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Clinicopathological spectrum of xanthogranulomatous pyelonephritis; a single-center experience over 7 years

Manish Jain[1], Faiz Ahmed Khan[2], Shailendra Patel[3], Ravinder Pal[4], Abhishek Shukla[5]

[1]Professor and Head, [2]Associate Professor, [3,4,5]Senior Resident, Department of Urology and Renal Transplant, Sri Aurobindo Institute of Medical Sciences, Indore, Madhya Pradesh, India

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ABSTRACT

Background: Xanthogranulomatous pyelonephritis (XGP) is chronic pyelonephritis subtype where in renal parenchymal destruction occurs and therefore results in progressive loss of kidney functions. Although middle age group is the predominant age group affected but it can be seen at any age. There is accumulation of macrophages (lipid-laden) leading to renal parenchymal destruction and fibrosis. In this study, we present our data of 15 patients who had undergone nephrectomy and were biopsy proven XGP. **Aims and Objectives:** The aim of the study was to describe the clinical and radiological features of XGP in adults. **Materials and Methods:** XGP constituted 4.31% of the 348 nephrectomies done for infective causes over a period of 7 years. All our patients had undergone unilateral total nephrectomy. Demographic and clinical records were analyzed after consent from all the patients. **Results:** The age range in our study was 18–65 years with mean 42.93 ± 15.66 years. Nine of our patients, that is, 60% were females. Diabetes was present in 53.3% of our patients. Three patients had imaging suggestive of pyonephrosis, three patients had perinephric collection and 9 patients (60%) had concomitant nephrolithiasis. All the kidneys were grossly enlarged and were non-functional on renal scintigraphy. **Conclusion:** XGP is a form of chronic pyelonephritis which although being less common but is devastating given the destruction of renal parenchyma it does and associated morbidity. Clinicoradiologic correlation cannot be overemphasized. The definitive diagnosis is established after histopathologic examination.

Key words: Chronic pyelonephritis; Pyelonephritis; Pyonephrosis; XGP

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INTRODUCTION

The clinical spectrum of xanthogranulomatous pyelonephritis (XGP) is varied as is its etiology. XGP was first described by Schlagenhaufer more than a century ago.[1] Due to its varied presentation, the differential diagnosis for it is also wide. It is a chronic pyelonephritis subtype where in renal parenchymal destruction occurs and therefore results in progressive loss of kidney functions. Although middle age group is the predominant age group affected but it can be seen at any age. XGP is usually unilateral; however, bilateral conditions have also been reported.[2-4] The most often reason for XGP is obstructive pathology which most commonly is nephrolithiasis and in children can be secondary to congenital abnormalities of kidney and urinary tract.[5,6] The condition is usually seen in females and people who have chronic conditions such as diabetes and autoimmune diseases or are on immunosuppression.[7-9] There is accumulation of macrophages (lipid-laden) leading to renal parenchymal destruction and fibrosis. The process of inflammation and damage thereby can extend beyond kidney and involve surrounding structures. The kidney is usually non-functional and surgical intervention in the form of nephrectomy is the only definitive treatment.[10] In this study, we present our data of 15 patients who had undergone nephrectomy and were biopsy proven XGP.

Address for Correspondence:
Dr. Abhishek Shukla, Senior Resident, Department of Urology and Renal Transplant, Sri Aurobindo Institute of Medical Sciences, Indore - 453 555, Madhya Pradesh, India. **Mobile:** +919917909417. **E-mail:** shukla.abhii.13@gmail.com

Aims and objectives

The aim of the study was to describe the clinical and radiological features of XGP in adults.

MATERIALS AND METHODS

We present our study of 15 cases of XGP, diagnosed on the basis of histopathology findings. The study was pre-approved by the Institutional Ethics Committee for the final permission. They constituted 4.31% of the 348 nephrectomies done for infective causes over a period of 7 years. All our patients had undergone unilateral total nephrectomy. Demographic and clinical records were analyzed after consent from all the patients. Specimens were fixed in 10% formalin and grossed. Paraffin embedded sections were cut into 5-μm slices followed by staining with hematoxylin and eosin. Sections were studied under light microscope both in low and high magnification.

RESULTS

The age range in our study was 18–65 years with mean 42.93±15.66 years. Nine of our patients, that is, 60% were females. Diabetes was present in 40% of our patients. Tables 1 and 2 show the clinical and biochemical parameters of patient. None of our patient had bilateral disease. Flank pain in all our patients and fever in 12 patients (80%) were the most common symptoms in our patients at presentation. All of our patients had undergone ultrasound examination followed by contrast-enhanced computed tomography and radio scintigraphy (Diethylene triamine penta acetic acid). Urine culture reports were positive for nine patients. Three patients had imaging suggestive of pyonephrosis, three patients had perinephric collection and 9 patients (60%) had concomitant nephrolithiasis. All the kidneys were grossly enlarged. Figure 1 showing the gross specimen of xanthogranulomatous kidney. Figures 2 and 3 show light microscopic findings on low and high power magnification, respectively.

Table 1: Clinical parameters of patients

Parameter	N
Age (Mean±SD)	42.93±15.66
Male n (%)	6 (40%)
Female n (%)	9 (60%)
Diabetes n (%)	8 (53.3%)
Hypertension n (%)	9 (60%)
Hematuria n (%)	5 (33.3%)
Fever n (%)	12 (80%)
Flank pain n (%)	15 (100%)
Abdominal lump n (%)	4 (26%)
Nephrolithiasis n (%)	9 (60%)

Table 2: Biochemical parameters of patients

Parameter	Mean±SD
Hemoglobin (g/dl)	10.44±1.33
Creatinine (mg/dl)	1.45±0.39
WBC	15600±5440
Platelet	2.16±0.93
Albumin	3.31±0.68
Urine Culture	Positive: 9 Negative: 6
Pyuria	15

DISCUSSION

XGP is a relatively rare subtype of chronic pyelonephritis with varying incidence as reported in the literature which ranges between 0.6% and 1%.[1] XGP is usually diffuse (involving most of the kidney) and focal variant is less common. All age groups may be affected but more often occurs in middle aged and elderly patients. XGP has been more commonly reported in females patients.[11] Our study too observed that XGP is more common in females as compared to males. All of our patients had unilateral presentation although it has been very rarely seen bilaterally.[2]

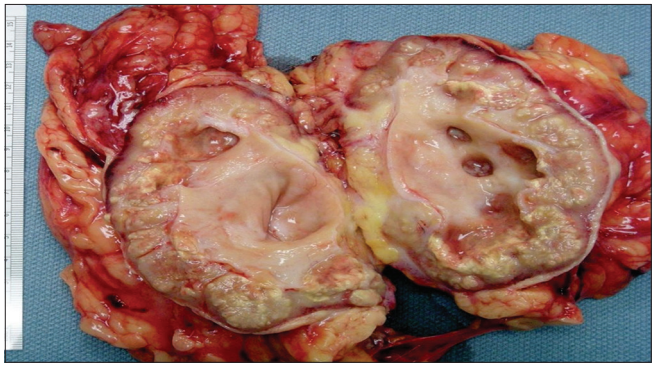

Figure 1: Gross photograph of nephrectomy specimen with dialated pelvicalyceal system, poor corticomedullary differentiation with deposition of yellowish specks limited to renal parenchyma and sparing perinephric fat in a case of xanthogranulomatous pyelonephritis

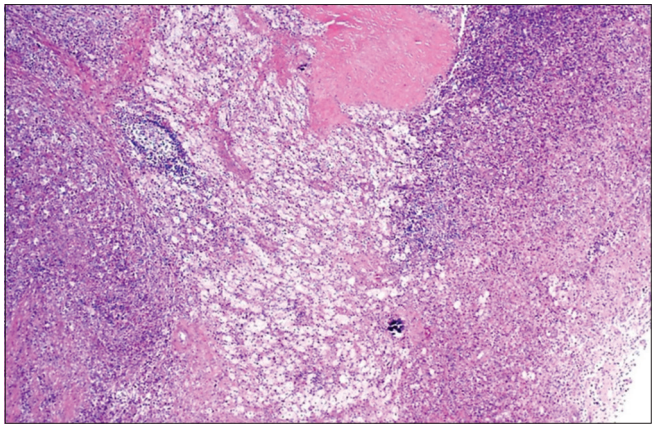

Figure 2: Photomicrograph showing renal parenchyma infiltrated with dense inflammation and foam cells on low magnification suggestive of xanthogranulomatous pyelonephritis on hematoxylin and eosin staining

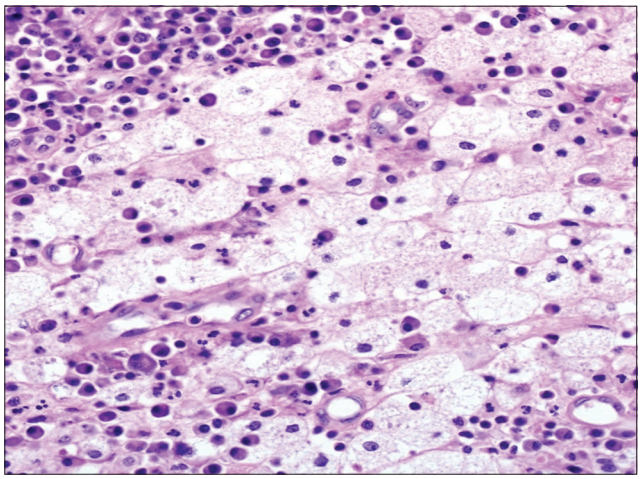

Figure 3: Photomicrograph showing foam (xanthoma) cells and lymphoplasmacytic inflammation in a case of xanthogranulomatous pyelonephritis on high magnification in hematoxylin and eosin staining

Obstruction of urinary tract and recurrent UTI is common in XGP.[1] In a study from India, nephrolithiasis was observed in 90% of patients with XGP.[12] Similarly, in our study, nephrolithiasis was observed in 9 patients (60%) diabetes mellitus, urinary stasis due to obstructive pathologies, renal neoplastic lesions, and immunocompromised states predispose patients to XGP.[13,14] In our study, diabetes was present in 8 (53.3%) patients and none of our patients was on immunosuppressive medications or had underlying malignancy. Varying clinical presentation has been observed in different studies. All of our patients had abdominal pain on presentation which is similar to most of the published series.[7,12,14] In a recent study, pyonephrosis and perinephric abscess was observed in 25.0% and 7.5% patients, respectively[12] where as in our study, 3(20%) patients had pyonephrosis and 3 patients had perinephric collection. In a study from Turkey, leukocytosis and pyuria were detected in one (7.7%) and 6 patients (46.1%) were anemic.[15] Korkes et al., analyzed 41 cases of XGP; anemia, pyuria, and leukocytosis were reported in 63%, 57.6%, and 41% of the patients, respectively.[5] In our study, 11 (73.3%) patients were anemic and leukocytosis was seen in 9 (60%) patients and all our patients had pyuria.

In the study by Kundu et al.,[12] XGP was diffuse in 31 (77.5%) cases and focal in 9 (22.5%) cases where as in our study, it diffuse disease was seen in 11 (73.3%) patients which is similar to their study. Moreover, they observed biopsy diagnosis of XGP in 23 patients (57.5%), 10 patients (25.0%) had pyonephrosis with XGP, 3 patients (7.5%) had XGP with diabetic nodular glomerulosclerosis and renal cell carcinoma with concomitant XGP in one patient. In our study, pyonephrosis with XGP was seen in in three patients and three patients had perinephric collection. Diabetic glomerulosclerosis with XGP was seen in one of the six diabetic patients in our study and in none of our patients, a neoplastic lesion was seen.

XGP has been divided into three stages according to Malek and Elder's Classification with Stage I (Nephric XGP: Confined to renal parenchyma), Stage II (Perinephric XGP involvement of perirenal space and Gerota's fascia), and Stage III (Paranephric XGP: Involvement of pararenal space and retroperitoneal structures).[16] In our study, 12 patients had Stage I disease and two patients had Stage II disease and one patient was in Stage III. On histopathology in XGP, there were lipid-laden foamy macrophages with infiltration of varying mixture of inflammatory cells along with fibrotic changes and cholesterol deposition. We too observed most of these changes in our specimens.

Limitations of the study

Patient selection was arbitrary and patients of pediatric age group not included in this study.

CONCLUSION

XGP is a form of chronic pyelonephritis which although being less common but is devastating given the destruction of renal parenchyma it does and its associated morbidity. Clinicoradiological correlation cannot be overemphasized. The definitive diagnosis is established on histopathology.

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Authors' Contributions:
MJ- Participated in research design and concept; **FK-** Participated in research design and performance of the research; **SP-** Participated in the writing of the paper; **RP-** Participated in research performance and data analysis; and **AS-** Participated in manuscript preparation and revision and in data analysis.

Work attributed to:
Department of Urology and Renal Transplant ,Sri Aurobindo Institute of Medical Sciences, Indore - 453 555,Madhya Pradesh, India.

Orcid ID:
Dr. Abhishek Shukla - https://orcid.org/0000-0003-1090-6140
Dr Manish Jain - https://orcid.org/0000-0002-8547-2116
Dr Faiz Ahmed Khan - https://orcid.org/0000-0002-1329-7646

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