Case Report

Xanthogranulomatous pyelonephritis presenting with thrombocytopenia and renal mass

Majid Malaki, Masood Jamshidi, Farzad Ilkhchooyi

Department of Pediatric Nephrology, Tabriz University Medical School, Children Hospital, Tabriz, Iran

Abstract Xanthogranulomatous pyelonephritis (XGP) is a rare event in children without any predisposing factor like calculi, obstruction or vesicoureteral reflux. In this case we report a four-year-old girl who presented with a renal mass, hematuria, flank pain, anemia and thrombocytopenia—these signs and symptoms misled us to Wilms tumor. Thrombocytopenia which is a strange event in XGP resolved after nephrectomy. Normal contra lateral kidney was infected four months after right nephrectomy. This suggests that these patients should be under strict surveillance and antibiotic prophylaxis as they are a high-risk group for urinary tract infection, and thrombocytopenia should be considered as a laboratory test finding in XGP.

Key Words: Renal mass, thrombocytopenia, xanthogranulomatous pyelonephritis

Address for correspondence:
Dr. Majid Malaki, Tabriz Children Hospital, Post code: 5136735886, Tabriz, Iran. E-mail: madjidmalaki@gmail.com
Received: 31.10.2010 **Accepted:** 26.12.2010

INTRODUCTION

Xanthogranulomatous pyelonephritis (XGP) is defined as a chronic inflammatory disorder of the kidney characterized by a destructive mass that invades the renal parenchyma. Though Schlagenhaufer first described XGP in 1916, the first pediatric case was not described until 1963. Although XGP is rare in the pediatric population, it is found in approximately 16% of pediatric nephrectomy specimens. The exact etiology of XGP is unknown, but it is generally accepted that the disease process requires long-term renal obstruction and infection.[1] In this case report we present a four-year-old girl with renal mass and thrombocytopenia diagnosed as XGP in spite of lacking any predisposing factors like as stone or reflux. She came Although anemia, leukocytosis and elevation of blood sedimentation are considered as the principle laboratory findings in XGP[2] thrombocytopenia can be considered as a new finding in this group of patients, which resolved after nephrectomy in our case.

Access this article online

Quick Response Code:

Website: www.urologyannals.com

DOI: 10.4103/0974-7796.91625

CASE REPORT

A four-year-old girl presented with flank pain that had begun two months ago along with hematuria, pyuria, anemia and thrombocytopenia in her laboratory test results [Table 1]. She also had low-grade fever which resolved after two days by antibiotic therapy. Her growth indices were in normal range (weight 18 kg above +1 SDS and height 98 cm (0 to +1 SDS), her sonography showed a hypo echoic mass, technetium-99m-dimercaptosuccinic acid scan (Tc^{99} DMSA) failed to take radiotracer in the upper and middle part of the right kidney [Figure 1]. Thus contrast computerized tomography (CT) has been done [Figure 2].

Urine culture was positive (50000 colony count of *E. coli*). After a complete antibiotic therapy for *E. coli* infection, her voiding cystourethrography showed nothing.

She went under operation for excising mass, (sized

10 × 7 × 6 cm, weighed 350 g), and spherical creamy shaped mass removed and diagnosed as chronic pyelonephritis. In the first month after her operation her thrombocytopenia resolved.

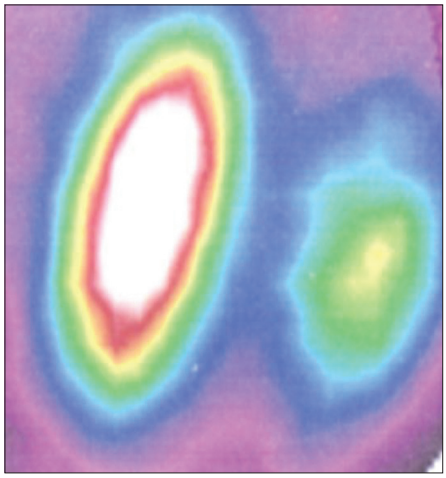

Figure 1: At first admission, Tc99 DMSA scan shows upper part of right kidney has not taken radiotracer, left kidney shows homogenous uptake without scar or acute pyelonephritis. Technetium 99 dimercaptosuccinic acid: TC99 DMSA

In the second admission four months later she was febrile with pyuria, her computerized renal scan showed a new episode of pyelonephritis in her normal left side; [Figure 3] which was not affected earlier, according to her previous DMSA and CT scans. She treated with antibiotics for 14 days. She left hospital with antibiotic prophylaxis with attention to her recurrent urinary tract infection.

Her cystourethrogram showed no vesicoureteral reflux. [Figure 4].

DISCUSSION

Xanthogranulomatous pyelonephritis (XGP) was considered as a rare form of chronic pyelonephritis, which is usually caused by calculus obstructive uropathy[3] and most often occurs in middle-aged women with a history of recurrent urinary tract infection.[4] It seems that XGP in children occurs more

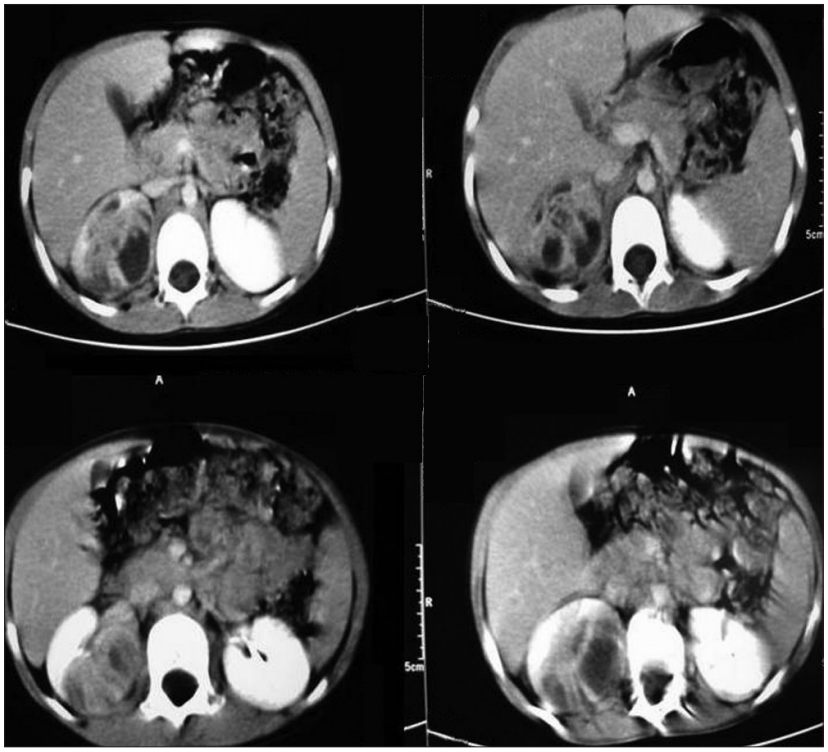

Figure 2: Computerized tomography scan with contrast shows heterogeneous mass in the upper and middle part of the right kidney

Table 1: In first and second admissions her laboratory findings revealed leukocytosis with mild normocytic anemia, high ESR, and thrombocytopenia which recovered after nephrectomy

	Hb (g/dl)/MCV (fL)/ WBC (×1000/μL)	Platelet (×1000/μL)/ESR	Urine WBC	Urine RBC	Specific gravity	Imaging
At admission	10.5/78/11000	76000/60	20-25 HPF	30 HPF	1025	A heterogeneous mass spaced upper, middle part of right kidney
Four months later	11.1/76/13000	178000/40	20 HPF	20 HPF	1020	Hypodense foci in upper part of left kidney

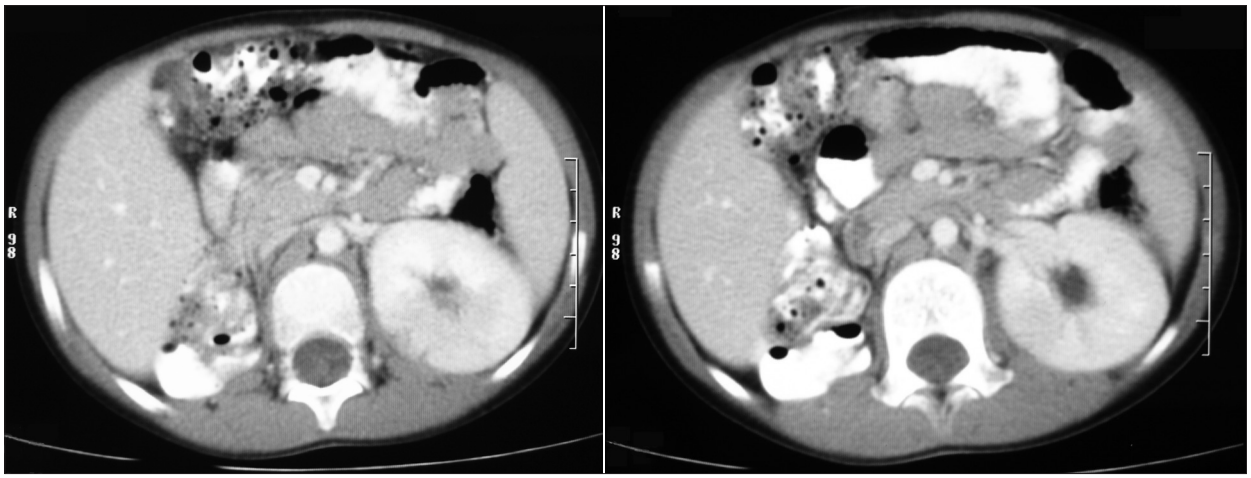

Figure 3: Computerized tomography scan shows hypodense foci of left kidney suggestive of acute pyelonephritis

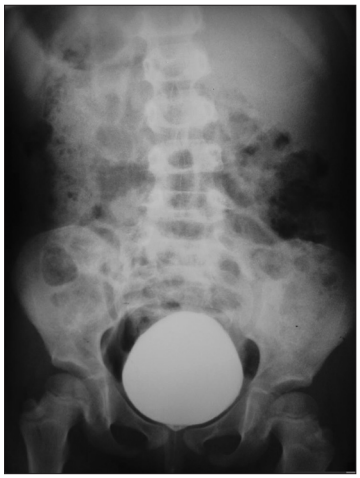

Figure 4: Voiding cystourethrography shows no vesicoureteral reflux, mild trabeculation in bladder wall and vertebral deviation against affected side can be seen

commonly than what was imagined before although only in 18% of children it can be diagnosed correctly.[5]

The typical presenting symptoms include flank pain, fever, malaise, anorexia and weight loss; blood tests reveal nonspecific abnormalities including anemia, an increased erythrocyte sedimentation rate and liver function abnormalities.

The urine analysis reveals urinary tract infection mainly with *E. coli*, proteus mirabilis, pseudomonas, *Streptococcus faecalis* and klebsiella.[6]

There are two different presentations in children. The most common form affects boys and girls equally and involves the entire kidney. The other form occurs more frequently in girls, it is localized and may mimic a tumor, may present with flank and abdominal pain, fever and growth retardation.[7,8]

Two factors mostly associated with XGP include of obstruction and nephrolithiasis (77.8-100%). Blood changes that may occur in them like anemia (100%) and leukocytosis (77-84%) and positive urine culture (30-88.9%).[2,9] Reports of thrombocyte count changes in XGP are scarce but thrombocytosis has been reported by some.[10,11]

Our case was a four-year-old girl without preexisting structural renal disease.

Diagnosed as XGP. Her laboratory findings were similar to previous studies about XGP except a moderate thrombocytopenia, a confusing sign, lead us to neoplasia diagnosis which resolved shortly after nephrectomy. Her left normal side kidney infected after 4 months which pose an question to us that damaged excised renal has protected normal renal side until now? Is total nephrectomy a safer and better management for XGP? Or prophylactic antibiotic should be considered for these groups of patients in spite of having no risk factors like as reflux or stone?

CONCLUSION

We should consider thrombocytopenia as a new laboratory test for XGP in addition in children with XGP strict surveillance for new episodes of pyelonephritis should be considered in spite of lacking predisposing factors.

REFERENCES

1. Tiguert R, Gheiler EL, Yousif R, Tefilli MV, Mills K, Grignon DJ, *et al*. Focal xanthogranulomatous pyelonephritis presenting as a renal tumor with vena caval thrombus. J Urol 1998;160:117-8.
2. Samuel M, Duffy P, Capps S, Mouriquand P, Williams D, Ransley P. Xanthogranulomatous pyelonephritis in childhood. J Pediatr Surg 2001;36:598-601.
3. Parsons MA, Harris SC, Longstaff AJ, Grainger RG. Xanthogranulomatous pyelonephritis: A pathological clinical and aetiological analysis of 87 cases. Diagn Histopathol 1983;6:203-19.
4. Chuang CK, Lai MK, Chang PL, Huang MH, Chu SH, Wu CJ, *et al*. Xanthogranulomatous pyelonephritis: Experience in 36 cases. J Urol

1992;147:333-6.
5. Matthews GJ, McLorie GA, Churchill BA, Steckler RE, Khoury AE. Xanthogranulomatous pyelonephritis in pediatric patients. J Urol 1995;153:1958-9.
6. Goodman M, Curry T, Russell T. Xanthogranulomatous pyelonephriti a local disease with systemic manifestations. Report of 23 patients and review of the literature. Medicine (Baltimore) 1979;58:171-81.
7. Malek RS, Elder JS. Xanthogranulomatous pyelonephritis. A critical analysis of 26 cases and of the literature. J Urol 1978;119:589-93.
8. Moller JC, Kristensen IB. Xanthogranulomatous pyelonephritis. A clinic-pathological study with special reference to pathogenesis. Acta Pathol Microbiol Scand A 1980;88:89-96.
9. Al-Ghazo MA, Ghalayini IF, Matalka II, Al-Kaisi NS, Khader YS. Xanthogranulomatous pyelonephritis: Analysis of 18 cases. Asian J Surg 2006;29:257-61
10. Eckoldt F, Riebel T, Wolke S. Xanthogranulomatous pyelonephritis in children: diagnostic and therapeutic aspects. J Med Ultrasound 2009;36:33-7.
11. Chang JW, Chen SJ, Chin TW, Tsai H, Pan CC, Chu YK, *et al*. Xanthogranulomatous pyelonephritis treated by partial nephrectomy. Pediatr Nephrol 2004;19:1164-7.

How to cite this article: Malaki M, Jamshidi M, Ilkhchooyi F. Xanthogranulomatous pyelonephritis presenting with thrombocytopenia and renal mass. Urol Ann 2012;4:51-4.

Source of Support: Nil, **Conflict of Interest:** None.