

Inflammation and Infection

Xanthogranulomatous Pyelonephritis Can Simulate a Complex Cyst: Case Description and Review of Literature[☆]



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ABSTRACT

Xanthogranulomatous pyelonephritis is a rare and peculiar form of chronic pyelonephritis and is generally associated with renal lithiasis. Its incidence is higher in females. The peculiarity of this disease is that it requires a differential diagnosis, because it can often simulate dramatic pathologic conditions. In fact, in the literature are also described cases in association with squamous cell carcinoma of the kidney. The radiologic clinical findings simulate renal masses, sometimes in association with caval thrombus. We describe a case of xanthogranulomatous pyelonephritis with radiologic aspects of a complex cyst of Bosniak class III in a man 40-year old.

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Introduction

Xanthogranulomatous pyelonephritis (XGP) is an uncommon and distinct type of chronic infective pyelonephritis in which yellow lobulated masses diffusely replace the renal architecture. XGP predominantly affects middle-aged women, although infants and very old men are also affected. The most common symptoms include abdominal pain, fever, a palpable mass, anorexia and weight loss, a urinary tract infection resistant to antibiotics, hematuria, and dysuria.

The disease is characterized by an accumulation of foamy histiocytes, macrophages with mature adipocytes, and occasional giant cells. Anemia, leukocytosis, and increased erythrocyte sedimentation rate comprise the usual laboratory findings.

Its etiology remains unclear, although as many as 6 causes have been proposed: (1) urinary obstruction, (2) urinary tract infection, (3) abnormal lipid metabolism, (4) lymphatic obstruction, (5) altered immune response, and (6) vascular occlusion.¹ *Escherichia coli* and *Proteus mirabilis* are the most common offending microorganisms despite sterile urine in approximately one-third of patients.

Two forms of XGP have been described: diffuse (83%–90%) and focal (10%–17%). The latter has been described as “the great imitator”, as it is often misdiagnosed as a renal mass.² Malek and Elder³ proposed a staging system for XGP: stage I, the lesion is confined to the kidney; stage II, there is an infiltration of the Gerota space; and stage III, XGP extends to the perinephric space and other retroperitoneal structures. Pseudoinflammatory tumors that are similar to XGP can affect many organs, including the gallbladder, appendix, bone, ovaries, bladder, rectum, prostate, epididymis, and endometrium.

Case presentation

According to the guidelines of our ethics committee, the patient has signed the consent to the publication of his case and of all the photographic material relating to him. A 40-year-old man presented with left lumbar back pain. He had a medical history of left lumbar pain, meteoric bowels, and a drug allergy (nonsteroidal anti-inflammatory drugs). The urologic examination detected a monolateral left positive sign of Giordano, and the left kidney area and costovertebral angle were tender on palpation. The ureteral trigger points on the left side were negative to deep palpation, and the abdomen was tractable. The results of blood and urine tests were within the normal range.

The urologic ultrasonography (Fig. 1) showed an expansive cystic formation of approximately 80 mm in the middle third of the left

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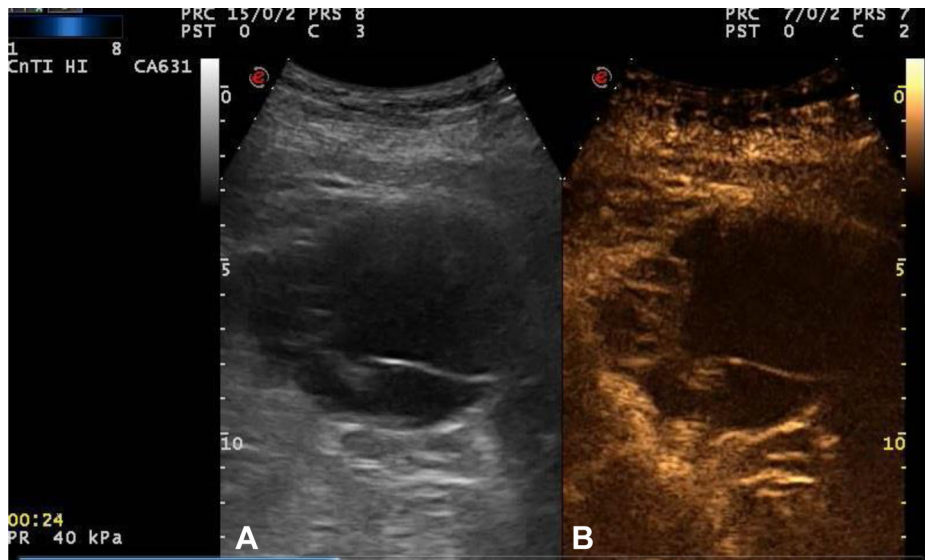


Figure 1. B-mode (A) and contrast-enhanced (B) ultrasonographic axial images. Grayscale ultrasonography shows a complex cystic renal mass with echogenic irregular nodular components along the right cystic wall as well as intracystic septa. Contrast-enhanced ultrasound depicts intense contrast enhancement of the solid intracystic mural nodules, suggesting a diagnosis of cystic renal cell carcinoma.

kidney, which was predominantly exophytic but at the same time had a lateral component wedged in the context of the renal sinus.

Uro-computed tomography (Fig. 2B) showed an expansive bulk on the left kidney of approximately 9 cm that extended from the renal sinus with an exophytic growth into the anterior perinephric space. The mass showed a fluid density and presented multiple septal structures characterized by contrast enhancement.

Suspecting a Bosniak type III cyst (Fig. 2B), we first attempted a cyst excision by laparotomy with a 22-minute warm ischemia time. However, the intraoperative histologic examination showed XGP; therefore, we performed a radical nephrectomy.

The histologic examination (Fig. 3) showed chronic pyelonephritis with xanthogranulomatous needle-like (Fig. 2A) deposits of cholesterol and macrocytic chronic hydronephrosis of the renal pelvis with intracystic hemorrhage.

Discussion

XGP is a rare atypical form of chronic pyelonephritis that is characterized by destruction of the renal parenchyma, which is replaced by granulomatous tissue containing lipid-laden macrophages.

Ultrasonography is the recommended first step for diagnosis and may differentiate between the 2 forms of XGP. In the diffuse form, imaging may show a generalized renal enlargement with multiple hypoechoic areas representing calyceal or pelvocalyceal dilatation and parenchymal destruction, hyperechoic foci with clean posterior acoustic shadowing representing renal calculi or a staghorn stone, and debris in the hydronephrosis. The focal form of XGP is usually confined to 1 part or pole of the kidney and therefore may not present findings similar to those of the diffuse form.⁴

Computer tomography (CT), which is considered the imaging modality of choice, reveals the XGP lesion, but differential diagnosis from hydronephrosis or pyonephrosis, malakoplakia, lymphoma, and especially renal neoplasms is sometimes difficult. CT features that have been considered characteristic of (but not pathognomonic of) XGP (especially in the diffuse form) are renal enlargement, strands in the perinephric fat, thickening of the Gerota fascia, and thick enhancing septa in the hypodense areas of the renal parenchyma. Round or egg-shaped areas of water density representing dilated calyces and abscess cavities with pus and debris in diffuse XGP may be described as the “bear paw sign”.⁵

CT usually depicts focal XGP as a clearly or poorly defined localized intrarenal mass with fluid-like attenuation.

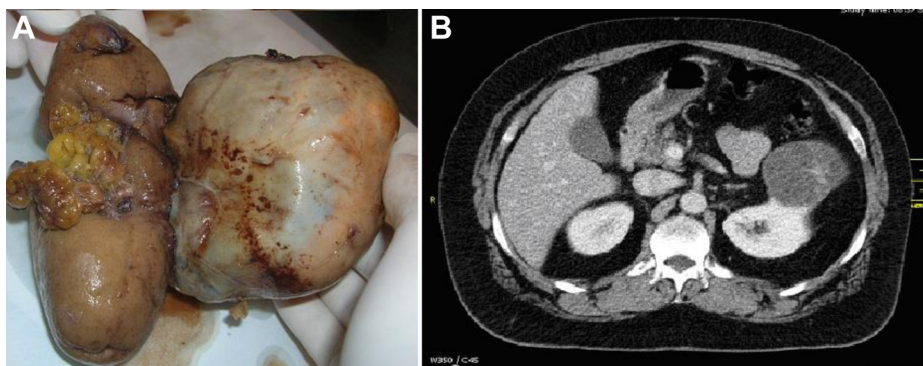


Figure 2. (A) Kidney and complex cyst, macroscopic appearances. (B) Contrast-enhanced nephrographic phase axial computer tomography (CT) image. Enhancement of irregular, thick intracystic septa is well appreciated.

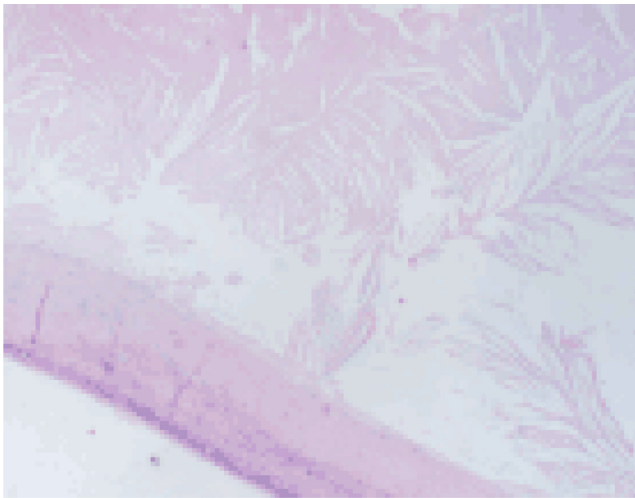


Figure 3. Cholesterol clefts within the cyst in renal parenchyma (hematoxylin and eosin stain; original magnification, $\times 100$).

In our case, the radiologic examinations did not assist with the diagnosis; all of the pathognomonic aspects were absent, and all of the images indicated a complex cyst. We assume that the XGP was initially triggered in the middle third of the kidney, creating the conditions for cyst formation, and, later, the inflammation involved the entire renal parenchyma.

Our case is unusual in its presentation; the patient had no history of kidney stones, and symptoms were absent or scarcely meaningful to suspect inflammation of the kidney. The intra-operative histologic examination identified the condition and enabled appropriate treatment.

Conclusion

Our experience suggests the opportunity of a simple intra-operative histological examination in all cases of complex cyst, otherwise the risk would be an under-treatment.

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