



XANTHOGRANULOMATOUS PYELONEPHRITIS IN AN ADULT MALE

Radiology

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ABSTRACT

Xanthogranulomatous Pyelonephritis (XGN) is a rare form of chronic pyelonephritis which is characterized by focal or diffuse destruction of the renal parenchyma and replacement by solid sheets of lipid-laden macrophages (xanthoma cells). It was first described in 1916 by Schlagenhafer. It can be seen in all age groups, but most commonly seen in middle-aged to elderly patients (45-65 years). More commonly seen in females with 4:1 female: male predilection, mostly related to an increased incidence of urinary tract infections and thus struvite (staghorn) calculi. Increased incidence is also seen in patients with diabetes mellitus.

In the past, the preoperative diagnosis was extremely difficult due to its nonspecific clinical presentation and radiographic appearances. Now a days, the ability to diagnose it preoperatively has considerably improved with the use of ultrasound and computed tomography.

Clinical presentation is non specific, consisting of constitutional symptoms such as anorexia, weight loss, malaise, and low-grade fever. Haematuria and flank pain can also be present.

CT scan is investigation of choice for its diagnosis.

KEYWORDS

Xanthogranulomatous Pyelonephritis, Xanthoma, Staghorn calculus

CASE HISTORY

A 67 years old Indian male patient presented to surgery OPD with complains of right sided flank pain, low grade fever and weight loss during last three months.

On clinical examination, a small lump was palpable in right lumbar region. The patient was referred to radiology department for further investigations.

Patient was advised USG, which showed enlarged right kidney with abnormal collection in the right renal parenchyma with gross dilatation of upper and mid calyces with thinned out cortex with internal echoes in it. Large staghorn calculus was seen in the renal pelvis causing mild to moderate hydronephrosis without any hydroureter. Multiple small calculi were also noted in right upper, mid and lower calyx of varying sizes.

Owing to findings of USG patient was then advised CE-CT KUB, which revealed enlarged right kidney with altered axis as hilum facing anteriorly.

A 3.2 x 2.7cm sized partially obstructive calculus (+981HU) was seen in the right renal pelvis causing mild to moderate right hydronephrosis with renal parenchymal thinning. Right ureter was non-dilated. Multiple variable sized non obstructive calculi were seen within the right upper, mid and lower pole calyces with sizes varying from 5-18mm Significant abnormal urothelial thickening and enhancement was seen involving the right renal pelvi-calyceal system with significant peri-nephric fat stranding.

Abnormal hypodense peripherally enhancing collections were seen within the right renal parenchyma and right peri-nephric fat stranding largest of size measuring 4.2 x 2.6cm with peri-nephric fat stranding and thickening of the right anterior and posterior renal fascia.

No excretion of contrast into the right renal pelvi-calyceal system was seen even on 2hrs delayed scan Histopathological report thus confirmed our diagnosis of xanthogranulomatous pyelonephritis with typical appearance of demonstrating abundance of lipid-laden macrophages (Xanthoma cells), lymphocytes and plasma cells

DISCUSSION:

Xanthogranulomatous Pyelonephritis (XGN) is a chronic renal infection that mostly leads to scarred contracted renal pelvis, dilated calyces and diffuse infiltration of the renal parenchyma by plasma cells and lipid laden macrophages (xanthoma cells). It was first described in 1916 by Schlagenhafer. The exact etiology is still not clear, but patients are typically middle aged and diabetic females, who commonly present with recurrent urinary tract infections, flank pain, hematuria, occasionally fever and weight loss.

The main predisposing factor for it is, an obstructing staghorn calculus present in almost 75% of cases. It is postulated that renal obstruction promotes recurrent renal tract infections that lead to an abnormal immune response that is responsible for the chronic changes and parenchymal destruction.

In earlier times, its nonspecific clinical presentation was responsible for difficult preoperative diagnosis, and as a result, majority of the cases were diagnosed on histology after radical nephrectomy was performed.

While clinical diagnosis is difficult, the pathological features of XGN are usually characteristic. Histologically, there is diffuse infiltration of the renal parenchyma by plasma cells and lipid laden macrophages (**xanthoma cells**).

Computed tomography is investigation of choice for it, as the findings on USG and urography are nonspecific.

On CT, it presents with (a) a large staghorn calculus; (b) enlargement of the kidney (or of a segment); (c) poor or no excretion of contrast into the collecting system; and (d) multiple focal low-attenuation (-10 to +30 HU) masses scattered throughout the involved portions of the kidney. The low attenuation collection represents dilated, debris-filled calyces and xanthoma collections. Small calculi are often present within the calyces, and scattered flecks of calcification may be detected within the destroyed parenchyma MRI findings of XGN are nonspecific.

On urine culture, bacteria are cultured from the urine in approximately two third of patients. Proteus mirabilis and Escherichia coli are most commonly present.

XGN can lead to formation of pyelocutaneous and ureterocutaneous fistulae if not detected on time. Its prognosis is good and death is rare.

Its treatment is surgical removal of the affected kidney (i.e) nephrectomy or sometimes partial nephrectomy.

DIFFERENTIAL DIGNOSIS

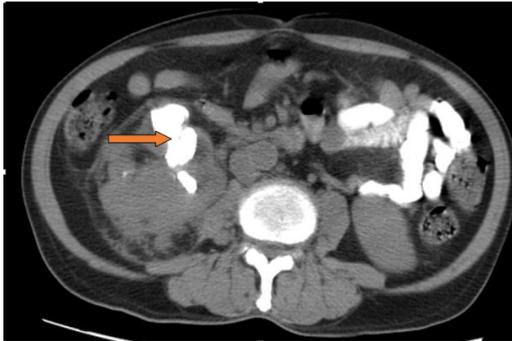
- Renal cell carcinoma (RCC)
- Renal tuberculosis
- Renal abscess
- Angiomyolipoma (AML): with minimal fat

CONCLUSION

Xanthogranulomatous Pyelonephritis is an uncommon condition presenting with non specific signs and constitutional symptoms.

The most typical features on CT are unilateral renal enlargement and extrarenal extensions of inflammatory changes presenting as perinephric fat stranding, peri renal collections or abscess.

Their presence in a patient with constitutional symptoms and urinary tract infection should alert the radiologist of this uncommon condition.



Axial plain image demonstrating asymmetric enlargement of right kidney, due to staghorn calculus(Orange arrow) in the right renal pelvis causing mild to moderate right hydronephrosis with renal parenchymal thinning.



Coronal plain image demonstrating ,multiple variable sized non obstructive calculi(yellow arrows) in the right upper , mid and lower pole calyces.

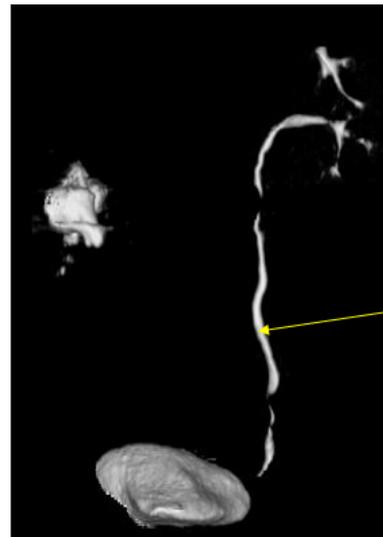


Axial post contrast image demonstrating abnormal hypodense peripherally enhancing collections (Blue arrows) within the right renal

parenchyma and right peri-nephric fat and thickening of the right anterior and posterior renal fascia (red arrows).



Axial 2 hour post contrast delayed image demonstrating, no excretion of contrast from the right renal pelvi-calyceal system even on 2hrs delayed scan. However left ureter(Red arrow) shows normal excretion of contrast.



2 hour post contrast 3D reconstruction of the image showing similar features as discussed above. Left ureter(yellow arrow)

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