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Case Report

Xanthogranulomatous Pyelonephritis- A Series of two Cases and a Review of the Literature

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Abstract

Xanthogranulomatous pyelonephritis (XGP) is a rare and severe manifestation of chronic kidney inflammation that can be critical if not recognized and treated appropriately, often requires surgical intervention along with antibiotics. Most commonly presented in the fifth or sixth decade of life with a prior history of nephrolithiasis, obstructive uropathy, or recurrent urinary tract infections.

Here, we discuss a 45-year-old male and a 73-year-old female who came with abdominal pain and weight loss for two months. An enlarged and distorted renal outline, with altered echotexture and calculus in renal pelvis was revealed on ultrasound examination. Abdominal computed tomography revealed staghorn renal calculi and thinning of renal cortex with involvement of adjacent structures.

Key words: pyelonephritis; ct scan;calculus

INTRODUCTION

Xanthogranulomatous pyelonephritis (XGP), first described by Schlagenhaufer in 1916, [1] is a rare, severe, chronic inflammatory disorder of the kidney characterized by a malignant mass that invades the renal parenchyma. XGP is mostly associated with Escherichia coli or Proteus infection.[2]

Characteristic laboratory findings include anaemia, high CRP and liver dysfunction. As for imaging investigation, computed tomography (CT) and magnetic resonance imaging (MRI) both show imaging findings of XGP and the extension of the disease.

CASE PRESENTATION

Case one:

A 45-year-old male was admitted to our hospital with vague right lumbar region pain and mild fever for the past two months. The patient mentioned that he had nearly 10 kg of unintentional weight loss during these last two months. There was no history of changes in urine frequency, colour change, burning micturition and hematuria. Physical examination of the patient revealed right-lumbar region tenderness with an ill-defined mass extending to the right iliac region.

Case two:

A 73-year-old female came to our hospital with left lumbar region pain, burning micturition and weight loss for the past four months. The patient

mentioned that she had lost nearly 16 kg of unintentional weight during these last four months. Physical examination revealed low-grade fever and left- sided costovertebral angle tenderness with an ill-defined mass extending to the lower coastal margin. There is no history of changes in urine frequency,colour change and hematuria.

INVESTIGATIONS

Both the patient had increased white blood cell counts in blood with neutrophil predominance. Urine analysis was indicative of urinary tract infection in both patients.

Case one

Abdominal ultrasound examination of the first patient showed a smaller right kidney with loss of normal cortico-medullary differentiation. An echogenic structure was seen in the renal pelvis, causing distal acoustic shadowing to represent a calculus (Which was also confirmed on the radiograph). A small ill-defined collection was observed in the right perinephric region. The presumptive diagnosis of obstructive uropathy was made, and the patient underwent a percutaneous nephrostomy to relieve the obstruction. However, the 24-hour urine output was almost negligible, and the patient did not improve clinically; a contrast-enhanced computed tomography (CECT) of the kidney-ureter-bladder (KUB) region was planned. The CECT demonstrated the whole affected kidney to be smaller with the proliferation of fatty tissue. A 23 mm staghorn calculus was noted in the renal pelvis with extensive perinephric fat stranding. [Figure 1A & 1B] There were multiple

low-density areas throughout the kidney, suggestive of necrosis or abscess. Small peri-nephric collection and retroperitoneal lymphadenopathy were also observed. [Figure 1B & 1C] The diagnosis of XGP was suggested. An ultrasound-guided renal fine needle aspiration cytology (FNAC) examination was performed, which confirmed chronic granulomatous inflammatory changes in the renal parenchyma. [Figure1D]

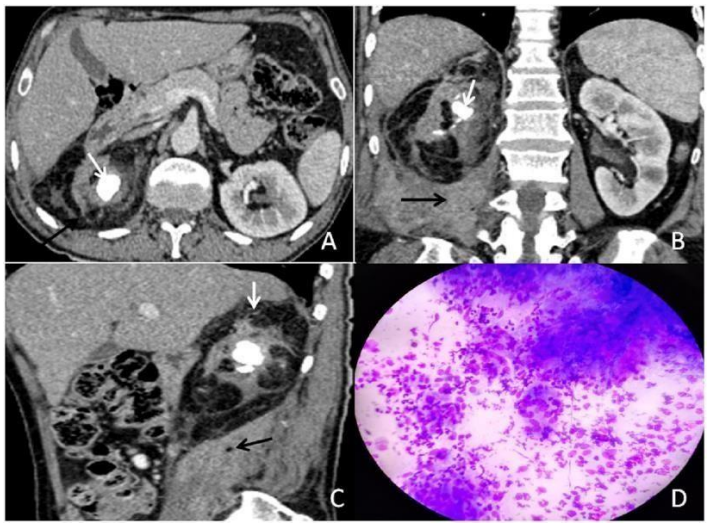

Figure 1: Contrast-enhanced CT scan of the KUB region, delayed venous phase (A) Axial image shows right kidney is replaced by fibro-fatty tissue with small residual renal parenchyma and renal pelvic calculus (white arrow). Extensive fat stranding and peri-renal collection are also evident in the perinephric region (black arrow). (B) Coronal reformatted image shows right renal pelvic calculus (white arrow) with grossly deformed renal contour and peri-nephric inflammatory changes (black arrow). (C) Sagittal reformatted image showing a small right kidney with multiple areas of low density suggestive of necrosis or abscess (white arrow). The peri-nephric collection is extending along with the right psoas muscle (black arrow), and, (D) Fine needle aspiration microscopic image (x40 stain: H&E) shows multiple scatters of epithelioid histiocytes plump to spindle shape cells, giant cells and extensive area of macrophages, neutrophils in the background of necrosis forming a granulomatous abscess.

Case two

The second patient underwent whole abdomen ultrasound that revealed an enlarged and distorted left renal outline, with loss of the typical left renal architecture and a centrally-located shadowing calculus.

A CECT scan of the KUB region revealed an enlarged left kidney with extensive inflammatory changes. Staghorn pelvicalyceal calculus measuring approx 46 mm with parenchymal thinning was noted. The ill-defined collection was noted in the anterior para nephric space, involving the pancreas and extending into the intraperitoneal compartment to involve the jejunal loops. Perinephric fat stranding and retroperitoneal lymphadenopathy were also observed. [Figure 2A, 2B, 2C] 3D, volume rendering technique coronal image of arterial phase CT scan shows abrupt cut off of left renal artery and non-visualization of the intrarenal capillary network. [Figure 2D]. The final radiological diagnosis of XGP was made. The patient underwent a total right nephrectomy, and a final histo-pathological examination confirmed our diagnosis of XGP. [Figure 3A & 3B]

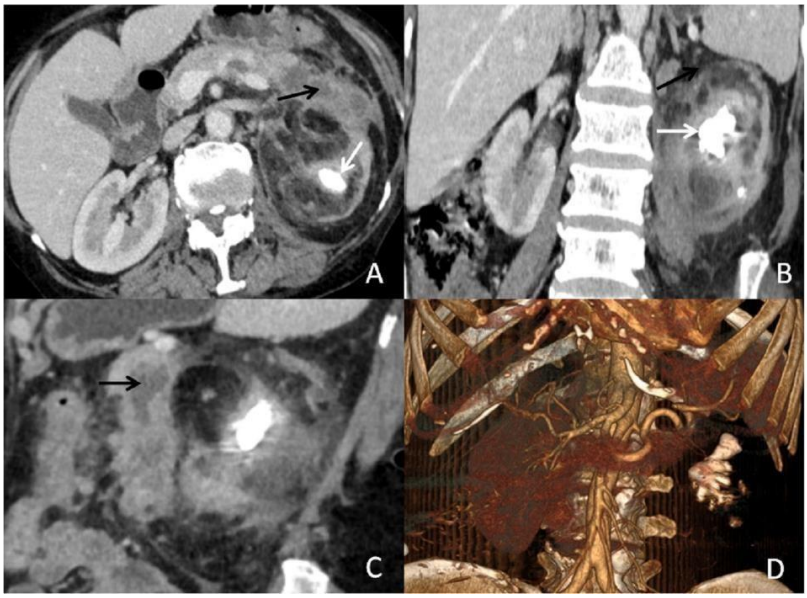

Figure 2- Contrast-enhanced abdomen and pelvis CT scan, delayed phase: (A) Axial image demonstrate a staghorn calculus in the left renal pelvis (white arrow) and peripherally enhancing hypodense collection in the perinephric region extending to the pancreatic tail (black arrow) and splenic flexure of colon. (B) Coronal reformatted image demonstrates enlarged left kidney with calculus in the renal pelvis (white arrow) and perinephric fat standing (black arrow). (C) Sagittal reformatted images showing multiple peripherally enhancing collection pockets suggestive of necrosis or abscess (black arrow), and (D) Arterial phase, 3D Volume rendering technique image, coronal plane shows non-visualisation of the intrarenal capillary network and abrupt cut off of the left renal artery at hilum along with a staghorn calculus.

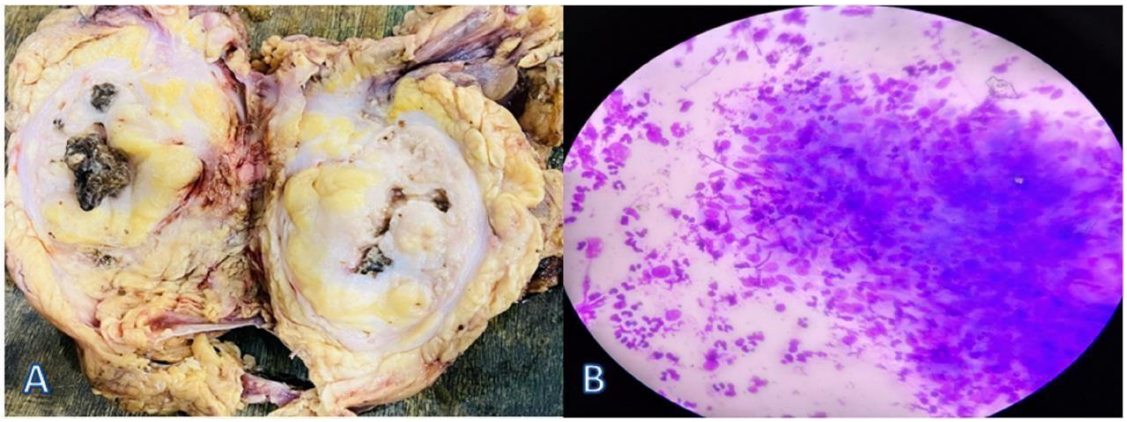

Figure 3: (A) Photograph of the resected gross specimen reveals extensive destruction and fatty replacement of renal parenchyma due to long-standing suppurative inflammation with staghorn calculus. (B) Microscopic image (x40 stain: H&E) fine-needle aspiration shows clusters and scatters of epitheloid histiocytes; few show reactive changes and prominent nucleoli in the background of necrosis, giant cells and extensive area of acute inflammatory cell-like neutrophils, tingible body macrophages forming a granulomatous abscess.

DIFFERENTIAL DIAGNOSIS

Xanthogranulomatous pyelonephritis (XGP) closely differentiated from renal cell carcinoma radiographically and clinically.[3]

TREATMENT

In acute infection higher antibiotics may be given, but if not cure with antibiotics then the treatment of choice is nephrectomy include with the removal of all the compromised tissue.

OUTCOME AND FOLLOW-UP

The first patient was kept on higher antibiotics and doing fine till now. She will be taken for surgery after the optimization of her co-morbidities. However, the second patient is recovered after the nephrectomy.

DISCUSSION

XGP is a distinctive form of pyelonephritis frequently occurring in repeated infections, chronic obstruction, and inflammation.Immuno compromisd conditions like diabetes mellitus, abnormal lipd metabolism are also considered as a risk factor in XGP. Clinical presentation is nonspecific XGP may present with fever, weight loss, and lower urinary tract symptoms being most common.[4] Though the precise pathophysiology is not yet demonstrated, it is thought that chronic obstruction and inflammation provoke the proliferation of lipid-laden macrophages, which leads to suppuration and renal parenchymal destruction. This theory is supported by observing that urinary tract calculi are present in 70–79% of patients with XGP.[1] XGP is the chronic inflammation of the renal parenchyma and is rarely encountered in clinical medicine. It is demonstrated as tubulointerstitial damage with chronic interstitial inflammation. Ultrasound and CT scans are both sensitive diagnostic tools. CT scan revealed the involvement of adjacent structures.Although ‘bear paw sign” is not present in our case but it has cystic changes with lipid laden macrophages seen in renal pelvis and calyces. Squamous ell carcinoma of the kidney, Wilms tumor and renal cell carcinoma also mimic XGP so a definitive diagnosis must be made histologically. [6,7] XGP causes renal parenchymal destruction which leads to non functional kidney, for which nephrectomy is the definitive treatment.[8]

Though XGP is mostly limited to the affected kidneys, it occasionally spreads to an adjacent structure. Malek and Elder classified the XGP into the following stages.[9] Stage I (Nephric). Spread is limited to the renal parenchyma. Stage II (Nephric and Perinephric). Disease extent both the parenchyma and the perinephric fat. Stage III. The disease is extending to the adjacent structure or retroperitoneum. In our cases, stage III, disease extent into adjacent structures like the pancreas, etc. These are primarily due to chronic inflammation leading to adherence and subsequent perforation of renal tissue to adjacent structures.

LEARNING POINTS/TAKE HOME MESSAGES

- Long standing cases of staghorn calculus with secondary infection is commonly seen associated with Xanthogranulomatous pyelonephritis.
- Bear paw sign is seen in early cases of Xanthogranulomatous pyelonephritis.
- Nephrectomy is the only treatment of choice.

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