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Xanthogranulomatous Pyelonephritis- Role of Clinical, Imaging, Microbiological and Pathological Correlation in Making Diagnosis.

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Abstract: *Xanthogranulomatous pyelonephritis (XGP) is a rare histological subset of pyelonephritis characterized by being a chronic destructive granulomatous inflammation of the renal parenchyma with presence of lipid laden macrophages (xanthoma /foam cells) on histology. We report two cases of this condition, both having Type II diabetes mellitus and urinary infections where preoperative diagnosis was suggested with the help of combination of clinical, microbiogical, pathological and imaging findings with subsequent confirmation on postnephrectomy histology, highlighting the importance of multipronged diagnostic strategy in such rare pathologies, resulting in prompt treatment and improved prognosis.*
Keywords: *XGP, CT, Microbiology, Histology*

I. Introduction

Xanthogranulomatous pyelonephritis (XGP), is a rare chronic renal inflammatory disease characterized by destruction and replacement of renal parenchyma with lipid-laden macrophages, which are also known as xanthoma (Greek for yellow) or foam cells due to their yellow pathological appearance [1,2]. XGP is mostly seen in middle-aged female patients and occurs in less than 1% of chronic pyelonephritis cases [3]. Urolithiasis and diabetes mellitus have been found to be associated with this condition. Due to rarity of the condition and varied imaging features preoperative diagnosis rate is low which can lead to delayed treatment and consequent increased morbidity and mortality [4,5]. We wish to share our experience in two different cases, both having urinary infection and diabetes mellitus where diagnosis was suggested preoperatively with the help of combination of clinical, microbiogical, pathological and imaging findings with subsequent confirmation on postnephrectomy histology. These cases highlight the importance of multipronged diagnostic strategy in such rare pathologies resulting in prompt treatment and improved prognosis.

II. Case report

Case 1

A 35 year old female with medical history of type II diabetes mellitus and previous cholecystectomy, presented with complaints of fever with chills on and off and right flank pain for around four weeks and recurrent urinary infections. On examiantion patient was febrile with tenderness and fullness of right flank. Her blood investigations revealed leucocytosis (13000/cumm) with neutrophilia. Her blood sugar and renal function tests were within normal limits with urine examination revealing pus cells with mutlidrug resistant *Proteus mirabilis* isolated on urine culture. Ultrasound of the abdomen revealed enlarged right kidney with gross hydronephrosis showing fine echoes and multiple calculi within. Her left kidney and rest of the abdominal viscera was found to be normal. Contrast enhanced CT examination was done for further evaluation which apart from confirming ultrasound findings revealed faintly enhancing right renal parenchyma with non opacification of pelvicalyceal system with inflammation of the perinephric space.(Fig. 1) In view of clinical and imaging findings suggestive of xanthogranulomatous pyelonephritis, right sided nephrectomy was performed. Grossly specimen showed completely distorted parenchyma with cystically dilated pelvicalyceal system with thickened walls (Fig.2) and histology revealed sheets of foamy histiocytes and lymphoplasmocytic infiltrates in interstitium with tubular atrophy and congestion confirming diagnosis of xanthogranulomatous pyelonephritis.(Fig.3a&b)

Case 2

A 48 years old male patient with medical history of type II diabetes mellitus and smoking, presented with complaints of left flank pain, fever and burning micturition.The patient had an elevated white blood cell

count (WBC) of 18 000/cumm and Serum creatinine of 0.9 mg/dL. Urine analysis revealed pyuria and bacteuria. Urine culture grew cephalosporins sensitive *Escherichia coli*. Plain radiography of abdomen was suggestive of multiple left renal calculi while ultrasound was suggestive of chronic pyelonephritis with nephrolithisis. CT examination with intravenous and oral contrast revealed dilated left pelviclyceal system with atrophic non contrast excreting renal parenchyma with proliferation of perinephric fibrofatty tissue and presence of hypodense collection within the left psoas muscle. Presence of gas was noted within the collecting system due to previous image guided aspiration of pelvicalyceal contents. Staghorn calculus was noted in lower calyx with few small calculi in other portions of collecting system. (Fig.4a &b) Subsequently, the patient underwent placement of a percutaneous nephrostomy and purulent fluid was drained which grew similar organism to the one that grew in the urine. Nephrostogram was performed and showed extravasations into the psoas muscle. In view of no significant urine output from the nephrostomy and significant morbidity, left nephrectomy was performed after fifteen days. Gross specimen revealed shrunken scarred kidney with dilated collecting system and sheets of foamy histiocytes with tubular degeneration on histopathology suggestive of XGP. (Fig. 5)

III. Discussion

Xanthogranulomatous pyelonephritis was first described by Schalegenhaufer in 1916 [6]. The worldwide incidence of XGP varies from 0.6% to 1% of all cases of pyelonephritis [7]. Incidence is more common in females usually during the fifth and sixth decades of life. Symptoms are flank or abdominal pain along with fever, gross hematuria and weight loss. The pathogenesis of XGP is thought to be chronic renal obstruction and infection with failure of local host immunity. Recurrent or chronic urinary tract infections, immuno-compromised patients including those with diabetes mellitus and abnormal lipid metabolism have been documented as risk factors [4,8].

XGP has been classified in to focal and diffuse forms [2,8]. The diffuse form is much more common (85%) than the focal form [9]. The diffuse form is divided into three stages according to the extent of perinephric tissue involvement [10,11].

- Stage I (Nephric): Disease is limited to the kidney.
- Stage II (Nephric and Perinephric): Disease extends to the renal pelvis or the peri-renal fat within Gerota fascia.
- Stage III (Nephric and Perinephric): Disease involves adjacent organs or retroperitoneum.

Our first case corresponds to stage II, while the second case is Stage III of the diffuse disease.

The classical imaging findings in diffuse disease described on CT imaging (which is the diagnostic modality of choice), are atrophic or nonfunctioning kidney and perirenal fibrofatty proliferation with centrally obstructing calculus and dilated calices (bear paw sign) [4,8].

Magnetic resonance imaging shows thinned renal parenchyma with lost cortico-medullary differentiation with dilated collecting system showing intermediate to high signal intensity on both T1 & T2WI (due to fat content in macrophages) and renal calculi seen as negative defects within dilated collecting system on T2WI [8,12].

Intravenous urography and DTPA renal scan usually show the non-functioning or poorly functioning affected kidney [7]. The laboratory findings in XGP include leukocytosis, anemia, microscopic hematuria, with proteinuria/ pyuria with isolation of specific bacterial species on urine culture, deranged liver function tests and elevated ESR [4,8,13]. There may be elevation of serum creatinine levels [4].

Escherichia coli, Proteus mirabilis, Staphylococcus aureus, Klebsiella species, Pseudomonas species and Enterobacter species are commonly cultured from the urine or the pathological specimen suggesting their role in pathogenesis [8].

The definitive diagnosis of this condition depends on histological examination of the operative specimen [4,8,11].Management of XGP depends on the extent of disease. In diffuse and advanced stage XGP, nephrectomy is the ultimate treatment option, while focal or segmental XGP if diagnosed early pre-operatively can be treated with antibiotics.4,8 Pre- and post-operative broad spectrum antibiotics and symptomatic management are important in the successful management and better prognosis [4].

In our first case the CT imaging findings clinched the diagnosis in a diabetic female with history of recurrent urinary infections and morbidity along with accurate preoperative disease staging. Post nephrectomy patient had relief from the symptoms with uneventful recovery.

Second case represents more extensive and advanced disease with rare involvement of psoas muscle which led to addition of tuberculous infection to the differential diagnosis. However the microbiological isolation of E. coli both in the urine culture and the culture of the aspirate from the collecting system in setting of appropriate clinical and imaging findings helped in making the preoperative diagnosis of XGP likely with post surgical histological confirmation. Patient was kept on antiobiotics post operatively with successful recovery.

Figures & legends

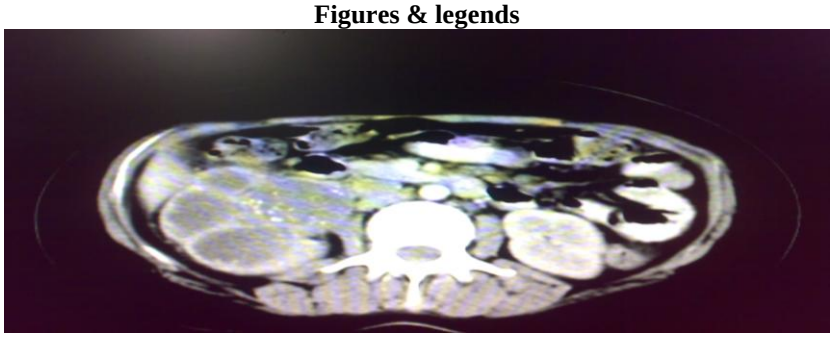

Fig. 1: CECT abdomen axial image showing faintly enhancing right renal parenchyma with dialed non opacified pelvicalyceal system (Bear paw sign) with multiple tiny calculi within along with inflammation of the perinephric space

Fig.2: Gross specimen: Completely distorted renal parenchyma. Cystically dilated pelvicalyceal system with thickened wall

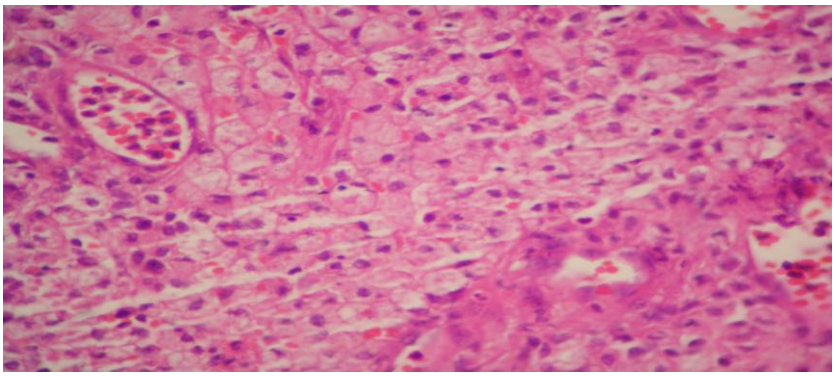

Fig.3a: High power view shows sheets of foamy histiocytes in interstitium (left upper) and degenerating tubules (right lower)

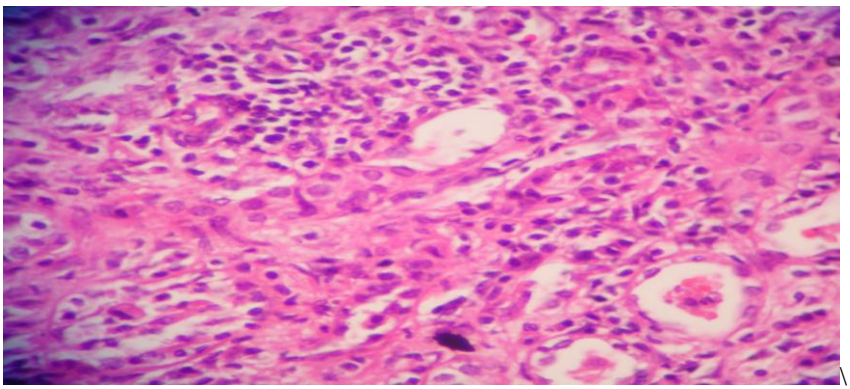

Fig.3b: High power view shows lymphoplasmocytic infiltrates in interstitium with tubular atrophy and congestion.(H&E 450X)

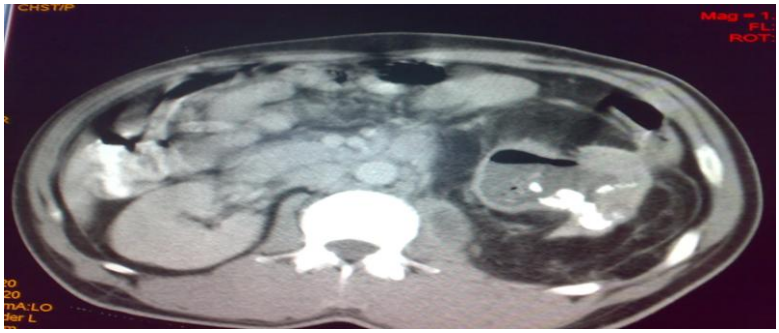

Fig. 4a: Axial CECT abdomen image shows atrophic left renal parenchyma with dilated collecting system with nephrolithiasis and perinephric fibrofatty proliferation. Iatrogenic air fluid level noted in the left renal pelvis.

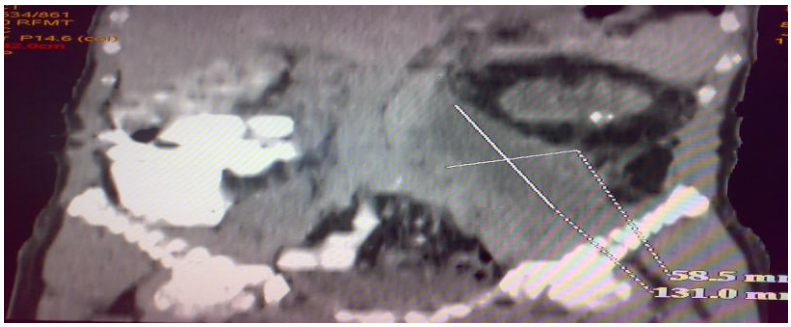

Fig. 4b: Coronal reformatted CECT image shows hypodense collection in left psoas muscle.

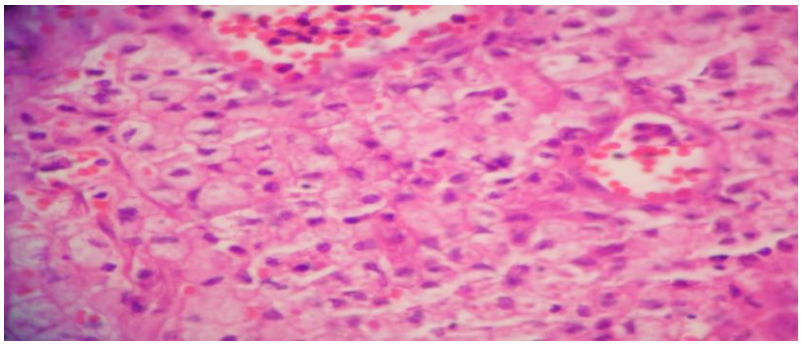

Fig. 5: High power microscopy shows sheets of foamy histiocytes with tubular degeneration. (H& E 450X)

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