

Case Report

Xanthogranulomatous pyelonephritis: a case review of two cases

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ABSTRACT

Xanthogranulomatous pyelonephritis is a chronic destructive granulomatous inflammation of the renal parenchyma. It was first described by Schlagenhauser in 1916 and then Oberling named the disease as Xanthogranulomatous pyelonephritis in 1935. It represents 1% of all renal infections. In this report we present two cases of Xanthogranulomatous pyelonephritis along with radiological assessment.

Keywords: Xanthogranulomatous pyelonephritis, Diffuse, Focal, Staghorn

INTRODUCTION

It is a chronic destructive granulomatous inflammation of the renal parenchyma. Xanthogranulomatous pyelonephritis was first described by Schlagenhauser in 1916.¹⁻⁸ Oberling named the disease as Xanthogranulomatous pyelonephritis in 1935.⁹ It represents 1% of all renal infections.⁹

CASE REPORT

Two patients, one 46 years old male patient presented to the department with complaints of pain on left half of the abdomen for the past one month. The pain was intermittent in nature.

- There was a past history of nephrolithotomy on the left side.
- The patient was non-diabetic, non-hypertensive and was not under any medication.

Another patient, a 65 years old male patient came with complaints of disorientation to time, place and person with high grade fever. Laboratory investigations showed markedly deranged urea and creatinine. The patient was non-diabetic, non-hypertensive.

- Abdominal CT examination was performed on Siemens Somatom Emo 6 machine with 6 mm and 2 mm sections after bowel opacification using oral and i.v iopamidol for patient one, while only plain study was done for patient two because of deranged renal function tests.

Case 1

- Left kidney showed variegated? Fat containing lesion seen replacing renal sinus fat and extending inferiorly causing thinning of renal parenchyma along lower pole.
- The lesion was seen surrounding left renal pelvis and upper ureter.
- Peri renal fat stranding seen along inferior pole of left kidney effacing left psoas muscle.
- Extra renal stranding seen extending upto subcutaneous planes along left lateral abdominal wall.
- Areas of calyctasis were seen especially in upper pole.
- Considering h/o operated left renal calculus findings could suggest Xanthogranulomatous pyelonephritis.

Case 2

- Left upper ureteric calculus with multiple left renal calculi was noted. Distorted left renal architecture with extensive fat stranding and peri-nephric component seen –s/o xanthogranulomatous pyelonephritis.
- Associated large vesical calculus noted.
- Large complex hypodense appearing lesion with cystic appearance in left para-spinal region extending along left ilio-psoas muscle laterally, s/o ilio-psoas abscess.
- In this patient, there was left ureteric calculi leading to xanthogranulomatous pyelonephritis and associated complication of left ilio-psoas abscess.

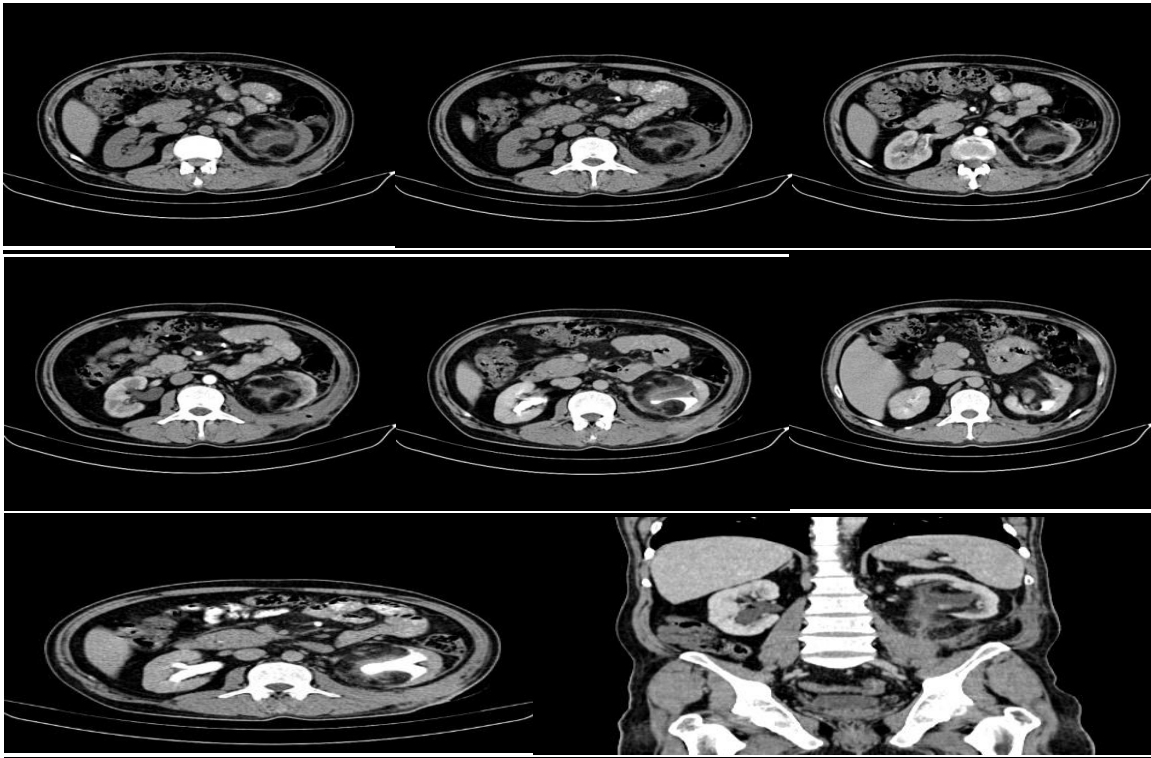


Figure 1: CT scan images of patient one showing findings s/o xanthogranulomatous pyelonephritis.

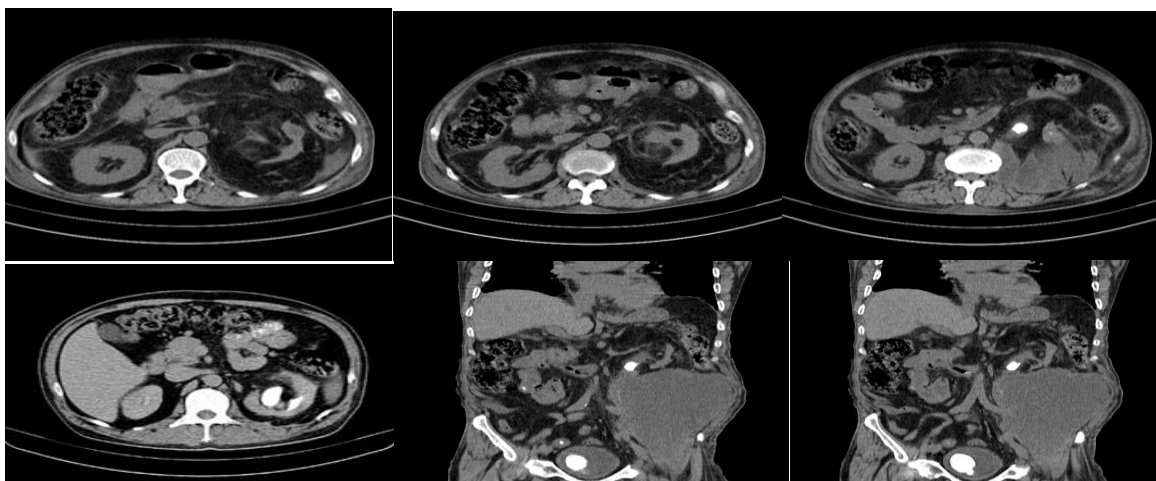


Figure 2: CT scan findings in patient two showing left ureteric and multiple left renal calyceal calculi, vesical calculus, Ilio-psoas abscess and features of xanthogranulomatous pyelonephritis.

DISCUSSION

It is a chronic destructive granulomatous inflammation of the renal parenchyma.¹⁻⁴

It is most commonly seen in immunocompromised patients and have associated urinary tract infection and/ or urolithiasis.^{3,8-11}

It is characterized by infectious phlegmon arising in the renal pelvis and extending into the medulla and cortex which are gradually destroyed and replaced by lipid laden macrophages (Xanthoman cells).^{2-6,9-12}

Three forms of the disease have been recognized.^{1,12} –

- Diffuse – It is most common in adults¹¹
- Segmental – There is segmental involvement of the disease.
- Focal – Located within the cortex with no pelvic communication. It is also known as tumefactive.¹¹ It is mostly seen in children and women.¹¹

Xanthogranulomatous pyelonephritis is seen in patients suffering from long term obstruction and infection.^{1,6,8,9}

It is mostly associated with calculi, especially staghorn.^{1,2,6,8,9}

The most common calculus is calcium oxalate. Other calculi that can be present is a combination of calcium carbonate and calcium phosphate.¹²

Other predisposing factors are

PUJ obstruction and Bladder tumor.^{1,8}

Malek and Elder proposed three stages of xanthogranulomatous pyelonephritis^{1,3,5,8} –

Stage 1

- It is also called the nephric stage.
- Disease here is confined to the renal parenchyma.

Stage 2

- It is also called the peri-nephric stage.
- Disease extends into the peri-nephric space (Gerota's fat).

Stage 3

- It is also called the para-nephric stage.
- The disease extends into peri- and para-nephric stage.
- The disease may spread to peri-nephric tissue with formation of abscesses and even fistula.^{5,6}
- It may also develop in renal transplant.⁸

- It is most commonly seen in 5th to 6th decade of life and is more common in females (1 to 12).
- It can however be seen in neonates as well.¹
- Flank pain is the most common symptom.^{1,2,4,5,12}
- Other symptoms are fever, gross hematuria, pyuria, lower urinary tract symptoms, palpable mass.^{1,2,4,5,12}
- It is usually unilateral but bilateral disease has also been reported and is generally fatal.^{5,6,8,9}
- The disease involves the left kidney most often.⁸
- Liver function tests may be abnormal because of hepatorenal syndrome and return to normal after surgery.^{4,8}
- The laboratory picture is that of leukocytosis with raised ESR and anemia.^{1,3,6,7,8,12}
- The most common organisms implicated in the development of disease are *E. coli* and *Proteus mirabilis*.^{1-5,7-10,12}
- Grossly, the kidneys are enlarged with thickened capsule and yellow nodules with or without central necrosis.⁸
- Microscopically, there are lymphocytes, plasma cells, neutrophils and large macrophages.^{1,5} There is granulomatous mixed inflammatory infiltrate with fibrosis and cholesterol clefts in the background.¹
- Other features seen histologically are – renal tubular atrophy, tubular dilatation and focal squamous metaplasia of the urothelium.¹
- It is CD 68 positive.¹

Radiological features

- No radiological feature is diagnostic.⁸
- CT – It is the mainstay of diagnosis as it differentiates the renal mass and also delineates the involvement of other organs.^{1,2,5,9,10,12}
- It helps in diagnosis of presence and extension of extra-renal involvement.⁹
- Diffuse disease is noted in most cases.¹

A triad of^{5,8}

- Unilateral renal enlargement²
- Non functioning kidney.²
- Large calculi in the renal pelvis.¹⁰

Other CT features are

- Hydronephrosis.
- Intra-parenchymatous collection.⁸
- Renal cortical atrophy.^{1,9,10}
- Inflammatory changes in peri-nephric fat.²
- Focal areas of parenchymal destruction filled with pus and/or debris, there is rim enhancement of these low density lesions and there is also extra-renal involvement.¹⁰
- Obscuration of ipsilateral psoas margin.²
- Thickening of Gerota's fascia and thick enhancing septa in the hypodense areas in renal parenchyma.¹¹

Bear paw sign

Rounded areas of water density representing dilated calyces and abscess cavities with pus and debris in diffuse xanthogranulomatous pyelonephritis.

Focal form

Poorly enhancing mass adjacent to calyx or in one pole of a kidney with duplicated collecting system.¹⁰

USG

Enlarged kidney with large amorphous central echogenicity that corresponds to a renal pelvic staghorn calculus.^{1,2}

Diffuse form¹²

Generalized renal enlargement with multiple hypo-echoic areas representing calyceal or pelvi-calyceal dilatation and parenchymal destruction, hyper-echoic foci with clean posterior acoustic shadowing representing staghorn calculus and hydronephrosis may be seen.

Focal form¹

Non-specific features and it is virtually impossible to differentiate from renal abscess or nephrotic renal cell carcinoma.

Abdominal X ray

Renal calculus is seen¹

MRI

- Sensitive for identifying the accumulation of lipid laden macrophages as high intensity signal on spin echo, T1WI.¹
- Treatment of diffuse or advanced disease is usually with nephrectomy.^{1,3,7-10}
- For focal xanthogranulomatous pyelonephritis, conservative nephron saving approach is used.^{3,9,10}
- Antibiotics alone is not effective.^{9,10}

Differential diagnosis

- Renal cell carcinoma.^{1,9,11,12}
- Parenchymal malakoplakia.⁹
- Megalocytic interstitial nephritis.⁹

CONCLUSION

The most typical features on CT are unilateral renal

enlargement and extra-renal extension of inflammatory changes.

Their presence in a patient with constitutional symptoms and UTI should alert the radiologist to the possibility of xanthogranulomatous pyelonephritis.

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