A Rare Cause of Chronic Pyelonephritis: Xanthogranulomatous Pyelonephritis

Kronik Piyelonefritin Nadir Bir Nedeni: Ksantogranulomatöz Piyelonefrit

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ABSTRACT

Xanthogranulomatous pyelonephritis (XGP) is a chronic, destructive, granulomatous inflammation of the renal parenchyma leading non-functioning kidney. Contrast-enhanced computed tomography (CT) is the most useful diagnostic method. A 6-year-old female patient admitted to us with complaints of abdominal pain, fever, fatigue, weakness. Recurrent urinary tract infection was present in her history. The patient's urine specimen revealed leukocyturia, hematuria, positive for nitrite and bacteria, white blood cell: 12.7x10^3 /microl, hemoglobin: 6.2 g/dl and C-reactive protein (CRP): 101 mg/dl. In the urinary system Ultrasound (US) imaging, the contours of the left kidney were irregular and lobulated, and medullary punctate echogenicity and calcifications were present. Contrast-enhanced abdominal CT showed increased left kidney size and opacities suggestive of stone in the collecting system; pararenal area was heterogeneous. Although the patient's gentamicin treatment was completed in 7 days and meropenem treatment in 14 days, acute phase reactants did not regress. No activity uptake was observed in the left kidney lodge in renal cortical scintigraphy. Left total nephrectomy was performed with the diagnosis of non-functioning left kidney and chronic pyelonephritis. Kidney biopsy material were reported as XGP. XGP is a rare and aggressive cause of chronic pyelonephritis with serious consequences such as nephrectomy requirement.

Key Words: Chronic pyelonephritis, Pyelonephritis, Urinary tract infections, Xanthogranulomatous pyelonephritis, XGP

ÖZ

Ksantogranülomatöz piyelonefrit (XGP), böbrek parankiminin fonksiyon kaybına yol açan kronik, yıkıcı, granülomatöz bir enflamasyonudur. Kontrastlı bilgisayarlı tomografi (BT) en yararlı tanı yöntemidir. Bu yazıda karın ağrısı, ateş, halsizlik şikayetleri ile başvuran ve XGP tanısı alan altı yaşında kız hastadan bahsedildi. Özgeçmişinde tekrarlayan idrar yolu enfeksiyonu öyküsü mevcuttu. İdrar incelemesinde lökositüri, hematüri, nitrit ve bakteri pozitifliği mevcuttu. Laboratuvar analizinde beyaz kan hücresi: 12.7x10^3 /mikrol, hemoglobin: 6.2 g/dl ve C-Reaktif Protein (CRP): 101 mg/dl saptandı. Üriner sistem ultrasonda sol böbreğin konturları düzensiz ve lobüler olup, medüller punktat ekojenite ve kalsifikasyonlar mevcuttu. Kontrastlı abdominal BT'de sol böbrek boyutunda artış ve toplayıcı sistemde taşı düşündüren opasiteler görüldü; pararenal alan heterojendi. Hastanın gentamisin tedavisi 7 güne, meropenem tedavisi 14 güne tamamlanmasına rağmen akut faz reaktanlarında gerileme olmadı. Renal kortikal sintigrafide sol böbrek lojunda aktivite tutulumu gözlenmedi. Fonksiyone olmayan sol böbrek ve kronik piyelonefrit ön tanıları ile hastaya total nefrektomi uygulandı. Böbrek biyopsi materyali XGP ile uyumluydu. XGP, nefrektomi gereksinimi gibi ciddi sonuçları olan kronik piyelonefritin nadir ve agresif bir nedenidir. Akut faz reaktanlarında beklenen düşüş sağlanamayan ve piyürisi devam eden çocuklarda ayırıcı tanıda düşünülmelidir.

Anahtar Kelimeler: Kronik piyelonefrit, Piyelonefrit, Üriner sistem enfeksiyonu, Ksantogranulomatöz Piyelonefrit, KGP

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INTRODUCTION

Xanthogranulomatous pyelonephritis (XGP) is a chronic, destructive, granulomatous inflammation of the renal parenchyma leading non-functioning kidney. The most frequently associated conditions are chronic urinary tract obstruction or ongoing infections due to kidney stones (1,2). Recurrent urinary tract infections due to Escherichia coli and Proteus mirabilis are frequently present in the history (3). The disease develops when renal or perirenal tissue is replaced by granulomatous tissue filled with lipid-laden macrophages due to a defect in macrophage-mediated bacterial degradation (4). Contrast-enhanced computed tomography (CT) is the most useful diagnostic method, demonstrating multiloculated abnormal kidney tissue consisting of dilated collecting systems surrounded by multiple hypoechoic areas with contrast enhancement. In addition, it can demonstrate the stones in the collecting system and the dissemination of the lesion in the kidney (5,6). XGP may imitate other neoplastic and inflammatory kidney diseases (7). It is a rare condition seen in 0.6% of histological examinations of chronic pyelonephritis and 19.2% of pyelonephritis resulting in nephrectomy (8). In this paper, we aimed to present our case, which is rare in childhood and treated with unilateral total nephrectomy, in the light of radiological findings and literature.

CASE

A 6-year-old, 8-month-old female patient applied to our emergency outpatient clinic with complaints of weakness that lasted for about 1 month and abdominal pain, fever, and fatigue for a week. She was born 2750 gr at 33 weeks, and she had no history of hospitalization before. It was learned from her history that she had occasional abdominal pain and that she had been treated with antibiotics several times with the diagnosis of urinary tract infection. She had no regular medication. On physical examination, weight: 17.5 kg (3-10p), height: 114 cm (10-25 p), cardiac pulse: 124/min, Blood Pressure: 108/66 mm/Hg, temperature was 38.2ºC. The skin and conjunctiva were pale and there was widespread tenderness in the abdomen. Other systemic examination was normal.

In laboratory analysis, white blood cell: $12.7x10^3$ /microl, hemoglobin: 6.2 g/dl, hematocrit 22.6%, MCV: 52 fL, RDW:21.7% (12.2-14.4), reticulocyte 2.3%, C-reactive protein (CRP): 101 mg/dl (0-8), procalcitonin 0.09 mcg/dl, iron 10 mcg/dl (50-120), total iron-binding capacity 222 mcg/dl (50-120), ferritin 16.9 mcg/L (11-307), urea: 22 mg/dl (17-47), creatinine:0.23 mg/dl (0.24-0.73), uric acid: 3.8 mg/dl, (2.5-7.2), albumin 33.1 g/L (35-55). Direct Coombs was negative, and distribution of hemoglobin electrophoresis was normal. The patient's urine specimen revealed a density of 1033, pH: 6.5, positive for nitrite and bacteria. There were 92 WBCs/hpf, 32 RBCs/hpf. Kidney functions, liver tests, serum electrolyte levels, and B12 levels were within normal limits. In the peripheral smear, neutrophil dominance (65%) was present, and no atypical cells and blasts were observed. Hypochromic microcytic erythrocytes were seen. The erythrocyte suspension was transfused to the patient. Empirical ceftriaxone treatment was initiated, and symptoms regressed within 48 hours and fever control was achieved. Urine culture was negative. At the 72nd hour of treatment, the patient was consulted with pediatric nephrology and pediatric infection, with an increase in CRP (132 mg/dl and 147 mg/dl, respectively) and ongoing leukocyturia in the control urinalysis. Her treatment was changed to meropenem and gentamicin.

In the urinary system Ultrasound (US) imaging, the long axis of the left kidney was 10 cm, the parenchyma thickness was 13 mm, and the Antero-posterior (AP) diameter was 11 mm. The contours of the left kidney were irregular and lobulated, and medullary punctate echogenicity and calcifications were present. Corticomedullary differentiation was lost. Dilatation and heterogeneous appearance were observed in the pelvicalyceal structures, and there was thickening of the calyx walls. The left pararenal area was heterogeneous and edematous. The long axis of the right kidney was 103 mm, the parenchyma thickness was 12 mm, and its parenchymal echogenicity was normal. Contrast-enhanced abdominal CT showed increased left kidney size and 7 mm diameter opacities suggestive of stone causing artifacts in the collecting system, and moderate to severe ectasia in the left kidney. The left pararenal area was heterogeneous, and the left ureter was dilated (Figure1). There was no vesicourethral reflux in the voiding cystourethrogram.

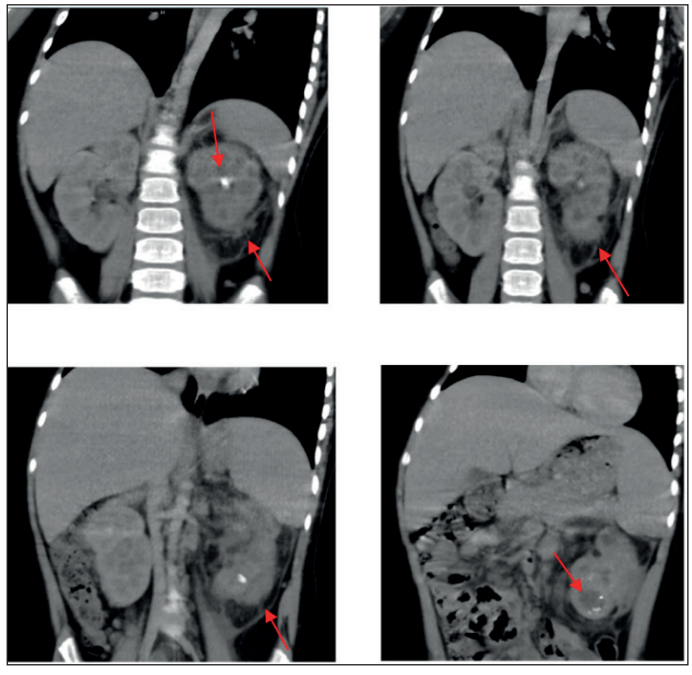

Figure 1: Contrast-enhanced abdominal computed tomography showing multiloculated appearance, ectasia, stones in the collecting system and heterogenous pararenal area of the left kidney

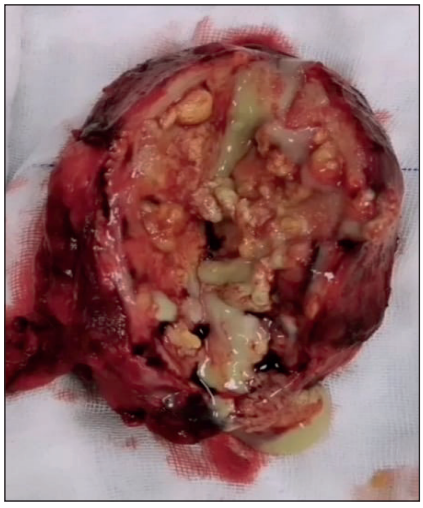

Figure 2. Nephrectomy material of non-functioning left kidney with yellow-green abscess areas in the cortex

Although the patient's gentamicin treatment was completed in 7 days and meropenem treatment in 14 days, acute phase reactants did not regress. In renal cortical scintigraphy with technetium-99m (Tc-99m) dimercaptosuccinic acid (DMSA), right kidney contours and size was normal, parenchymal uptake was homogeneous. No activity uptake was observed in the left kidney lodge. Left total nephrectomy was performed to the patient with the diagnosis of non-functioning left kidney and chronic pyelonephritis. Kidney biopsy material with yellow-green abscess areas in the cortex and the presence of pancytokeratin (-) and CD68 (+) staining were reported as XGP (Figure 2). The patient was discharged with oral cefixime on the 7th postoperative day without any complications. Acute phase reactants turned negative on the post operative 6th day. The patient is currently being followed up as an outpatient with normal renal function 4 months after surgery.

DISCUSSION

XGP is the chronic inflammation of renal parenchyma, causing non-functional kidney (1). The most common symptoms in children are fever, abdominal or flank pain and growth retardation. Urinary system symptoms such as dysuria, frequency and bloody urine may also be seen (3). Physical examination may reveal fever, pallor due to anemia, palpable unilateral or bilateral renal mass, costovertebral angle tenderness, nephrocutaneous fistulas, and rarely hepatomegaly due to liver invasion (3). Karabulut et al. (9) reported abdominal pain, weight loss and pallor; and Caixeta et al. (10) reported fever, pallor, and abdominal distension in their papers. In our patient, the initial complaints were fever, abdominal pain, weakness, and pallor, similarly. In patients with XGP, leukocytosis, anemia, increase in erythrocyte sedimentation rate, CRP, urea, and creatinine, as well as abnormalities in liver function tests due to mild biliary retention may be observed (11,12).

Urinalysis may reveal pyuria, bacteriuria, or hematuria. It has been reported that organisms such as Escherichia coli, Proteus mirabilis, Pseudomonas spp., Enterococcus faecalis and Klebsiella spp. grow in urine culture (11,12). In our case, resistant pyuria was present, and no microorganism growth was detected in the urine culture. Previously used antibiotics and the intense infection in the perinephric area were thought to be the cause of negative urine culture.

Staghorn stones can be shown radiologically on abdominal plain radiographs. CT is the most useful imaging in the diagnosis of XGP cases. With contrast-enhanced imaging, abnormal kidney tissue consisting of dilated collecting system with multiloculated appearance, multiple hypoechoic areas with contrast uptake around it, stones in the collecting system and the spread of the lesion in the kidney can be shown (5,6). Magnetic resonance imaging (MRI) can be performed in patients with contrast material allergy. Reduced kidney function can be demonstrated with Tc-99m DMSA scan. Biopsy and immunohistochemical staining (PAS positive staining) are other diagnostic methods (11,12). Karabulut et al. (9) reported parenchymal loss in the kidney, contamination in the perirenal fat planes, and nephrocalcinosis in their cases. Contrast-enhanced CT in our patient supported the diagnosis with a similar radiological appearance in the left kidney and pararenal region, and complete loss of function in the left kidney was revealed by renal parenchymal scintigraphy.

XGP is seen in 3 forms: diffuse, segmental, and focal. While segmental involvement is seen in the segmental type, there is localized cortex involvement not associated with the pelvic region in the focal type (13). The most common type is diffuse and has been divided into 3 stages by Malek and Elder. These stages are;

- Stage 1 (nephritic): Disease limited to the kidney
- Stage 2 (Peri-nephric): Disease with renal pelvis and perirenal fat involvement that has progressed into Gerato's fascia.
- Stage 3 (Paranephric): It is a disease with adjacent organ or retroperitoneal involvement (14).

Complications of XGP are psoas abscess, perinephric abscess, nephro-cutaneous fistula, intestinal fistulas, secondary amyloidosis, nephrotic syndrome, and sepsis (15). In our case, the abscess in the renal parenchyma opened to the calyces, causing sterile pyuria, and spread to the peri-nephritic and retroperitoneal areas with perforation of the renal parenchyma. Preoperative antibiotics and percutaneous drainage have a place in the treatment of cases with focal or segmental XGP. Partial or total nephrectomy can be performed in cases unresponsive to treatment (16). While Caixeta et al. (10) reported a complete recovery with antibiotic treatment in their pediatric case, nephrectomy was required in both cases of Karabulut et al. (9).

In conclusion, XGP is a rare and aggressive cause of chronic pyelonephritis with serious consequences such as nephrectomy

requirement. It should be considered in the differential diagnosis of patients who do not have the expected decrease in acute phase reactants and in children whose pyuria continues. The follow-up and treatment of these patients should be carried out in a multidisciplinary manner with the cooperation of pediatric nephrology, pediatric infectious diseases, oncology, urology/ pediatric urology, and radiology.

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