defined, mixed cystic and solid mass with a diameter of 4.5 cm in the superior pole of the right kidney

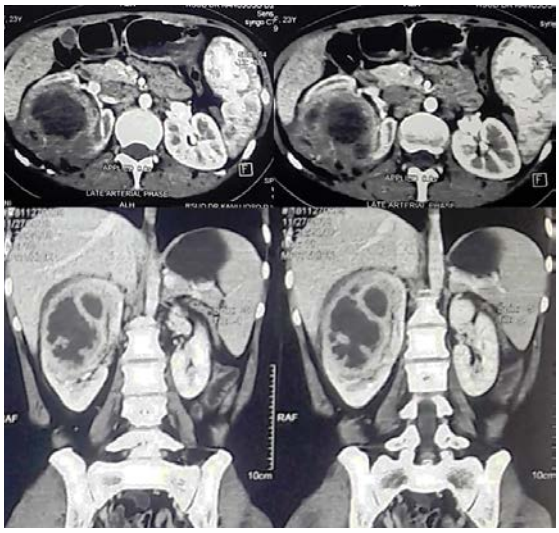

Figure 2. A contrast abdominal CT scan in the transverse plane (upper) and coronal plane (lower) showed a kidney mass measuring 7.72 × 8 × 10.6 cm in the superior pole of the right kidney suppressing the pelvico-calyceal system with perirenal fat obliteration and lymphadenopathy

ml of pus intraoperatively. The histopathological examination revealed the granulomatous inflammation of the renal parenchyma comprising many xanthomatous histiocytes with foamy cytoplasm and multinucleated giant cells surrounded by lymphocytes, areas of hemorrhage, and necrosis with numerous neutrophil infiltrations, thereby indicating XGP with renal abscess (**Figure 3**). Cultures from the

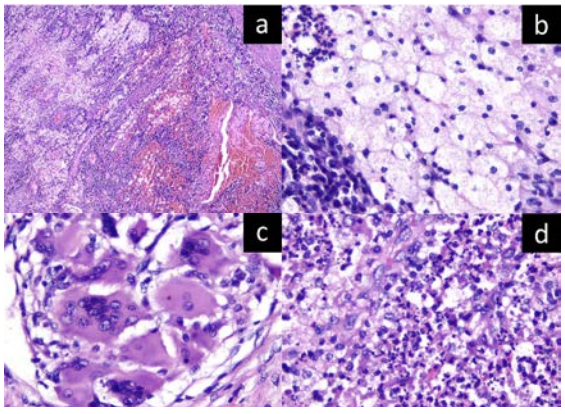

Figure 3. Histopathologic examination showed: (a) granulomatous inflammation of the renal parenchyma, xanthomatous histiocytes with foamy cytoplasm, and areas of hemorrhage and necrosis, indicating xanthogranulomatous pyelonephritis with renal abscess (hematoxylin and eosin (HE) stain, 40x). Higher magnification showed (b) xanthomatous histiocytes (HE, 400x), (c) foreign body-type histiocytic giant cells (HE, 400x), and (d) collection of inflammation cell infiltrate, dominated by neutrophils with few lymphocytes, plasma cells, and histiocytes (HE, 400x).

pus showed *Proteus mirabilis* infection without any antibiotic resistance. The patient was treated with parenteral ceftriaxone and metronidazole postoperatively. The patient was followed up thrice after the nephrectomy. The patient was discharged from the hospital one week after the surgery, when the operation wound was already dry; however, there was still a feeling of pain in the wound area. One week after the first follow-up, the pain had lessened and the wound edges appeared to have fused on inspection. Three months after the surgery, the surgical wound had healed completely and the patient did not have symptoms of any kind. Follow-up of renal function test showed normal results (blood urea 26 mg/dL, blood creatinine 0.8 mg/dL, and eGFR 103 ml/min/1.73 m^2).

DISCUSSION

The diagnosis of XGP is rare and challenging because of its non-specific clinical symptoms, laboratory, and radiologic findings. This condition typically occurs in middle-aged women and is mainly linked to urinary obstruction. However, the patient described in the presented case was a 23-year-old woman without any evidence of underlying urinary obstructions, thus presenting a potential diagnostic pitfall.

Epidemiologically, xanthogranulomatous pyelonephritis is seldom found in a young, otherwise healthy adult. An XGP case in a young adult has been reported previously but it was in the presence of a staghorn calculi.[7] The absence of urinary obstruction further makes the occurrence of XGP more unlikely in the previously described patient. Ultrasonography and CT scans did not find nephrolithiasis or ureterolithiasis. No anatomical abnormalities were observed from radiology and intraoperative assessment. Despite the low possibility, XGP was confirmed in the patient. Similar conditions have also been reported in other case reports where XGP was confirmed by the histopathologic examination in patients who did not have a history of urinary tract obstruction.[8,9]

The only related risk factor found in the aforementioned patient was recurrent and chronic urinary infections. XGP can arise as a complication of chronic urinary infections.[2]

The recurrence and chronicity of the urinary infections could occur from the patient's low fluid intake and infrequent urination. Drinking less than 1.5 L of total fluid daily is associated with more frequent urinary infections in women.[10] Adequate treatment and related lifestyle changes can prevent complications from a urinary infection.

The management of XGP depends on the extent of the disease. The current treatment of choice for diffuse XGP is surgical intervention. If XGP is found in the earlier stages with a smaller lesion size, then antibiotics can be attempted as an initial treatment.[6] The diffuse XGP in the patient had resulted in a non-functioning kidney; hence, the urologist decided treatment with nephrectomy. An early recognition and management of recurrent UTIs and the XGP itself might have prevented the necessity of nephrectomy in the described patient.

CONCLUSION

XGP is a rare type of chronic pyelonephritis, whose diagnosis is challenging preoperatively. The described patient was quite atypical as she was a young adult with no evidence of urinary obstruction. The only risk factor found in the patient was recurrent UTIs. Clinicians should still consider XGP in patients with recurrent UTIs in all ages, even without the presence of risk factors. An early diagnosis and treatment of the UTIs and XGP could prevent unnecessary morbidity caused by nephrectomy.

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