

Case Report

Xanthogranulomatous Pyelonephritis with Spontaneous Nephrocutaneous Fistula

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Abstract

Xanthogranulomatous pyelonephritis is a severe, rare, form of chronic pyelonephritis characterized by the destruction of the renal parenchyma and replacement by granulomatous tissue. This disease has been called as the “great imitator” because clinical and radiological findings often mimics other inflammatory or neoplastic renal disorders leading to diagnostic challenge before histopathologic analysis. Herein, we report a case of a 72-years-old woman who presented with a 4-year history of left flank pain with intermittent lumbar purulent discharge. The CT scan findings advocated a pyonephrosis. After a left nephrectomy, the per-operative and macroscopic appearance of the mass was in favor of a renal replacement lipomatosis. Microscopic examination finally revealed the diagnosis of Xanthogranulomatous pyelonephritis. Clinicians, radiologists and pathologists should be aware of this rare “great imitator” entity.

INTRODUCTION

Xanthogranulomatous pyelonephritis (XGPN) is an atypical severe form of chronic pyelonephritis characterized by the destruction of renal parenchyma and replacement with a chronic inflammatory infiltrate of lipid-laden macrophages known as xanthoma cells [1]. Its name is derived from yellow color on gross pathology and a granulomatous reaction histologically. The exact pathogenesis is still unclear but appears to be multifactorial including: recurrent urinary tract infection, genitourinary obstruction, nephrolithiasis, altered immunological anomalies and abnormal lipid metabolism [2]. XGPN is rare, varied from 0.6 to 1.4% of cases of renal inflammation [3]. Unlike chronic pyelonephritis, it may spread to the perinephric tissue with formation of abscesses and even fistulas [4].

This disease has been called as the “great imitator” because clinical and radiological findings often mimics other inflammatory or neoplastic renal disorders leading to diagnostic challenge before histopathologic analysis [2,5,6]. This disease has rarely been reported in Africa and is unknown in Cameroon

Herein, we report the case of a 72-year-old female presenting with chronic left flank pain and discharge. After abdominal CT

scan, the diagnosis of pyonephrosis was made. A left radical nephrectomy was performed and macroscopically pyonephrosis was eliminated and we thought about renal replacement lipomatosis. Finally, the histopathologic examination was consistent with the diagnosis of XGPN.

CASE REPORT

A 72-years old woman was admitted in our service complaining of chronic pain of the left flank. She had a 4-year history of left lumbar pain treated as rheumatism with anti-inflammatory drugs as automedication without improvement. Two years after the onset of the pain, she presented an intermittent purulent left lumbar discharge through a sinus and associated with off and on fever. This lumbar discharge was managed as an abscess, and she underwent incision and drainage 4 times by local practitioners with persistence of the discharge and recurrence of symptoms. Besides the pain and the fever, she has lost more than 15 kg for the last past years.

She didn't complain about hematuria or dysuria. She had no previous history of diabetes or documented urinary tract infection.

On physical examination, a sinus and skin scarring were noted

in the left lumbar region which was tender on palpation. Analytical evaluation revealed a leukocytosis with increase in neutrophil count, a mild anemia and no azotemia. Urine culture was sterile and pus culture positive for *Escherichia Coli*. An abdominal CT scan showed a large reniform left mass with severe renal parenchyma atrophy, a central staghorn calculus and peripheral enhancement. Renal parenchyma was replaced by multiple low attenuation areas advocating abscess cavities (Figure 1). The left kidney wasn't excreting and the right kidney was normal. The radiological diagnosis advocated was left pyonephrosis.

Patient underwent a left nephrectomy through open transabdominal approach. There was a diffuse inflammation and thickening of Gerota' fascia and it was difficult to dissociate the mass from adjacent organs mainly in the renal hilum because of many adhesions and intense inflammation. No abscess cavity was noted. Macroscopically, the resected specimen was yellowish. Cut section showed a friable yellow fatty mass replacing the entire renal parenchyma with a central staghorn calculus. There were similarity in color between the fat tissue inside the renal pelvis and perirenal fat with no purulent material. These per-operative and macroscopic findings were not compatible with pyonephrosis and we thought about renal replacement lipomatosis with nephrocutaneous fistula (Figure 1). Histopathology revealed diffuse histiocytes with abundant foamy cytoplasm, lymphoplasmatic inflammatory cells and Ziehl Nelsen stain was negative for acid fast bacilli. The final diagnosis was then Xanthogranulomatous pyelonephritis.

Post-operative broad spectrum antibiotics were given. The postoperative course was uneventful and the patient was discharged home at day 9.

DISCUSSION

XGPN was first described in 1916 by Schlagenhauer [7]. It's a chronic and unusual infectious inflammatory condition involving the renal parenchyma in which lobulated masses diffusely replace the renal architecture [2]. XGPN is considered as a granulomatous reaction to severe obstruction secondary to stone, stricture or rarely tumor [5]. In 83% of cases, a calculus is found and it's a staghorn one in half of the cases [8]. This disease is prevalent in all age group with a peak incidence in the 5th to 6th decade [5] and is more common in females [2,9]. XGPN affects both kidneys with equal frequency, but bilateral forms have been reported [2,10,11]. The most commonly reported symptoms are fever, abdominal and/or flank pain, weight loss and lower urinary tract infections [5]. Urinary cultures are usually positive to *Escherichia coli*, *Proteus mirabilis* and rarely *Staphylococcus aureus*, *Pseudomonas* and *Klebsiella* [12].

Three features make this case a distinctive one.

First, the low incidence of the disease. XGPN is a rare type of pyelonephritis found in 0.6 to 1.4% of cases of renal inflammation [3] and to our knowledge this is the first reported XGPN case in our country.

Second, the presence of nephrocutaneous fistula. Nephrocutaneous fistulas are rare complications of XGPN, reported only in 5% of the cases [13,14]. Renocolic fistula has also been described [15].



Figure 1 Abdominal CT scan showing a large reniform left mass with severe renal parenchyma atrophy, a central staghorn calculus and peripheral enhancement. Renal parenchyma was replaced by multiple low attenuation areas advocating abscess cavities.

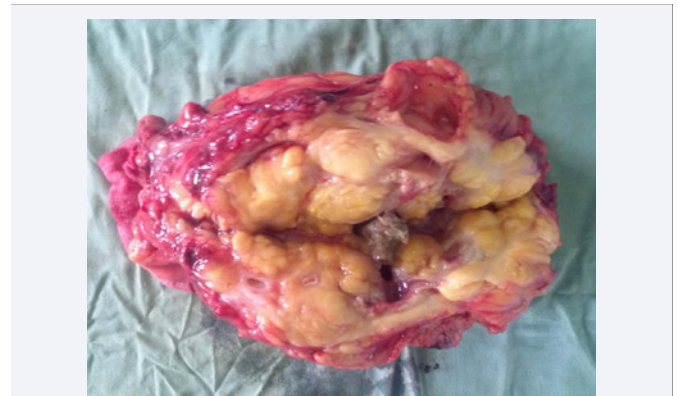


Figure 2 Macroscopic cut section showed a friable yellow fatty mass replacing the entire renal parenchyma with a central staghorn calculus. There was similarity in color between the fat tissue inside the renal pelvis and perirenal fat with no purulent material.

Third and main point, the diagnosis difficulties: The varied clinical presentation of XGPN and its non-pathognomonic radiologic findings can often make this diagnosis a real challenge for physicians. Even in developed countries with all health care structure facilities like CT scan and magnetic resonance imaging, misdiagnosis before histopathology analysis have been reported and XGPN can be confused with: pyonephrosis, renal cell carcinoma and renal replacement lipomatosis [3,5,6,16,17]. This led some authors to call this disease the "great imitator" and in our case it justified this name. The differential diagnosis of XGPN include neoplastic diseases such as clear-cell carcinoma, lymphoma, leukemia, Wilm's tumor, neuroblastoma and inflammatory processes (renal or peri-renal abscess, pyonephrosis, renal tuberculosis, focal and diffuse nephritis, fungal infection, renal replacement lipomatosis disease) [13,14]. Between all these conditions, the main differential diagnosis is renal replacement lipomatosis. Table (1) resumes the main features of both of them and they appear to be very similar [5,17,18,19]. However, we missed the diagnosis because of lack of knowledge about this clinical entity. In developing countries

Table 1: Comparative characteristics of xanthogranulomatous pyelonephritis and renal replacement lipomatosis [5,17,18,19].

	Renal Replacement Lipomatosis	Xanthogranulomatous Pyelonephritis
Pathogenesis	Severe loss of renal parenchyma with deposition of fat and fibrous tissue in the sinus and perirenal space associated with a long-standing inflammation Calculus disease, bacterial infections and renal obstruction involved	Chronic granulomatous inflammatory disorder of the kidney with increased lipid-laden foam cell infiltrating and substitute renal tissue resulting in a lipomatous degeneration Calculus disease, bacterial infections and renal obstruction involved
Clinic		
Age	Usually between fifth and seventh decade of life	Usually between fifth and seventh decade of life
Signs and symptoms	Flank pain or mass, recurrent urinary tract urinary, hematuria, fever, weight loss Sometimes asymptomatic	Flank pain or mass, recurrent urinary tract urinary, hematuria, fever, weight loss Sometimes asymptomatic
CT-scan	Marked parenchymal atrophy, abundant adipose tissue in renal sinus and perirenal spaces.	Large reniform mass with peripheral enhancement. Loss of renal architecture with large and heterogeneous low density areas and extra-renal extend of the disease. True fat density not found and presence of air inside the urinary tract
During surgery	Fat more fibrous than normal fat	Severe adhesions and infiltration
Macroscopically	Enlarged kidney with gross fibrofatty appearance. Atrophic renal parenchyma replaced by fatty tissue proliferation. Similarity between the fat tissue inside the renal pelvis and the perirenal fat	Enlarged kidney with gross fibrofatty appearance. Mass of yellow/green tissue with focal necrosis and hemorrhage. Fatty tissue inside the renal parenchyma is paler than perirenal fat.
Histology	Large fat cells outside the renal parenchyma with a sharp demarcation between the adipose tissue and the renal parenchyma showing that there is no real invasion of the kidney by the fat but merely of fat as it atrophies	Increase of lipid-laden macrophages (xanthoma cells) inside the renal parenchyma

as our own, the low incidence of XGPN associated to mal-practice and lack of awareness by clinicians, radiologists and pathologists may be responsible for misdiagnosis and underreporting. The diagnosis in our patient was missed by practitioners for 4 years. Even if XGPN don't have pathognomonic signs, CT scan remains the most valuable method for pre-operative diagnosis; absence of true fat density, presence of air inside the urinary tract and perinephric extension to adjacent organs are indicative of XGPN. Misinterpretation of "foam cell" as "clear cell" consistent with renal adenocarcinoma is the most important diagnostic challenge at histology.

Definitive treatment of XGPN is en bloc surgical resection of the involved kidney.

CONCLUSION

Xanthogranulomatous pyelonephritis should be considered in the differentials of flank mass. Especially in patients with long standing fever and flank pain. Clinicians, radiologists and pathologists should be aware of this rare "great imitator" entity.

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