Xanthogranulomatous pyelonephritis in a 1-year old boy

Derek P. Ottem, MD, Andrew E. MacNeily, MD, Iain W. McAuley, MD
Division of Pediatric Urology, BC Children's Hospital, Vancouver, British Columbia, Canada

OTTEM DP, MACNEILY AE, MCAULEY IW. Xanthogranulomatous pyelonephritis in a 1-year old boy. The Canadian Journal of Urology. 2005; 12(6):2901-2903.

Xanthogranulomatous pyelonephritis (XGP) is a rare, chronic inflammatory lesion of the kidney associated with both chronic infection and obstruction. Most common in middle-aged females, it is important to recognize in children because it may be confused with childhood renal malignancies particularly Wilms' tumor. We report a case of a 1-year old male with antenatally detected and postnatally confirmed hydronephrosis. Further investigation revealed a left incomplete duplex collecting system with obstructing lower pole stones, pelvicaliectasis and widespread foci of calcification. He underwent a left nephroureterectomy with the pathological report noting XGP.

Key Words: xanthogranulomatous pyelonephritis, childhood renal mass, chronic infection

Introduction

XGP is a rare but well recognized variant of pyelonephritis. It is most commonly found in middle-aged females and is infrequently noted in children. Lesions are characterized by the destruction of renal parenchyma with replacement by granulomatous tissue containing lipid laden macrophages (foam cells).[1] There may be invasion of adjacent tissue mimicking renal tumors.[2] The disease is associated with urinary obstruction and urolithiasis with cultures often positive for *E. coli* or *Proteus* species. In most cases there is nonfunction of the kidney.[3]

The clinical picture is typically non-specific and variable. Preoperative diagnosis is difficult because the lesion may resemble malignant conditions radiologically. Treatment typically involves nephrectomy or partial nephrectomy, although percutaneous drainage with antibiotics and renal preservation has been reported.[2]

Case report

An asymptomatic male was followed since birth for antenatally detected left hydronephrosis. A VCUG

Accepted for publication November 2005

Address correspondence to Dr. Andrew E. MacNeily, Head, Division of Pediatric Urology, BC Children's Hospital, Level Ambulatory Care Bldg., 4500 Oak Street, Vancouver, BC V6H 3V4 Canada

performed at 6 weeks of age was normal. A catheter specimen taken during the VCUG grew *Proteus mirabilis* which was treated. MAG-3 renogram at 6 weeks revealed a delay in left lower pole uptake and excretion. Follow up ultrasound at 7 months revealed persistent moderate hydronephrosis with a new finding of multiple stones in the lower, middle and upper calyces, Figure 1. Metabolic stone disease was ruled out. Cystoscopy and retrograde pyelography performed shortly thereafter displayed an incomplete duplication of the left collecting system with a stricture distal to the confluence, and a likely UPJ obstruction of the lower pole with obstructing calculi, Figure 2. The MAG-3 renogram was repeated at 10 months of age and showed delayed perfusion with obstruction of the upper pole and nonfunction of the lower pole. The patient remained asymptomatic.

Initially, at 13 months of age, a left lower pole heminephrectomy through a posterior lumbotomy incision was planned, but examination under anesthetic revealed the new finding of a large palpable abdominal mass dictating the need for greater exposure via a transverse peritoneal approach. Intraoperatively, both the upper and lower poles were filled with pus which was drained prior to resection. The kidney was adherent to surrounding structures with multiple parasitic vessels identified. Macroscopically, the kidney was grossly distorted with an abundant amount of necrotic debris filling a dilated pelvicalyceal system and destruction of the overlying parenchyma. Microscopically there was massive destruction of the lower pole parenchyma and replacement with a granulomatous layer of tissue with fat laden macrophages. Severe interstitial nephritis of the upper pole was noted.

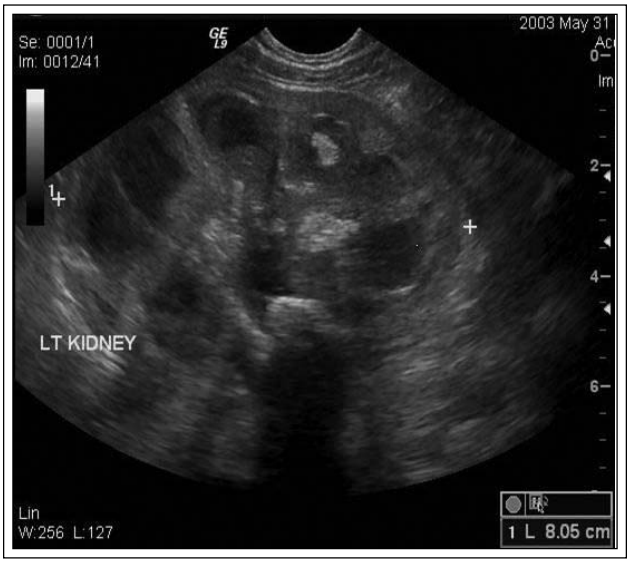

Figure 1. Renal ultrasound at 7 months. Moderate hydronephrosis with multiple calculi in upper, middle and lower poles.

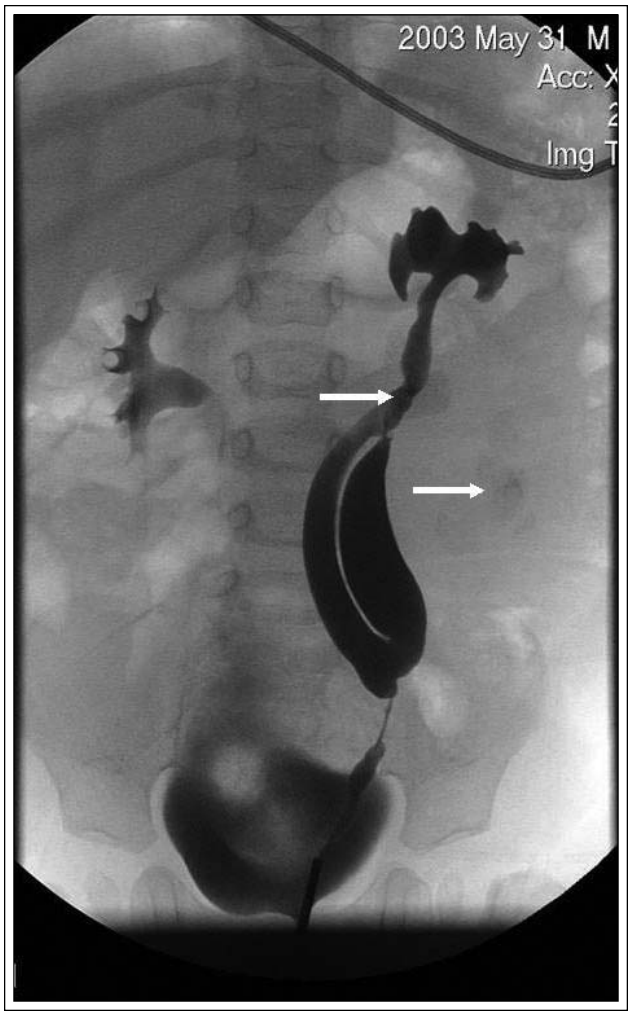

Figure 2. Retrograde pyelography demonstrating incomplete duplication of the left collecting system with a stricture at the confluence, and a UPJ obstruction of the lower pole with an obstructing calculi.

Discussion

The diagnosis of XGP in children is difficult and elusive. Patients typically present with nonspecific complaints such as fever, fatigue, anorexia and weight loss.[4] Flank pain and hematuria may help point to a renal source. Almost all cases are anemic, and this is a useful clue to diagnosis.[5] XGP usually results from an underlying urological condition so the knowledge or discovery of this in a suspected case should further raise suspicion.

In a large Turkish series the common underlying conditions were renal or bladder calculi, reflux, ureteropelvic junction obstruction, and exstrophy.[2] Renal calculi are particularly important for radiological differentiation from other causes of a renal mass.6 Most series report a slight propensity for the left kidney, and the frequency appears to be equal between boys and girls unlike in adults.[7] Our case appears to be typical in clinical presentation, with minimal vague symptoms but profound anemia noted preoperatively. His hemoglobin was 84. An interesting feature of our case, which has been noted in other series, is the relatively rapid progression. XGP is generally regarded as a slowly progressing disease evolving from a milieu of chronic infection and inflammation typically in the setting of obstruction. Matthews et al reported XGP in two cases younger than 6 months and noted the acute evolution in a compromised renal unit.[9] Similarly, in our patient we could appreciate the rapidity of progression. In the 2 months between the last clinical visit and the date of operation the renal mass had evolved to a large palpable flank mass extending medially towards the midline where before no mass was detected. In fact, we changed our operative approach to account for this new finding.

Ultrasound is recommended as the most useful initial imaging study when working up a patient with the above symptoms and a suspected urological cause.[8] Typical findings are a massively enlarged kidney which retains it's reniform shape and calculi. Duplication is often noted.[1] IVP's were once commonly performed, and showed a non-functioning kidney in as many as 80% of cases.2 Now nuclear medicine studies are used to evaluate renal function. CT demonstrates peripheral enhancement with calyceal dilatation, the "bear paw" sign. XGP is occasionally misdiagnosed as Wilms' tumor and, rarely, visa versa. Features on CT favoring a diagnosis of XGP rather than Wilms' tumor are perirenal inflammation, and obstructing calculi. Wilms' tumors rarely invade the collecting system.[6] As mentioned above XGP may evolve rapidly. In our case it was interesting to observe the radiological progression over time. What initially presented as antenatal hydronephrosis developed into an obstructed infected system promoting the development of stones and renal destruction. A viable lower pole became non functional over several months time.

Conclusion

XGP should be considered when a renal mass is associated with an underlying urological condition, particularly obstruction. Anemia is very common. Distinguishing radiological features include a non-functional kidney with calculi that retains its reniform shape. The disease may progress rapidly, and in children early diagnosis may allow heminephrectomy rather than nephrectomy. □

References

1. Clapton WK et al. Clinicopathological features of xanthogranulomatous pyelonephritis in infancy. *Pathology* 1993;25:110-113.
2. Bingol-Kologlu M et al. Xanthogranulomatous pyelonephritis in children: Diagnostic and therapeutic aspects. *European Journal of Pediatric Surgery* 2002;12:42-48.
3. Hammadeh MY et al. Xanthogranulomatous pyelonephritis in childhood: pre-operative diagnosis is possible. *British Journal of Urology* 1994;73:83-86.
4. Kural AR et al. Xanthogranulomatous pyelonephritis in children and adults. *British Journal of Urology* 1987;59:383-385.
5. Goodman TR et al. Paediatric xanthogranulomatous pyelonephritis. *International Journal of Clinical Practice* 1998;52:43-45.
6. Groeneveld D et al. Intrapelvic Wilms' tumor simulating xanthogranulomatous pyelonephritis. *Pediatric Radiology* 1995;25:68-69.
7. Braun G et al. Xanthogranulomatous pyelonephritis in children. *The Journal of Urology* 1985;133:236-239.
8. Cousins C et al. Xanthogranulomatous pyelonephritis in childhood: ultrasound and CT diagnosis. *Pediatric Radiology* 1994;24:210-212.
9. Matthews G et al. Xanthogranulomatous pyelonephritis in pediatric patients. *The Journal of Urology* 1995;153:1958-1959