# Xanthogranulomatous Pyelonephritis With Unconnected Liver Lesion

**Abdurrahman Karaman, Emine Şamdancı, Metin Doğan, Rauf Tuğrul Aksoy, Ahmet Sığırcı, and Mehmet Demircan**

Xanthogranulomatous pyelonephritis is a chronic renal inflammation characterized by destruction and replacement of its parenchyma with granulomatous tissue. This uncommon condition is rare in children. We report on a 5-month-old male infant with a left renal and hepatic mass detected by ultrasound. The case was preoperatively misdiagnosed as Wilms' tumor and total nephrectomy and biopsy from liver lesion were performed. The subsequent histopathological findings confirmed the diagnosis of xanthogranulomatous pyelonephritis for renal and liver lesions. Increasing awareness of this disease should lead to the diagnosis being suspected preoperatively even if it is with unconnected tissue lesions. UROLOGY 78: 189–191, 2011. © 2011 Elsevier Inc.

Xanthogranulomatous pyelonephritis (XGP) is a distinct, atypical, and chronic renal parenchymal infection. This condition is a rare but well-recognized clinicopathological entity characterized by destruction of the renal parenchyma and replacement by granulomatous tissue containing foamy lipid-laden macrophages (xanthoma cells).[1-3] Classically, this entity is usually associated with urinary tract obstruction, kidney stones, and infection. *Proteus mirabilis* and *Escherichia coli* are the most frequent microorganisms.[4,5] This rare pathology is usually observed in middle-aged women, and is much rarer in children.[1-6] XGP is classified into diffuse and focal forms that is often misdiagnosed as a renal tumor.[1-3]

The spread of this pathology into perirenal fat or connected tissues has been described, such as the psoas, duodenum, spleen, skin, colon, and diaphragm.[2,3] However the association of XGP with unconnected tissue abscess is very rare. Here we report a child with focal XGP in the left kidney and its extension into the right liver lobe without connection.

## CASE REPORT

A 5-month-old male infant was admitted with persistent crying, irritability, and loss of appetite for 2 days. There were no relevant prenatal and labor histories. At admission, body temperature was normal; physical examination was unremarkable. Laboratory findings were as follows: white blood cell count 15,300/mm$^3$, hemoglobin 9.8 g/dL, platelet count 477,000, and markedly elevated C-reactive protein (82.6 mg/L). Blood biochemistry results revealed mildly elevated aspartate aminotransferase levels (47 U/L), and markedly elevated lactate dehydrogenase levels (311 U/L). Urine culture was positive for *Escherichia coli*. At admission, renal ultrasound showed a tumor mass in the middle of the left kidney. Computed tomography (CT) scan revealed a heterogenous and hypodense mass in the middle of the left kidney and a hyperechoic mass in liver segment 7 (Fig. 1). Wilms' tumor with liver metastasis was strongly suspected and the left nephrectomy and biopsy from hepatic lesion was performed. Cefoperazone/sulbactam was initiated on the preoperative period. Seven days postoperatively, the cefoperazone/sulbactam regimen was changed to ampicillin/sulbactam.

The cut section of nephrectomy specimen showed a well-circumscribed multicystic mass that contained purulent material (Fig. 2). There were no gross developmental abnormalities of kidney and calculi.

Microscopically, the mass of kidney had a granulomatous inflammatory infiltrate that consisted of predominantly foamy histiocytes, lymphocytes, neutrophils, plasma cells, and scattered multinucleated giant cells.

Microscopic findings of the liver biopsy were similar to the mass of kidney, but there were no hepatocytes, portal areas, and central vein. Immunohistochemically, bile ducts were stained with cytokeratin 19 antibody (Fig. 3) (immunoperoxidase) and foamy histiocytes were stained with CD 68 antibody (immunoperoxidase).

In the postoperative period, examination of the patient did not reveal an underlying disease. Treatment with ampicillin/sulbactam was continued for 6 weeks.

Follow-up abdominal ultrasound after 6 months revealed that the size of the liver lesion had markedly decreased. The patient is currently doing well, with no complications.

*From the Department of Pediatric Surgery, Inonu University, Faculty of Medicine, Turgut Ozal Medical Center; Department of Pathology, Inonu University, Faculty of Medicine, Turgut Ozal Medical Center; and Department of Radiology, Inonu University, Faculty of Medicine, Turgut Ozal Medical Center Malatya, Turkey*

*Reprint requests: Abdurrahman Karaman, M.D., Assoc. Prof. of Pediatric Surgery, Inonu University Tip Fakultesi, Turgut Ozal Tip Merkezi, Cocuk Cerrahisi Anabilim Dali, Malatya, Turkey. E-mail: akaraman@inonu.edu.tr*

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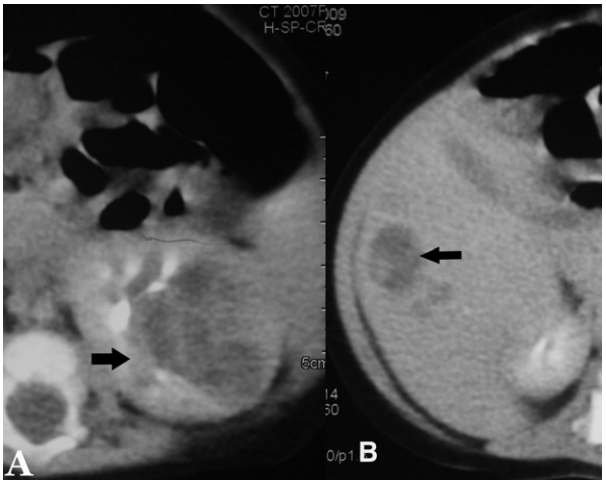


**Figure 1.** CT scan revealed a 38 × 35-mm heterogenous and hypoechoic mass in the middle of the left kidney (**A**, *arrow*) and 2 × 2-cmhyperechoic mass in liver segment 7 (**B**, *arrow*).

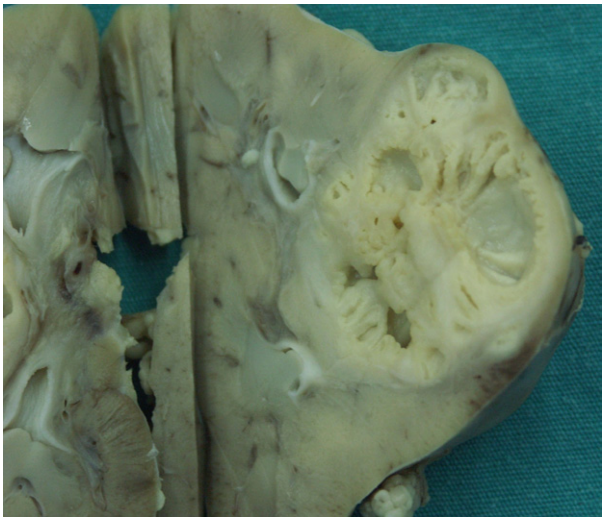

**Figure 2.** Cut surface of the left nephrectomy presented the mass localized in the upper-mid pole.

## COMMENT

XGP is an unusual complicated form of pyelonephritis. The first descriptions of the specific macroscopic features of this disease were made by Schlagenhaufer in 1916.[2] Its name is derived from the yellow (xantho) color on gross pathology and a granulomatous reaction histologically.

XGP is classified as diffuse (92%) or focal (8%)[1] and is extremely rare in children younger than the age of 1 year.[1,2] The etiology of XGP is still unclear but it is clearly demonstrated that a combination of impairment of the urinary flow and chronic bacterial infection are the 2 most consistently associated factors that predispose to the development of XGP. *Escherichia coli* and *Proteus* species are the most commonly implicated organisms.[3-6]

XGP has been named as the "great imitator" because of its close resemblance clinically, radiologically, and pathologically to the other renal disorders.[2] Unlike adults, the symptoms in children are usually nonspecific and associated with more vague symptoms of general malaise, anorexia, failure to thrive, and weight loss.[1,3,4] The radiological features of XGP are nonspecific. In diffuse form, increased renal size, replacement of the normal parenchyma with multiple fluid-filled masses, and renal stones can be found on ultrasonography. With focal XGP, the involved portion of the kidney may appear masslike. The most characteristic CT finding is a strongly enhanced rim of tissue surrounding the hydronephrotic areas (bear paw sign).[4,5]

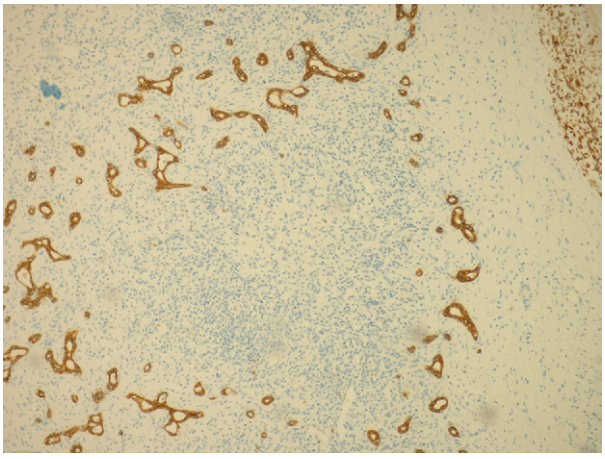

**Figure 3.** Liver replaced by fibrosis and granulomatous inflammation that include bile ducts. Immunohistochemistry revealed that the bile ducts were positive for cytokeratin19 (original magnification ×100).

XGP usually resembles the other renal neoplastic lesions such as Wilms' tumor, clear cell sarcoma, and mesoblastic nephroma. This condition may result in misdiagnosis, as in our case. The correct diagnosis and the differentiation from renal malignancy is established mostly by the histopathological examination only.[1-4]

Total nephrectomy remains the standard and curative treatment for diffuse type, but partial nephrectomy is advocated whenever possible, especially in children with focal type. Different surgical approaches can be scheduled for the treatment of XGP, such as open with flank incision or the anterolateral transperitoneal approach or laparoscopic surgery. It has been suggested that the standard kidney approach is adequate for total or partial nephrectomy for pediatric XGP, although others have recommended the anterolateral transperitoneal approach because of the difficulties during nephrectomy, with the feasibility of the safe excision of surrounding tissue. On occasion, preoperative percutaneous drainage of an extrarenal soft tissue abscess may be performed in an attempt to minimize potential surgical complications. Compared with open surgery, laparoscopic nephrectomy can be more complicated because of perirenal inflammatory changes. However, there are rare reports of successful treatment of focal XGP with antibiotics only in children.[1,3,4,7] Histologically, it is a granulomatous inflammation characterized by solid sheets of lipid-laden macrophages

(xanthoma cell) and inflammatory cells destroying renal parenchyma and invading adjacent tissues.[5]

It is demonstrated that inflammation resulting from XGP may extend into the perirenal and pararenal spaces and this condition may progress to perinephric abscess formation. In a review of the literature, we found 2 reports of liver involved in the process. In one, Esposito et al reported a large mass in the upper pole of the right kidney, which was adherent to the liver and the diaphragm but with no extension into the liver tissue.[8] In another report, Taskinen et al presented a patient who had XGP in conjunction with caliceal diverticulum infiltrating the liver.[9] In both of these 2 reports, XGP was in right kidney and the extension of inflammation developed via tissue connection, and cases of XGP that infiltrated unconnected liver tissue had not been published previously. We report here for the first time a patient with XGP in the left kidney with inflammatory infiltration in an unconnected liver segment that was detected in early childhood. Its mechanism is not clear but septic spreading may play an important role. Hortling et al presented an unusual case of XGP with septic spreading into the lungs mimicking pulmonary metastasis and with an inflammatory infiltration of the descending colon.[10]

XGP is a very rare condition in children, particularly in infants. Enhancing the clinical awareness of pediatricians about XGP is very important to achieve the correct preoperative diagnosis. Surgical treatment still remains the cornerstone of treatment for this rare disease, but early diagnosis and treatment may change the natural history, particularly in focal XGP, and prevent the need for total nephrectomy.

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