Original Article

Xanthogranulomatous Pyelonephritis due to calculi: Report of 63 cases and review of literature

Haq Nawaz, Saadat Khan, Israr Hussain, Sheir Ahmed, Masha Khan, Nadeem Niazi
Department of Urology, Bolan Medical College, Quetta.

Abstract

Objective: To examine the relationship between clinical history and results of renal investigations in patients with xanthogranulomatous pyelonephritis.

Methods: A retrospective review was conducted on 63 cases presenting with histopathological diagnosis of xanthogranulomatous pyelonephritis between 1995 to 2002, at the department of Urology, Bolan Medical College and Sandeman Provincial Teaching Hospital Quetta.

Results: There were 26 males and 37 females having an average follow up of two years. Positive findings on examination and investigations at presentation were fever and flank pain in 59 (93.6%) patients and pyuria in 34 (53.9%) patients. All the patients had renal and ureteric calculi with no or severe reduction in the function of the affected kidney on DTPA scan. Hypertrophy of the contra lateral kidney was seen in 56(88.8%) patients.

Conclusion: Xanthogranulomatous pyelonephritis is a common entity in this part of the world. Late referral leads to loss of the kidney. Pre-operative diagnosis of the condition is desirable and if surgery is mandatory then all infected tissues have to be removed. (JPMA 55:387;2005).

Introduction

Xanthogranulomatous pyelonephritis is a rare, severe atypical form of chronic renal parenchymal infection usually due to obstruction and is characterized by a mass. It was first described by Schlangenhafer in 1916.[1] It is common in middle age diabetic females with a peak incidence in 6th to 7th decades, accounting for 6/1000 proved cases of chronic pyelonephritis.[2,3]

In this uncommon form of chronic renal infection and obstruction, inflammation destroys and replaces the renal parenchyma with sheets of foamy lipid containing macrophages (xanthoma cells) diffuse infiltration with plasma cells and histiocytes.[4] Nephrolithiasis has been noted in as many as 83% of patients in various studies.[5-7] These changes may involve the perinephric fat and may extend to other retroperitoneal organs.[8-10] The disease often mimics other inflammatory or neoplastic renal disorders and is frequently misdiagnosed clinically.[3]

The aim of this study was to determine the frequency of organisms causing this rare, severe atypical form of chronic renal parenchymal infection and surgical outcome.

Patients and Methods

This study was conducted at the Urology department, Sandeman Provincial teaching Hospital, Bolan Medical College, Quetta. A retrospective review of 63 patients of Xanthgranulomatous pyelonephritis presenting to our unit between July 1995 and July 2002 is reported. All the patients were assessed by a complete history and full physical examination. Investigations done before surgery were complete blood count, ESR, blood sugar, renal function tests, liver function tests, urine detailed report, urine culture and sensitivity, ultrasonography, intravenous urography and DTPA scintigraphy as part of our routine practice. Any active urinary tract infection was treated before surgery with appropriate antibiotics. The flank approach through the 12th or 11th rib was used for surgery in all the cases. After nephrectomy, fluid from the kidney was sent for culture and sensitivity and the kidney as a whole was sent for histopathological examination. The diagnosis was confirmed on the histopathological examination report of the removed kidney.

Results

There were 26 males and 37 females aged between 11 to 70 years, sixteen (25%) patients were diabetic and 19 (30%) were on anti-hypertensive drugs. There was a history of urinary tract infection in only 34 (53.9%) patients and the symptoms were of long duration (more than 2 years) in all cases. Diagnosis of xanthogranulomatous pyelonephritis was made preoperatively in 51 (80.9%) patients.

Majority of patients (93.6%) presented with fever and flank pain. Few other presenting symptoms were anorexia (76%), pyuria (53.9%), dysuria (46%), proteinuria (42.8%), weight loss (33%) and haematuria (30%).

In complete blood count, 42 (66.6%) patients had a hemoglobin less than 80 gm/l, 57 (90.4%) had polymorph leucocytosis. ESR was raised to more than 30 mm in the first hour in 48 (76%) patients.

In 19 (30%) patients, the urine was sterile but pyuria was present in 34 (53.9%) cases. Proteus mirabilis and Escherichia coli were the most common organisms isolated from the voided

urine and the fluid removed from the kidney after nephrectomy as shown in Table. In 23 (26.5%) patients, urine culture was negative despite positive cultures from the kidneys.

Table. Culture reports of urine (before surgery) and fluid from the kidney (during surgery).

Organisms isolated	No.	Percentage
Proteus mirabilis	21	33%
Escherichia coli	16	25%
Klebsiella	10	15.8%
Staphylococcus aureus	7	11%
Pseudomonaus	6	9.5%
Enterobacter species	3	4.7%

Radiological investigation, Figure 1, revealed multiple renal calculi in 56 (88.8%) patients and in 17 (26.9%) patients there were ureteric stones associated with multiple renal calculi. In 56 (88.8%) patients intravenous urography and ultrasound scan of the kidney revealed hypertrophy of the contralateral non-affected kidney. In all patients DTPA (diethyl triamine penta acetic acid) renography was performed which revealed no function in the affected kidney in 61 cases and poor function (< 10% relative function) in 2 (3%) patients. The left kidney was affected in 42 (66.6%) patients.

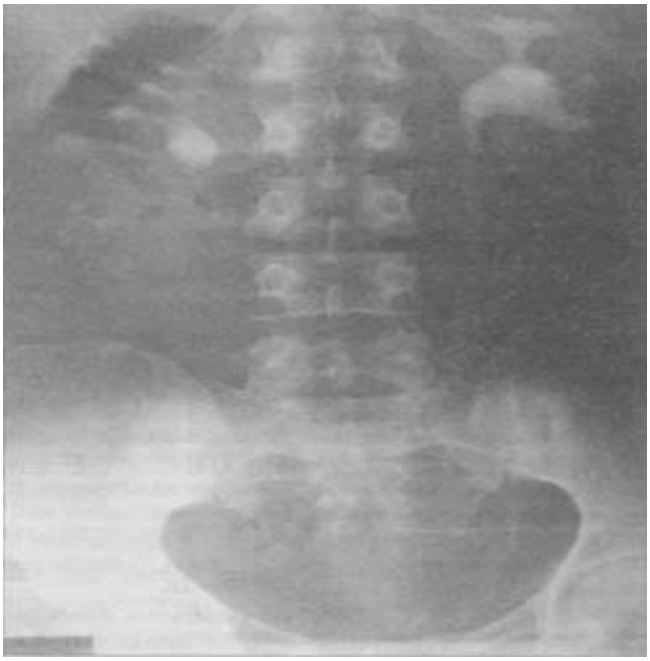

Figure 1. Intravenous Urogrogram of a 52 years old lady shows normal function/ excretion on the left side but a radio -opaque calculus and no renal function on right side. A diagnosis of Xanthogranulomatous pyelonephritis was confirmed at surgery.

On exploration, the affected kidney was enlarged in 55 (87%) patients and densely adherent to the surrounding tissues in all the patients. In 3 (4.7%) patients, the affected kidney was adherent to the descending colon, necessitating a partial colonic resection with the kidney in one patient. Perinephric abscess cavities were present in 11(17%) patients. Total nephrectomy was performed in all patients. In 6 (9.5%) patients there was clear evidence of pelviureteric junction obstruction along with calculi.

The cut surface of all removed kidneys showed diffuse cortical scarring with considerable effacement of the normal renal architecture. Areas with the distinctly yellow tinge were seen in most patients, either as scattered foci or as a discrete rim around dilated pus filled calyces or other cystic spaces (Figure 2).

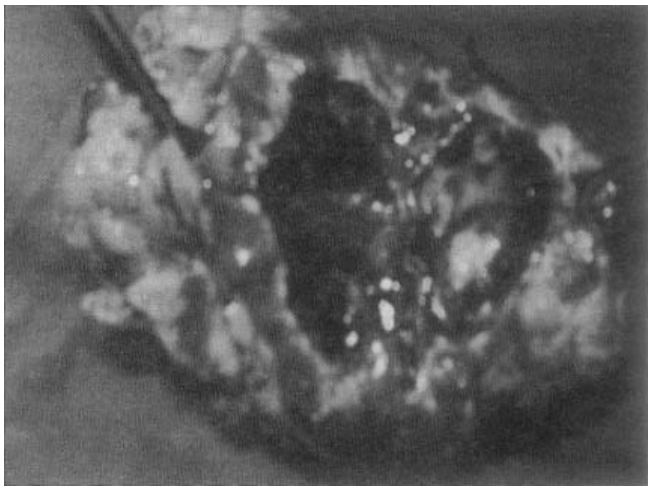

Figure 2. Gross specimen of a Xanthogranulomatous kidney showing dilated calyces, yellowish tinge of fat and a renal calculus lodged at the bottom of photograph.

Microscopically the kidneys showed a mixed acute and chronic inflammatory cell infiltrate with giant cells and lipid-laden macrophages. Such foci were usually surrounded by dense fibrous tissue and both inflammation and fibrosis clearly extended behind the renal capsule in majority of the cases.

Discussion

Xanthogranulomatous pyelonephritis is a rare and aggressive form of chronic pyelonephritis accounting for 6/1000 surgically proved cases of this pathology.[11] Women are affected more frequently than men, with a male to female ratio of 1:3.4[3,12], which is different from this study, although the number of females was higher as compared to males.

Several interrelated aetiological features are thought to be responsible for the pathogenesis of Xanthogranulomatous pyelonephritis. They include calculus or noncalculus urinary tract obstruction, ineffectively treated urosepsis, chronic renal ischemia causing localized alterations in renal metabolism, lymphatic obstruction, alterations in lipid metabolism and finally an altered immune response.[5,6,13-15] In our study, all the cases were secondary to calculi, only in a small number of cases, there was clear evidence of uretero pelvic junction obstruction with calculi.

The extent of the pathological process within the affected kidney varies. In more localized form, the lesion can be confined to one or the other pole. Most commonly, a diffuse process involving the whole kidney leading to a

decrease in renal function[16,17], which is consistent with our findings. The inflammatory process with extension into the perirenal tissues and adjacent organs causes great variety of clinical presentation of Xanthogranulomatous pyelonephritis leading to a misdiagnosis.[18]

Malek and Elder[8] proposed the extent of Xanthogranulomatous pyelonephritis inflammatory process into three stages. In the first or nephric stage, the lesion is confined to the kidney, in perinephric or stage two, the Gerota's fat is affected in addition to the kidney. In paranephric or stage three the process affects the kidney and its surrounding fat with wide spread retroperitoneal involvement. According to this staging, 57 (90%) of our patients had stage three disease. Majority of our patients were anemic, chronically ill and had painful tender flank mass.

Many reports of this disease concluded[18-22] that preoperative diagnosis is not easy and this viewpoint seem to have been generally accepted. However, we think that because of the increasing awareness of the condition, this is a diagnosis that can and should be made preoperatively. In this study, a large number of patients were diagnosed preoperatively as the Xanthogranulomatous pyelonephritis is more common in this part of world due to lack of health facilities, late referral to tertiary care centers and poor nutrtional status. The characteristics of the condition that might help in a pre operative diagnosis include usually a unilateral involvement, renal function is absent or grossly impaired on the affected side, presence of renal calculi, anemia, raised ESR and leucocytosis. There is also compensatory hypertrophy on the opposite side.[20,23] Our study patients also, had hypertrophy of the contralateral kidney suggesting chronic poor function of the diseased kidney.

Reports suggested that there is no predilection for any particular side.[11,24] On the contrary, our study showed a bias towards left side involvement. In all the cases of xanthogranulomatous pyelonephritis, there was a typical mass of yellow tissue with regional necrosis and hemorrhage. The microscopic features were lipid laden "foamy" macrophages accompanied by both chronic and acute phase inflammatory cells. Focal abscess were also observed in some cases.[4] Urine culture before surgery and culture of fluid from the kidney like other studies[4,16,25] mainly showed E. coli and proteus mirabilis growth.

Treatment is always surgical excision of the diseased tissue (total or partial nephrectomy) depending on the extent of involvement. In rare cases, where the localized form of the disease is found involving only a single pole of the kidney, partial nephrectomy[11,13,26] might be carried out. In our study, total nephrectomy along with entire inflammatory mass was done in all the cases, as all were in paranephric or stage three process.

To conclude, it is generally accepted that once the diseased kidney has been removed, the prognosis is excellent and the aim should be to remove all the infected tissues.

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