XANTHOGRANULOMATOUS PYELONEPHRITIS: ANALYSIS OF 18 CASES

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SUMMARY
Xanthogranulomatous pyelonephritis is a chronic inflammatory renal infection usually seen in middle aged women but extremely rare in children. In this study 18 patients (6 of them were children) with xanthogranulomatous pyelonephritis are reported. Preoperative picture was similar to calculous pyonephrosis, and malnutrition and maldevelopment were seen in these children. Although ultrasonography and computed tomography (CT) suggested perinephritic and retroperitoneal invasion (like psoas abscess), the final diagnosis was established by histologic examination. Following surgical therapy (15 nephrectomy and 3 nephroureterectomy) patients showed excellent improvement.

Key words: Xanthogranulomatous pyelonephritis, urinary infections, pyelonephritis

INTRODUCTION

Xanthogranulomatous pyelonephritis is a very rare, severe chronic renal paranchimal infection and usually calculous nonfunctioning renal mass with chronic urinary infection and obstruction is seen. Its original description was introduced by Schlagenhaufer in 1916, increasing number of cases have been reported in the last decade and reached nearly to 500 cases in the literature (1). Inflammatory process may either involve all the kidney (diffuse) or may be segmental or focal, and also may spread into the surrounding structures. Histologically, it's characterized by a cellulary infiltration of lipit-laden macrophages ("foam cell") (2).

In this study,18 patients with xanthogranulomatous pyelonephritis were analysed and results have been discussed.

MATERIALS AND METHODS

18 cases were treated and histologically diagnosed as xanthogranulomatous pyelonephritis in our clinic between 1979 and May 1988 and reviewed with respect to the symptoms, physical examination, laboratory, radiological findings. The results were discussed and compared with previous reports in the literature.

RESULTS

This study includes 18 cases aged between 3 and 62, 11 of them were male. Twelve of the cases were adults (mean age 41) and 6 of them were children (mean age 8). The patients were admitted with their multiple complaints which had been present for 15 days or 4 years. The symptoms and findings are in Table I.A 15 year old boy who was admitted with a pain radiculating to his knee and unability to stand had flexion deformity on his hip and subsequently diognosed as psoas abcess. One of our patients who was 5 years old had undergone to pyelolithotomy and contralateral ureteral catheterization for urinary obstruction secondary to bilateral renal stones two months ago. Another boy at age 10 in the study although with congenital cataract and paraplegia had pyelolithotomy on the affected kidney. One of the adult women in the study had nephrectomy at another clinic which was unsuccessful. Another female patient was diabetic. One of our patients who was paraplegic had been sistostomized because of spinal cord injury and urethral stricture. Another had postraumatic urethral fistula. There was a recurrent urinary stone disease in 3 patients. There were no special findings in the previous history of our 8 patients.

The right kidney was affected in ten patients and the left kidney was affected in eight patients. The 9 of our 13 patients had multiple kidney stones and 4 had only one stone, another 4 patients had ureteral stone (Fig. 1). Multiple bladder stones were observed in one patient with ureteral stone.

Ultrasonography showed usually an exteremely enlarged hidronephrotic kidney giving hyperechogen shadows due to the stones and including multiple anechoic sonolucent areas due to purulent collection. In one patient perinephritic and psoas abcess showed collection. In another patient perinephritic and psoas abcess showing the spread of xanthogranulomatous process was demonstrated by CT (Fig. 2).

All the patients were explored. 15 patients had

undergone nephrectomy and 3 patients with ureteral stones had undergone nephroureterectomy. In most of the cases the kidneys were extremely enlarged, stretched, oedematous and hard. They partly included nodulary hardness and hidronephrotic areas where paranchyma was quite thin. The kidneys were usually full of yellow, purulent collection. Aspiration of the purulent collection with trochar in order to make the surgical dissection easy was necessary. Various amount of pus ranging between 100 cc and 3500 cc was drained. Usually the affected kidneys were extensively adherent to the surrounding tissues and organs. The xanthogranulomatous inflammotory process was confined to the kidney in 2 cases (Stage 1 or nephric), involved the kidney and perinephritic area in 2 cases (Stage 2 or perinephritic) and affected the kidney and its surrounding fat and involved the retroperitoneum widely in 14 cases (Stage 3 or paranephric) (3). In one case psoas abcess was drained and nearly 150 cc purulent collection was aspirated.

Microscopically, lipit-laden macrophages, plasma cells, giant histiocytes and leukocyts replacing normal renal paranchime were seen. It had been established that the xanthogranulomatous disease invested the surrounding tissue with extensive tissue necrosis.

DISCUSSION

Although xanthogranulomatous pyelonephritis may occur at any age (from 1 month to 87 years) it is most commonly seen during the age of 50 and 60 and affects females more than males (1.5 to 4.1/1) (4,6). Our patients were younger and most of them were men. Xanthogranulomatous pyelonephritis is very rare in children and about 30 cases have been reported in the literature. Six of our cases were children and 4 of them boys. Yazaki et al. reported a boy and they saw that the 4 of 15 patients were boys in English literature and 7 of 8 children were boys in Japanese literature (7). The 7 of 16 patients reported by Kural et al. were children and 6 of these cases were boys (8).

The symptoms and findings of the patients were similar to results of large series dealing with xanthogranulomatous pyelonephritis. But malnutrition and anemia which we observed in children were significant. Anemia shows that inflammatory process is chronic and severe. In xanthogranulomatous pyelonephritis, urine and kidney cultures may give different results. The affecting microorganism cannot be detected in the urine in a septic patient even though the overall clinical condition is guite serious. This situation may occur because of infected urine cannot reach bladder due to urinary obstruction which usually secondary to a stone. Malek et al. found positive kidney cultures in 22 of 23 patients, but they found contaminated urine in only 9 of them (3).

Although, the exact etiology of xanthogranulomatous pyelonephritis isn't known, a lot of factors dealing with each other cause this disease. Experimentally it had been shown that obstruction and infection by ureteral ligation and intravenous administration of specific E. coli serotype, could produce xanthogranulomatous pyelonephritis (10). Most of our patients had urinary obstruction too. They didn't have any therapy or had unsufficient antibiotic therapy. Whether the malnutrition and the maldevelopment we observed in children were the cause of xanthogranulomatous pyelonephritis or occured because of the disease; must be defined. The fact that our patients came fr[illegible] the poor part of the society and their nutritional insufficiency makes the first possibility more reasonable.

The diseases and factors which we have learned from the previous history of our patients, causing stasis, obstruction and infection like paraplegic condition, neurogenic bladder, urethral stricture and diabetes mellitus and increasing the risk of urinary tract infection may play a role in the etiology. However, when we have a look at the literature and evaluate our patients in xanthogranulomatous pyelonephritis it appears that no single etiologic factor is responsible.

Although radiologic findings in xanthogranulomatous pyelonephritis are variable and nonspecific they may give us some signals in diagnosing the disease. The involved kidneys show nonfunction and poor function in IVP. A poorly or nonfunctioning kidney was encountered in 59 to 89 percent of the patients in the literature (2-8). Contralateral kidneys are usually normal or show compansatris hypertrophy. Bilateral xanthogranulomatous pyelonephritis is very rare and only a few patients were reported (8,11).In our series the right kidneys were affected more than the left kidneys, this proportion was 42 and 62 percent in two studies (3,6). In xanthogranulomatous pyelonephritis the incidence of stone is very high; the 94 percent of our patients had urinary tract stone. In our three patients, distal ureteral stones causing hydroureteropyonephrosis are very interesting, because of their sizes and giving late symptoms. Incidence of calculi in xanthogranulomatous pyelonephritis ranges between 59 and 80 percent (2-9).

Ultrasonography and especially CT are very useful in the nonfunctioning or poorly functioning kidneys, where lesions and the stage of the disease have to bedeterminet. Recently, the characteristic occurance of xanthogranulomatous pyelonephritis in CT have been described (12-14). In one of our child patients we demonstrated psoas muscle involvement of the disease. We claim that this is the first report that xanthogranulomatous pyelonephritis causes psoas abcess spreading into the psoas muscle. In our three cases since entire ureter were involved because of the obstruction secondary to the stone in the distal ureter, ureterectomy had been required in addition to nephrectomy. In the literature, the

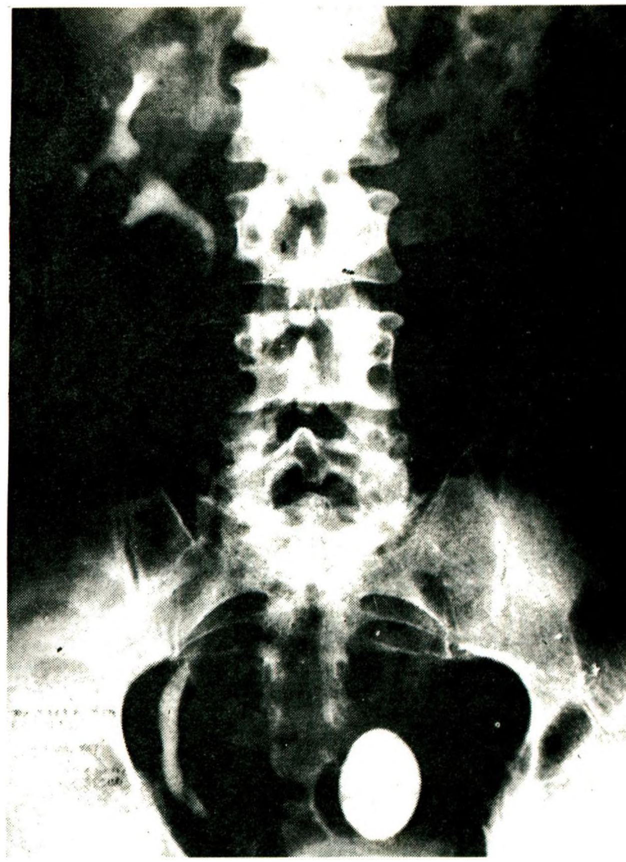
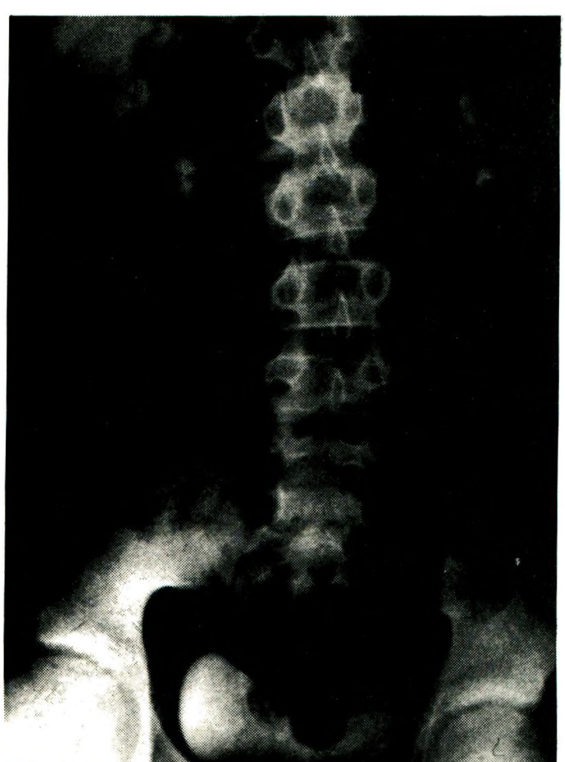
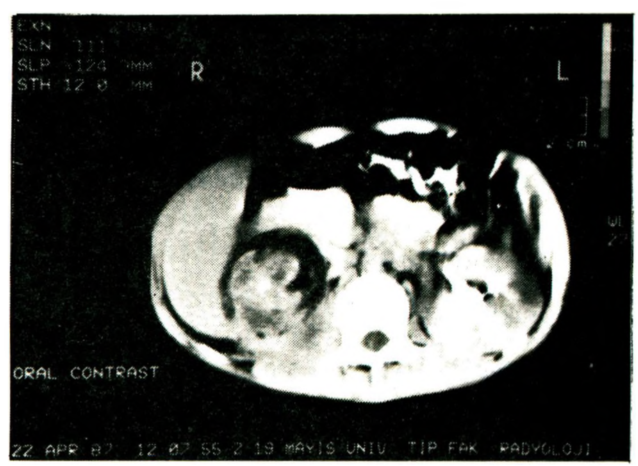


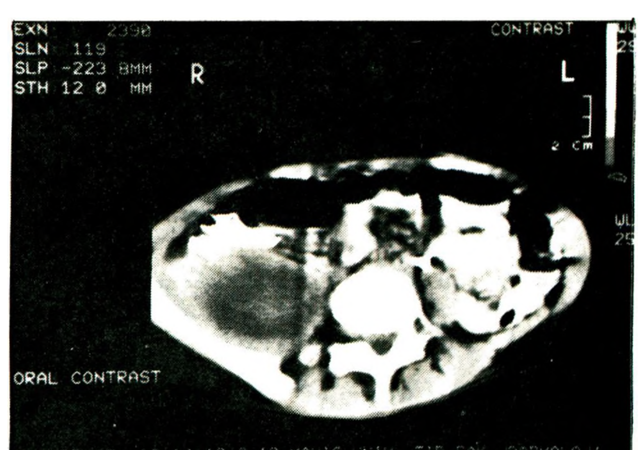
Fig. 1.
Alarge elipsoid stone in the distal left ureter caused by nonfunctioning kidney in a 40 year old male patient who complained of left lumbar pain and dysuria.

Fig. 2.

A— Intravenous pyelogram of 15 year old male patient shows nonfunctioning kidney with multiple stones. Obliteration of left psoas shadow and lumbar scoliosis with the concavity toward the involved side.
B— CT scan at renal hilus level of the same patient reveals calculous nonfunctioning right kidney with spherical low-density areas and irregular posterior pararenal space with low-density area in enlarged right psoas muscle.
C— CT scan of lower lumbar area demonstrated very large low-density area lying within the right psoas muscle which includes diffuse purulent collection.







spreading of the disease into jejunum and pleural area, fistula formation on the skin, in gastrointestinal tract, in lungs and in other parts of urinary system were reported (2,9,15-18). The widespreading of the disease so much and causing complications both spoil the health of the patient and make the operation more difficult. However, xanthogranulomatous pyelonephritis sometimes can also be focal or segmental (5-8).

In summary, xanthogranulomatous pyelonephritis in our series showed some differences than in the literature, because it was frequently seen at younger ages and in males, and in children it was also combined with malnutrition and maldevelopment. Fever and/or chills, lumbar pain, dysuria, lumbar tenderness, lumbar mass and loosing weight are the most prominent symptoms and signs. In addition to anemia, leukositosis and urinary infection, IVP usually reveals a nonfunctioning enlarged kidney with obstructive calculi. Ultrasonography and especially CT show the spreading of the disease into perirenal and also retroperitoneal area and suggest the disease.

An excellent treatment of xanthogranulomatous pyelonephritis requires an affective therapy for com lications in addition to nephrectomy.

Table . The Characteristics of 8 Patients with Xanthogranulomatous Pyelonephritis.

		Patient No.	%
A —	SYMPTOMS		
	Lumbar and/or abdominal pain	7	94
	Fever and/or chills		6
	Dysuria	7	39
	Nausea and vomiting	7	39
	Knee pain and unability to stand.	1	5
	Oliguria - uremia	1	5
B —	PHYSICAL FINDINGS		
	Lumbar tenderness	3	72
	Lumbar mass		6
	Loss of weight	7	39
	Malnutrition and maldevelopment	6	33
	Hepatomegalia		5
C —	LABORATORY FINDINGS		
	Anemia	0	56
	Leukocytosis	9	50
	Proteinuria	5	83
	Hematuria	6	33
	Urine culture		
	E. coli and/or proteus	4	22
	Enterobacter.		5
D —	RADYOLOGIC FINDINGS		
	Renal function (IVP)		
	Nonfunctioning	3	72
	Poorly functioning	5	28
	Urolithiasis	7	94

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