

A case of xanthogranulomatous pyelonephritis mimicking Wilms tumor

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Received: 29 September 2014, Revised: 14 November 2014, Accepted: 26 November 2014

SUMMARY: Tüysüz G, Tayfun F, Canpolat F, Zeytun H, Goya C, Keleş AN, Özdemir N. A case of xanthogranulomatous pyelonephritis mimicking Wilms tumor. *Turk J Pediatr* 2015; 57: 409-412.

Xanthogranulomatous pyelonephritis (XGPN) is a very rare, unusual variant of pyelonephritis characterized by destruction of renal parenchyma. It usually occurs in adults with a history of recurrent urinary tract infections. The condition is rare in children and the disease can imitate renal tumors. Here, we describe a 12-year-old boy who presented with abdominal pain. He did not have any history of urinary tract infection. Computed tomography and magnetic resonance imaging showed a cystic lesion in the left upper kidney. The patient underwent radical nephrectomy with a provisional diagnosis of Wilms tumor however histopathological examination of specimen revealed XGPN. Xanthogranulomatous pyelonephritis should be kept in mind in the differential diagnosis of renal lesions in childhood, during surgery if any suspicion from the diagnosis, a frozen biopsy should have been taken.

Key words: *pyelonephritis, Wilms tumor, children, magnetic resonance imaging, computed tomography.*

Xanthogranulomatous pyelonephritis (XGPN) is a rare form of chronic renal infection with unknown pathogenesis. It is usually associated with recurrent urinary tract infections and renal calculus causing obstructive uropathy. The disease process is characterized by the destruction of the renal parenchyma and surrounding tissues and replacement with lipid-laden macrophages. Xanthogranulomatous pyelonephritis can involve the entire kidney or may be localized, mimicking a renal tumor. It is a challenge in the differential diagnosis of renal tumors. We report a case of XGPN in a 12-year-old boy, who underwent radical nephrectomy due to left kidney upper pole located cystic lesion with the provisional diagnosis of Wilms tumor. The histopathological examination of the removed kidney showed XGPN.

Case Report

A twelve-year-old boy presented with abdominal pain ongoing for the last 17 days. Medical

history of the patient was unexceptional; he did not have any previous history of hospitalization, unexplained fever or urinary tract infection before. His body temperature was normal. General and systemic examination of patient was unremarkable except tenderness on left upper quadrant at deep palpation. The blood pressure was within the normal limits. Blood chemistry tests were normal except the mildly elevated C-reactive protein (value: 10.34 mg/dl, normal: < 0.5 mg/dl). Complete blood count was unremarkable: white blood cell count, 9710/mm³ of which 60.2% neutrophils, 28.7% lymphocytes, 9.89% monocytes, 1.21% eosinophils. The urine analysis was normal. Ultrasonography of the abdomen showed a renal lesion on left upper lobe of kidney (Fig. 1). The patient was subsequently hospitalized for further investigation. An intravenous contrast enhanced computed tomography (CT) scan was performed which revealed a renal mass, 54x47 mm sized, originating from the left upper pole

of the kidney and extending to the medial part. There was thickening and mild density increase at anterior pararenal fascia. Both of the kidneys were in normal size and location (Fig. 2). A T1 weighted abdominal magnetic resonance imaging (MRI) showed 60x58 mm sized cystic lesion containing a few septa with an 8 mm thickness wall on left upper pole of the left kidney. Around the lesion, there was diffusion restriction on diffusion weighted scan and contrast enhancement at septa and at the wall of the lesion (Fig. 3). The patient was evaluated at tumor council; on the basis of radiologic findings, despite the rare

incidence above 10 years of age, the patient was clinically diagnosed as Wilms tumor. A total nephrectomy was planned according to the national Wilms tumor staging (NWTs) protocol. Total nephrectomy was done. Pathological examination of the specimen showed upper middle pole located, predominantly cystic, 4.5x3.5x3.5 cm sized, a brown colored mass containing 2.8x1x1 cm solid area on the wall in 10.5x6x5 cm sized left kidney operation material. Microscopic examination of the mass showed macrophage predominant mixed type inflammatory zone lining the cyst wall and a restrictive fibrous zone characterized by connective tissue surrounding the inflammatory zone (Fig. 4) which revealed the presence of xanthogranulomatous pyelonephritis. He is doing well now with no complaints or any signs of urinary tract infection at three-month follow-up after surgery.

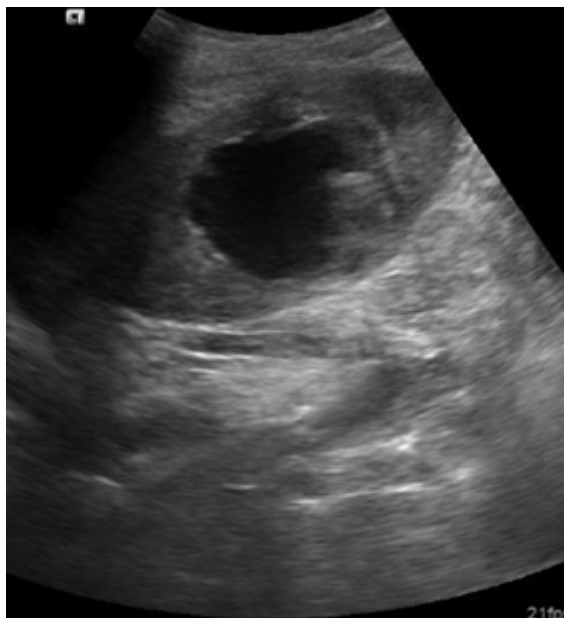


Fig. 1. Ultrasonography showed anechoic cystic lesion with a thick irregular wall on upper pole of left kidney.

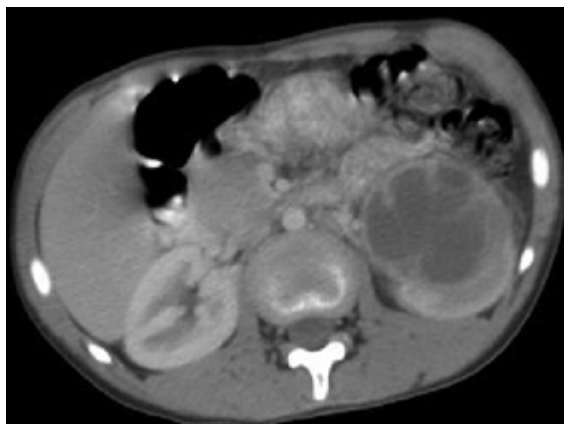


Fig. 2. Abdominal contrast CT scan showed cystic lesion with enhancing septa and wall at upper pole of left kidney in axial plan.

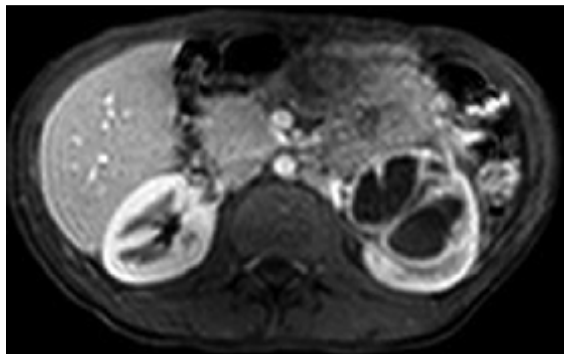


Fig. 3. Axial T1-weighted fat-suppressed MRI image with contrast showed cystic lesion with septa on upper pole of left kidney.

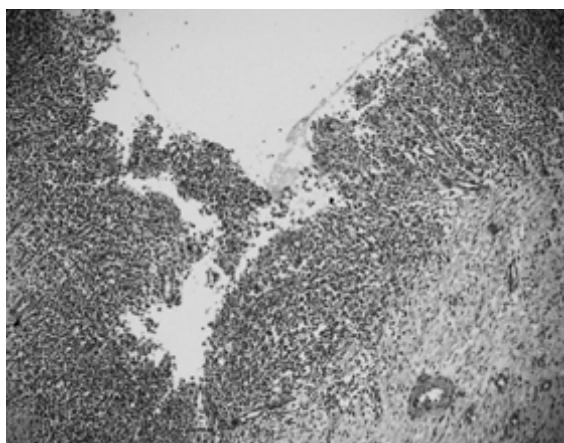


Fig. 4. Pathological examination of renal mass (H&E X100) showed macrophage predominant mixed type inflammatory zone lining the cyst wall and a restrictive fibrous zone characterized by connective tissue surrounding the inflammatory zone.

Discussion

Xanthogranulomatous pyelonephritis is a severe form of chronic renal infection characterized by destruction of renal parenchyma and replacement by granulomatous tissue containing lipid filled (xantoma cells) macrophages¹. These macrophages form the special yellow color of the disease. Although chronic bacterial infection accompanied with urinary tract obstruction is the common pathology, exact etiology still remains unknown^{2,3}. The urinary tract obstruction is usually secondary to nephrolithiasis, ureteropelvic junction obstructions, renal tumors or severe vesicoureteral reflux. The most common causative organisms for infection are *Escherichia coli*, *Proteus mirabilis* and *Pseudomonas aeruginosa* (4). Other factors that might have an effect in development of disease are renal ischemia, arterial insufficiency, lymphatic blockage, bacteremic seeding, alterations in lipid metabolism and altered immune response in renal transplant recipients⁵⁻⁷.

The disease is mostly seen in middle aged women but can occur at any age. In adults, male to female ratio is 1:2⁸. In pediatric cases, it is very rare; girls and boys are equally affected, the age of onset varies but usually patients are diagnosed before age of 5⁹. Xanthogranulomatous pyelonephritis usually affects one kidney, but rarely can involve both kidneys. Clinical symptoms and signs are nonspecific and include abdominal pain, flank mass, fever of unknown origin, weight loss, pyuria and rarely hematuria^{8,10}. These symptoms are reported in most of the cases. Laboratory tests are usually non-diagnostic like mild anemia, leukocytosis, elevated erythrocyte sedimentation rate and increased C-reactive protein levels^{9,11}. Pathologic urine sediment and positive urine cultures are prognostic for XGPN but are found only in 70% of patients¹².

Radiologic investigations including ultrasonography and contrast-enhanced computed tomography can help in identifying XGPN. Radiologic image of XGPN differs according to the type of the disease. In rather frequently seen, diffuse form; there are more characteristic lesions like diffuse enlargement of kidney, loss of renal architecture, perirenal spread of infectious process and accompanying fistulae in CT. Diffuse forms can be divided

into 3 stages, depending on the extension of the inflammation: in stage I (nephric XGPN) the lesion is confined to kidney, in stage II (perinephric XGPN) the lesion extends to Gerota's fascia and in stage III (paranephric XGPN) extends to paranephric space and other retroperitoneal structures¹³. In the focal form, like in our patient, ultrasonography shows a non-enhancing or minimally enhancing localized renal mass that can mimic renal tumors and this form is usually combined with staghorn stones. Although radiologic images are suitable with Wilms tumor in our patient, renal cell carcinoma, renal abscess, infected renal cystic disease and tuberculosis should be kept in mind in the differential diagnosis of these cystic lesions of kidney. If any suspicion from the diagnosis, during the surgery, a frozen biopsy should be taken even though this can split the tumor to abdomen in case it turns out to be Wilms tumor.

The management of XGPN differs in literature according to the dissemination of disease. In diffuse forms, nephrectomy is usually the standard treatment. In focal forms partial nephrectomy, drainage of perirenal/renal abscess and concomitant antibiotic therapy has been recommended but most of the cases had gone under renal explorations because focal XGPN resembles renal tumors^{14,15}. Because there is no clear guideline on the management of XGPN, conservative and surgical treatments should be considered for each individual case.

In our case, there was no history of urinary tract infection or unexplained fever. Urine sample and CBC tests showed no abnormality. Blood chemistry and systemic inflammatory reactants were unremarkable except the increased C-reactive protein level. Ultrasonography and CT showed a cystic lesion originating from the left pole of kidney but there weren't any renal calculi. The preoperative distinction between XGPN and malignant kidney tumor is often difficult especially in focal forms and in the absence of previous urinary tract complaints. Here, we wanted to emphasize that XGPN should be kept in mind in the differential diagnosis of renal masses even in the absence of predisposing factors. A high index of suspicion and clinical awareness is needed to accomplish preoperative diagnosis.

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