

M. Alehossein MD¹,
M. Mollaeian MD²,
F. Shahnazari MD³.

Xanthogranulomatous Pyelonephritis in a Child

Abstract: Xanthogranulomatous pyelonephritis (XGPN) is a rare chronic inflammatory disorder in children that results from infection usually associated with longstanding urinary obstruction. There are two morphologic types: diffuse and focal. In the more common diffuse form, the entire kidney is involved. We present a case of diffuse type, in which some foci in the kidney are spared.

Keywords: xanthogranulomatous pyelonephritis, kidney, childhood

Introduction

Xanthogranulomatous pyelonephritis (XGPN) is a rare but well recognized morphologic variant of chronic pyelonephritis, which commonly occurs in middle-aged women with a longstanding history of chronic pyelonephritis and urolithiasis.¹

The first published case was described by Schlangenhauer in 1916, and the term was first used by Oberling.²

It accounts for 0.6 % to 1.4 % of histologically documented cases of pyelonephritis. There is destruction and replacement of renal parenchyma by granulomatous tissue containing histiocytes and foamy lipid-laden macrophages. Of the 213 cases of pediatric XGPN reported in the literature since 1963, the histopathologic process was diffuse, involving the whole kidney in 92%, and focal in 8%.¹ With about 75% having calculi and most having some obstructing lesion.² Patients present with fever, flank pain, weight loss and a palpable mass.³ The most frequent laboratory abnormalities include leukocytosis, anemia, pyuria, hematuria and positive urine cultures. Erythrocyte sedimentation rate (ESR) is elevated in virtually 100% of cases. The most common organisms are *Proteus*, *Escherichia coli*, *Pseudomonas*, *Klebsiella*, *Aerobacter* and *Staphylococcus*.²

We present the radiologic and pathologic description of a pediatric case of the diffuse type of XGPN with parts of the kidneys was spared.

Case report

A 13-year-old boy was referred to our imaging center for ultrasonography. He had a history of chronic pain in left flank for 5 years. He had been hospitalized two years ago due to fever, pyuria and failure to thrive but without a definite diagnosis. He complained of chronic continuous left flank pain with intermittent dysuria and mild fever. The boy did not look ill. He was alert but had marked growth retardation (5th percentile). There was no apparent palpable abdominal mass or hypertension. In the review of the recent lab data, there were noted pyuria (10-12 WBC per high power field), increased sedimentation rate (80 mm/hr) and urine culture positive for *E.coli* (> 100,000 colony forming units per milliliter). Complete blood cell count (CBC) was normal.

1. Department of Radiology,
Bahrami Children Hospital, Tehran
University of Medical Sciences,
Tehran, Iran.

2. Department of Surgery, Bahrami
Children Hospital, Tehran
University of Medical Sciences,
Tehran, Iran.

3. Saadatabad Pathobiology
Laboratory, Tehran, Iran

Corresponding Author:

M. Alehossein, School of Medicine,
Tehran University of Medical
Sciences, Tehran, Iran.

Tel: +98 21 7547971-4

Fax: +98 21 7551544

Email:

mehdialehossein@yahoo.com

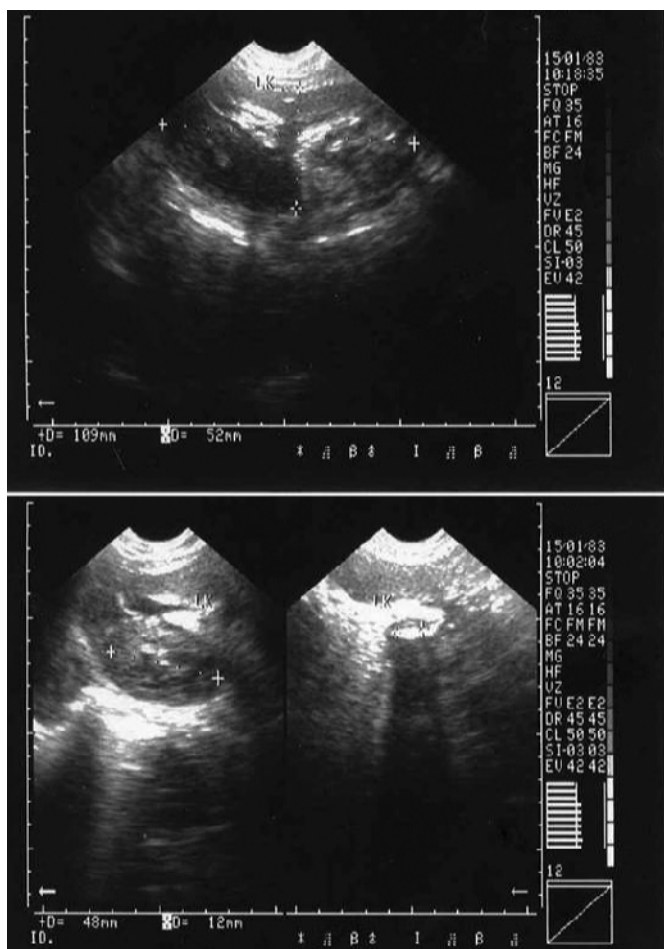


Figure 1: The left kidney is enlarged with hypoechoic mass in upper pole (top image). Lower images show staghorn calculi with posterior shadowing.

In ultrasound evaluation, the left kidney was enlarged (110x 52mm). There was nephrolithiasis in the pelvis and lower calyces compatible with staghorn calculi. However, the pelvis was contracted and without fullness. A few small hypoechoic areas were noted which were separated by apparently normal parenchyma. There was at least one hypo- to isoechoic mass, measuring 48mm, in the upper pole that stretched and mildly dilated upper infundibulum (Figure 1). Right kidney and bladder were normal.

Plain abdominal film showed an ill defined enlarged renal shadow with staghorn calculi in the pelvis and lower calyces. Voiding cystourethrography was negative. Pre-contrast computed tomography (CT) demonstrated an enlarged left kidney with two hypodense spherical masses in upper and middle portions measuring about 5 and 1.5 cm, respectively. There were a few central and lower calyceal calculi. Renal pelvis contracted around the pelvic stone (Figure 2). Post-contrast CT showed persistent low attenuation masses (or collections) without changing attenuation. Its peripheral rim showed no marked enhancement. Pelvis and lower calyces did not fill with contrast (Figure 3); however, upper calyces showed excretion of contrast media. (Figures 4, 5)



Figure 2: CT without contrast shows left pelvic calculi, contracted pelvis and perirenal fat stranding.

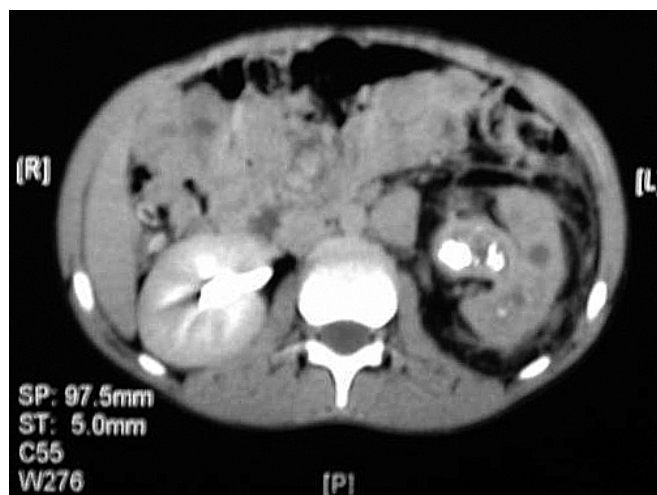


Figure 3: CT after IV contrast shows normal right renal excretion and no excretion is evident from the left kidney.



Figure 4: CT with contrast in nephrogram phase shows tiny suspending calyceal stone, low attenuation mass and normal parenchymal rim in posterior aspect of the upper pole of left kidney.

Thickening of Gerota's fascia and perinephric soft tissue strands were seen (Figures 2, 3). However, contiguous organs or spaces were spared (Malek's stage 2).^{4,5} Preoperative diagnosis was XGPN.

At operation, the left kidney was enlarged and firm with perinephric inflammation. There was no apparent fistula or other organ involvement. Nephrectomy was done.

In gross pathology (Figure 6), a deformed kidney of $11 \times 7 \times 6$ cm was observed. The renal cut surface showed yellow lobulated masses diffusely replacing the renal architecture. Areas of necrosis and pus accumulation with a few normal foci in upper pole

were seen with pushing the calyces. There were a few stones. Microscopic view (Figure 7) revealed abundant foamy histiocytes (xanthoma cells) aggregation, fibrosis, and acute and chronic inflammatory cell infiltration including giant cells. The inflammatory infiltrate was extended beyond the kidney into the perirenal fat. Michaelis-Gutmann bodies were absent in a specific PAS stain. The diagnosis of XGPN was confirmed by histopathology.

Thereafter, he made a good postoperative recovery. One-year clinical follow-up showed weight gain and normalization of the lab data.



Figure 5: Delayed phase of the similar section of the Figure 4 demonstrating excretion of the contrast media into the posterior upper calyces and persistent small low attenuation masses.



Figure 6: Gross specimen showing yellow lobulated masses diffusely replacing the renal architecture with relative sparing of the upper pole.

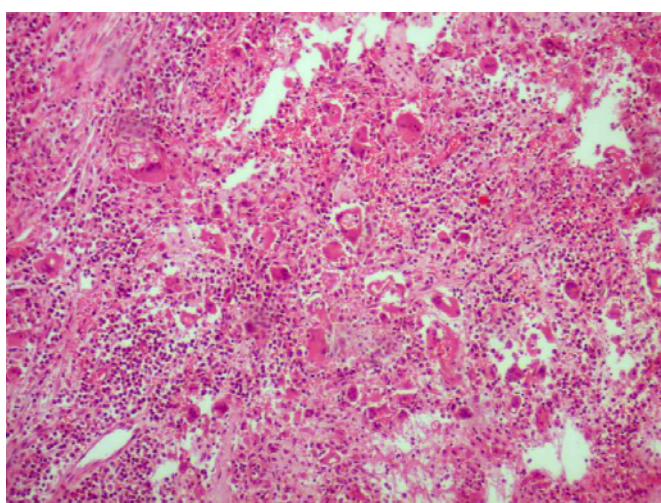


Figure 7: Microscopic view showing inflammatory cell infiltrates, foamy histiocytes and giant cells.

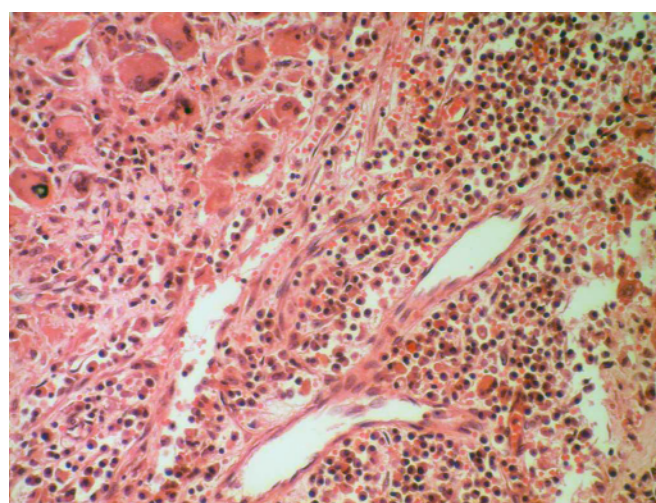


Figure 7 (continued): Higher magnification for detailed evaluation of inflammatory cell infiltrates.

Discussion

XGPN is a specific form of chronic inflammatory kidney disease classically occurring in middle-aged women and is rare but increasingly recognized in children.

Clinical and laboratory data are nonspecific. Patients usually present with signs and symptoms of acute or chronic urinary tract infection, but the presentation overlaps with that of other renal infections. In some cases, the presentation is more subtle and indolent, and neoplasm may be suspected clinically. In imaging, Wilms' tumor is the most important differential diagnosis of the focal form.

The pathology of XGPN is distinctive.⁶ Xanthoma cells are a hallmark of the disease (unlike pyonephrosis) and absent Michaelis-Gutmann bodies must be documented (to rule out malacoplakia).⁷ In the more common diffuse form (75% to 91%), the entire kidney is involved. Both focal and diffuse types of the disease may be found without stones, without obstruction and without documented infection. Imaging can be very useful. In earlier reports, diagnosis was usually made only pathologically after nephrectomy (in 51 cases from 1970 to 1985, not a single preoperative diagnosis was made)⁶; but currently, preoperative diagnosis is the rule.⁸ In one series, 20 to 23 cases examined with CT had XGPN diagnosed preoperatively. Of course, diagnostic difficulties remain particularly with focal form in the absence of calculi. In the most common diffuse form, CT scan demonstrates a complex of findings that allows preoperative diagnosis of XGPN.^{9,10} These CT features are summarized as follows: (1) a central calculus; (2) lack of contrast excretion; (3) low attenuation collections with peripheral enhancing rims; (4) preservation of the renal outline but generalized enlargement of the kidneys; and (5) frequent perinephric stranding. Eighty per cent of cases show calculi on CT. Some of them may be suspended within the pus rather than dependent in the calyx. Most often, a renal pelvic calculus, frequently a staghorn calculus is seen. The collecting system may be dilated, but typically the pelvis is contracted around the calculus resembling the footprint of a bear paw. Numerous collections are seen, spreading from central to peripheral locales. These represent the dilated collecting system filled with thick pus and

the adjacent xanthoma collections and inflammatory tissues. In our case, unlike classic diffuse cases, there was focal excretion of contrast media in upper calyces and negligible peripheral enhancement of collections.

Sometimes, XGPN involves a segment of a duplex kidney with an only partial staghorn. In CT of the focal XGPN, there is a focal bulge with one or more low attenuation collections within. Often, there is a calculus in the associated calyx or infundibulum, or some other obstructing lesion, while the other parts of kidneys are normal.

But in diffuse form, there is no fat density in the renal hilum and calyces never fill with contrast media even on delayed images. In focal (or segmental) form, only a portion of the kidney is involved. But, may diffuse form spare one segment?

We present a case with foci of normal parenchyma in the upper renal pole and excretion of contrast into calyces in the delayed phase of CT scan. There was no evidence of duplex system. Does XGPN make a spectrum with focal form in the one end and diffuse in the other? Is the presented case between the two ends? More facts and cases can be conclusive.

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