Original Article

Xanthogranulomatous Pyelonephritis

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Abstract

Introduction: Xanthogranulomatous pyelonephritis is a chronic destructive granulomatous inflammation of the kidney. This pathology was firstly described in 1916 and a small number of patient series were reported in the literature. In this study, we aimed to report the patients with xanthogranulomatous pyelonephritis in our nephrectomy cases.

Methods: The patients who underwent nephrectomy and were diagnosed with xanthogranulomatous pyelonephritis in our hospital database were reviewed retrospectively. Preoperative laboratory results, radiological imaging findings, patients' age and history of predisposing diseases were recorded.

Results: A total of 13 cases of xanthogranulomatous pyelonephritis treated in our hospital from January 2003 to December 2015 were included in the study, consisting of 7 male and 6 female patients with a mean age of 56 ±_16.09 years. In urine analyse, pyuria was positive in 6 patients (46.15%) and leukocytosis was detected in one patient (7.7%). The disease site was the right kidney in 4 patients (30.7%) and the left kidney in 9 patients (69.23%). Radiological findings of the patients were pyonephrosis, renal ectasia, pyelonephritis, hydronephrosis, renal tumor, xanthogranulomatous pyelonephritis and non-functioning kidney with renal calculi. All patients were treated by open surgical techniques and perioperative and postoperative complications did not occur. Partial and radical nephrectomy was performed in one patient and the other patients were treated with simple nephrectomy.

Conclusion: Xanthogranulomatous pyelonephritis is an uncommon histologic variant of the kidney for patients who are surgically treated for pyelonephritis. Early diagnosis and treatment is very important for decreasing morbidity and mortality. Although radical surgery is the main treatment of choice for patients with diffuse xanthogranulomatous pyelonephritis, nephron sparing surgery is an alternative for patients who have the focal form, if technically possible.

Keywords: Nephrectomy, staghorn, xanthogranulomatous pyelonephritis

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Introduction

Xanthogranulomatous pyelonephritis (XGP) is an uncommon chronic inflammatory process of the kidney resulting in renal destruction.[1] This pathology is characterized by destruction and replacement of renal or peri-renal tissue with infiltration of granulomatous tissue that contains lipid-filled macrophages.[2] Since these lipid-filled macrophages appear yellow in pathological appearance, the term "*xantho*" (Greek for yellow) is used in its name.[3] XGP is usually seen in middle-aged female patients and occurs in less than 1% of chronic pyelonephritis cases.[4]

The most common symptoms of patients with XGP are flank pain and fever.[5] Other complaints include hematuria, palpable mass and voiding symptoms. XGP may be life-threatening in some cases and must be generally managed with radical surgery.[3] Additionally, conservative management with antibiotics fails to treat the underlying the pathological process. XGP can involve adjacent organs such as liver, duodenum, colon and great vessels in the severe form of the disease.

We retrospectively reviewed the cases of XGP who underwent nephrectomy in our hospital with pathologic documentation.

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Materials and Methods

All nephrectomy specimens collected from January 2003 to December 2015 in Haydarpaşa Numune Training and Research Hospital were evaluated retrospectively. A total of 13 patients were included in this study. The preoperative laboratory tests including white cell count, creatinine, alkaline phosphatase, aspartate aminotransferase, alkaline aminotransferase, urine analysis and culture, hemoglobin and hematocrit levels were recorded.

The patients' age and laboratory results were calculated as Mean ± Standard deviation using MedCalc Statistical Software demo version 14.12.0 (MedCalc Software bvba, Ostend, Belgium; www.medcalc.org; 2014).

Results

There were 13 patients in the current study. Of these patients, 7 were male (53.84%) and 6 (46.15%) were female. The patients' characteristics and laboratory results are shown in Table 1. The mean age of the patients was 56 ± 16.09 years. Respectively, 3 and 4 patients had a positive history of diabetes mellitus and hypertension. In urine analysis, pyuria was positive in 6 patients (46.15%) and urine culture was available for 10 patients. One patient had *Escherichia coli* as a bacterial agent (10%) and 90% of the patients had a sterile urine culture. Leukocytosis was detected in one patient (7.7%) and the mean level of white cell count was 7830 ±_2110*10^6/L. Azotaemia (7.7%) and elevated liver enzymes (7.7%) were detected in one patient separately.

Table 1. Patient characteristics, laboratory findings and clinical presentations.

Number of patient *n* (%)	13(100)
Male *n* (%)	7 (53.84)
Female *n* (%)	6 (46.15)
Age (years)	56 ± 16.09
White cell count (*10^6/L)	7830 ± 2110
Hemoglobin (g/dL)	11.45 ± 2.18
Anemia *n* (%)	6(46.15)
Pyuria *n* (%)	6(46.15)
Azotaemia *n* (%)	1(7.7)
Abnormal liver enzymes *n* (%)	1(7.7)
Clinical symptoms *n* (%)	
Flank pain	7(53.84)
Fever	4(30.77)
No symptom	2(15.38)

Urinary calculi were present in 7 patients (53.84%), with ureteral stone in two patients (15.38%) and staghorn calculi in one patient (7.7%). In all patients, except a 49-year-old female patient, unilateral disease (92.3%) was reported. The disease site was the right kidney in 4 patients (30.7%) and the left kidney in 9 patients (69.23%). The clinical presentations of the patients included flank pain in 7 patients, fever in 4 patients, and incidental discovery in 2 patients (Table 1).

The radiological findings of the patients were reported as pyonephrosis (28.57%), renal ectasia (57.14%) and pyelonephritis (14.28%) on ultrasonography imaging. Computed tomography findings were reported as pyelonephritis (25%), hydronephrosis (25%), renal tumor (25%) and xanthogranulomatous pyelonephritis (25%). Intravenous urography was performed in two patients and non-functioning kidney was diagnosed with renal calculi. Magnetic resonance imaging was performed in one patient and renal mass was detected. The patient was treated with open nephron sparing surgery. The other patients' (12 cases) treatment modalities included open simple nephrectomy in 11 cases and open radical nephrectomy in one patient because of tumor suspicion.

Enlarged kidneys associated with calyceal, ureteral, staghorn calculi and hydronephrosis were detected. Pathologically, replacement of renal parenchyma with foamy macrophages (xanthoma cells), neutrophils, lymphocytes, plasma cells and multinucleated giant cells were found. The corticomedullary area was replaced with yellow nodular areas. Cortical atrophy and abscess formation were other pathological findings. An immunohistochemical study was performed in one patient. The patient was positive for CD 68 and negative for vimentin, desmin, CD10 and cytokeratin.

The overall incidence of xanthogranulomatous pyelonephritis was 2.45% in our nephrectomy cases. The incidence decreased to 0.7% in renal tumor nephrectomy series and increased to 4.3% in simple nephrectomy patients.

Discussion

XGP is a rare inflammatory condition of the renal parenchyma that is a result of chronic obstruction and suppuration.[6] This renal pathology was first described by Schlagenhaufer.[7] The worldwide incidence of XGP varies from 0.6% to 1% of all cases of pyelonephritis.[8] The disease is seen in all age groups and is more common in female patients. The authors[8] reported the mean age of the patients as 51.5 years with a female-to-male ratio of 1.66. Uğraş *et al.*[4] demonstrated that the mean age of the patients was 59 years and female patients were predominant with a ratio of 1.75. In another study from Brazil,[6] the investigators revealed that the patients had 47 years of mean age and 85.4% of the patients were female. The mean age of the patients was 56 ± 16.09 years and the male patients accounted for 53.84% of the patients.

The Clinical presentations of the patients include fever, flank or abdominal pain, lower urinary tract symptoms, palpable mass, gross hematuria and weight loss.[9] One-third of the patients have complications such as psoas abscess, nephrocutaneous fistula, nephrocolonic fistula and paranephric abscess. Kim *et al.*[5] found that the most common complaints of the patients were flank pain and fever. In another study from India, fever and dysuria were the main symptoms of the patients.[8]

The laboratory findings of XGP include leukocytosis, anemia, hematuria and increased creatinine level.[4] Korkes *et al.*[7] analyzed 41 cases of XGP; anemia, pyuria and leukocytosis were reported in 63%, 57.6%, and 41% of the patients, respectively. In another study from Turkey, the authors revealed that all of the patients had anemia and leukocytosis.[4] In this study, leukocytosis and pyuria were detected in one (7.7%) and 6 patient (46.15%), respectively. Anemia was detected in six patients (46.15%)

The pathogenesis of XGP is not clear.[4] Obstruction with or without calculi and destructive infections are the main factors. The other predisposing factors are recurrent urinary tract infection, altered immunological anomalies and abnormal lipid metabolism.[8] History of diabetes mellitus and hypertension were positive in 3 (23%) and 4 patients (30.77%), respectively.

XGP was classified as focal or diffuse.[2] The diffuse form is more common (85%) than the focal form.[10] The diffuse form is divided into three groups according to the extent of involvement in the adjacent tissues.[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.

Although XGP has atypical findings on radiologic imaging, diagnosis or differential diagnosis of XGP is difficult.[10] The classic ultrasound finding is diffuse renal enlargement with a central echogenic focus representing staghorn calculus.

The main diagnostic imaging modality for XGP is computed tomography.[9] Hydronephrosis, renal calculus, pyonephrosis, intraparenchymatous collection, abscess and peri-nephritic fat accumulation are the typical appearance of diffuse form of XGP on computed tomography. The focal form of XGP could be presented as a pseudotumoral lesion.[10] Magnetic resonance imaging is another modality that is sensitive for identifying the accumulation of lipid-laden foamy macrophages as high-intensity signal on T1 weighed images.[9] Intravenous urography and DTPA renal scan usually show the non-functioning or poorly functioning kidney.[8] Preoperative diagnosis rate is very low; Kim *et al*.[5] reported that the diagnosis rate was 19% and the authors from Brazil[6] found positive diagnosis in 22% of the patients. Differential diagnosis of XGP includes renal cell carcinomas, malakoplakia, leiomyosarcoma, and megalocytic interstitial nephritis.[9] We revealed that the positive diagnosis of computed tomography was 25% in the current study.

The best treatment of XGP is surgery with partial or total nephrectomy.[4] Surgery can be performed by open and laparoscopic techniques.[2] Conservative management with antibiotics can be an alternative in some cases with small lesions.[5] The management of the patients were open simple, radical and partial nephrectomies.

We have reported the rare kidney pathology findings with preoperative findings. The small number of patients and retrospective design are the limitations of this study. XGP must be identified and treated carefully because of aggressive complications. Laboratory results may be unremarkable and clinicians should consider this pathology in treatment failure.

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