Acta Radiologica Short Reports
3(1) 1–4

DOI: 10.1177/2047981613513763
arr.sagepub.com



# Xanthogranulomatous pyelonephritis mimicking a renal cell carcinoma: a unique and challenging case

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## Abstract

We describe an unusual case of xanthogranulomatous pyelonephritis (XGPN) in a 73-year-old woman diagnosed after a blunt abdominal trauma. This case is unique because of the atypical presentation, with absence of symptoms, normal laboratory exams, and unusual computed tomography and magnetic resonance imaging findings. The patient underwent radical nephrectomy because a renal cystic tumor was suspected. Only the histopathological findings suggested the final diagnosis of XGPN.

**Keywords**

Computed tomography (CT), magnetic resonance imaging (MRI), ultrasound, kidney, pyelonephritis, urinary

Date received: 24 September 2013; accepted: 31 October 2013

## Introduction

Xanthogranulomatous pyelonephritis (XGPN) is an atypical form of chronic pyelonephritis characterized by the destruction of the renal parenchyma and replacement with a chronic inflammatory infiltrate of lipid-laden macrophages, known as xanthoma cells (1,2). XGPN is usually classified in diffuse and focal forms, with the diffuse form accounting for >90% of cases (3,4). It usually affects middle-aged women and is extremely uncommon in children (5,6), accounting for 0.6% of histologically documented cases of chronic pyelonephritis (5–8). Its exact etiology remains unknown. However, it usually occurs in association with nephrolithiasis, urinary tract obstruction, and/or chronic urinary infection, with common pathogens such as *Proteus mirabilis*, *Escherichia coli*, *Pseudomonas*, *Klebsiella*, and *Staphylococcus* (9). This disease has been called as the ''great imitator'' because the clinical and radiological findings closely resemble other pathological entities such as renal cell carcinoma (10). The preoperative distinction between XPGN and malignant kidney tumors is often difficult.

We report an unusual case of a 73-year-old woman presenting with asymptomatic right renal mass, incidentally discovered in a post-traumatic screening ultrasound. Right open radical nephrectomy was performed and the final histopathologic examination, despite the absence of symptomatology (lumbar pain, fever, anorexia, and weight loss) was consistent with the diagnosis of XGPN.

## Case report

A 73-year-old woman was admitted to our hospital after a blunt abdominal trauma. On admission the patient was afebrile and in stable clinical condition.

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On clinical examination the patient only complained of urinary frequency and nycturia. Complete blood count was unremarkable: red blood cell count, 4,360,000 cells/$mm^3$; white blood cell count, 7040 cells/$mm^3$ of which 72.8% neutrophils, 18.5% lymphocytes, and 6.0% monocytes; platelets, 308,000 cells/$mm^3$. The inflammatory markers were normal. An ultrasound (US) examination of the abdomen showed a voluminous cystic mass in the lower pole of the right kidney, with associated severe hydronephrosis. She was subsequently hospitalized for further investigation of the case. To better evaluate the US findings, a computed tomography (CT) scan of the abdomen was performed, after the intravenous bolus administration of iodinated non-ionic contrast agent. The CT images showed severe hydronephrosis of the right kidney, with marked thinning of the cortical parenchyma and the presence at the lower pole, of a voluminous cystic mass of 9.7 cm, which had non-uniform wall thickening, with heterogeneous contrast enhancement (Fig. 1a and b). Subsequently, a magnetic resonance (MR) scan of the abdomen was performed, using a 1.5 T scanner and confirmed the presence at the lower pole of the right kidney of a voluminous expansive formation of 9.7 cm; this mass appeared heterogeneously hyperintense in T1 and T2 and showed only a slight contrast enhancement in its peripheral and cranial portion (Fig. 2a and b), compatible with a complex cystic mass, with predominantly hemorrhagic content (Bosniack IIF-III). The patient underwent surgery for radical nephrectomy with open access that showed a friable yellow lesion, partially cystic (Fig. 3); it was difficult to dissociate the mass from adjacent organs because of many adhesions. Microscopic examination showed a partially cystic lesion, with marked hemorrhagic components and an intense xanthogranulomatous inflammatory reaction, in the context of which there were nests of kidney ducts that resulted positive for cytokeratin immunohistochemistry survey-pool with associated inflammatory cellular atypia.

## Discussion

XGPN is a severe form of chronic pyelonephritis, characterized by the destruction of the renal parenchyma and replacement by granulomatous tissue. Its name is derived from yellow color on gross pathology and a granulomatous reaction histologically. XGPN is

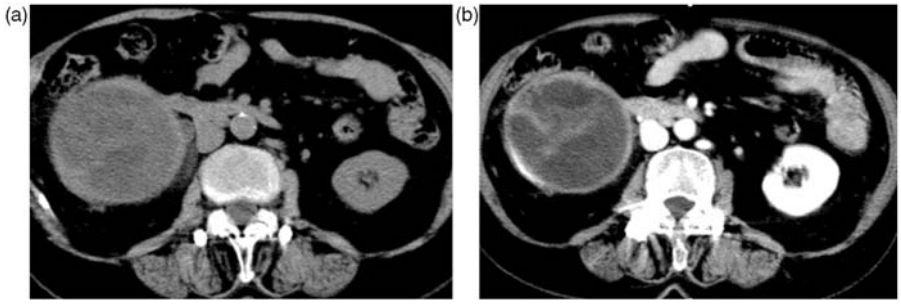


**Fig. 1.** CT scan of abdomen (a) before and (b) after intravenous iodine contrast agent administration, showing a voluminous cystic mass of 9.7 cm, with non-uniform wall thickening and heterogeneous contrast enhancement, at the lower pole of the right kidney.

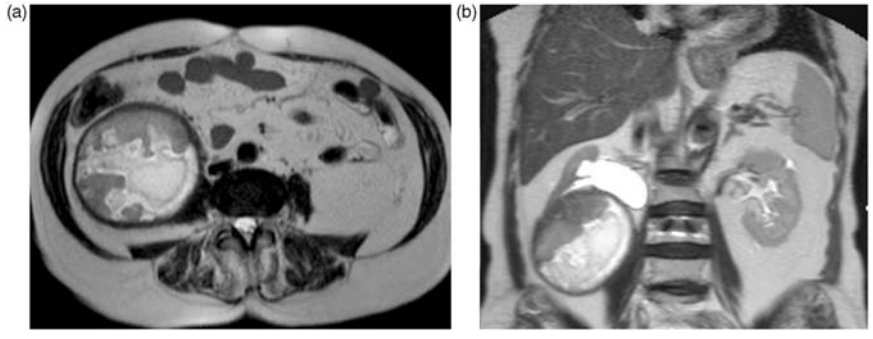


**Fig. 2.** (a) Axial and (b) coronal T2W MR image of the abdomen showing a 9.7 cm rounded mass, with thick wall, in the right lower kidney. The mass shows inhomogeneous content with a mix component both cystic and solid, more pronounced in its cranial and peripheral portion.

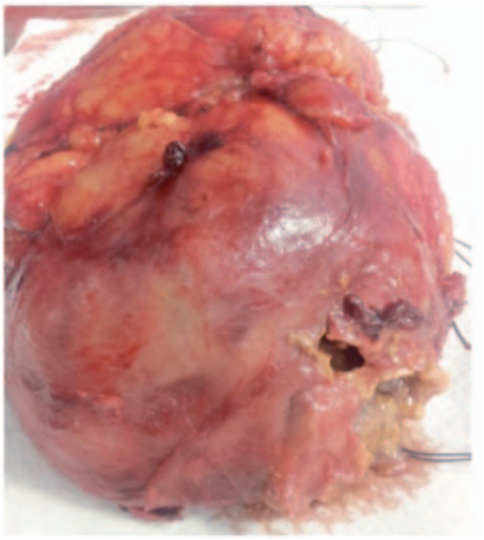

**Fig. 3.** Surgical excised right kidney.

usually divided into three stages: stage I (nephric XGPN), the inflammation is confined to the kidney; stage II (perinephric XGPN), the inflammation involves both the kidney and peri-renal fat; stage III (paranephric XGPN), the inflammation involves the kidney, peri-renal fat, and the retroperitoneum (11). The etiology is still unclear, but appears to be multifactorial. It is clearly related to a combination of renal obstruction and chronic bacterial infection and usually occurs in association with nephrolithiasis, urinary tract obstruction, and chronic urinary infection (12,13). Other causes of obstruction include congenital abnormalities such as uretero-pelvic junction obstruction and tumors that occur mainly in the adult population (renal cell carcinoma, ureteral carcinoma, bladder carcinoma) (11,14). Other factors implicated in the etiology of XGPN include altered immune response and intrinsic disturbance of leukocyte function, alterations in lipid metabolism, lymphatic obstruction, malnutrition, arterial insufficiency, venous occlusion and hemorrhage, and necrosis of the pericalyceal fat (3,9,11,14,15). The most commonly reported symptoms are fever, abdominal and/or flank pain, weight loss, malaise, anorexia, and lower urinary tract symptoms. Pyuria is present in 60–90% of patients. Common findings at physical examination are a palpable mass and flank tenderness. Rarely, in 5% of patients, a draining renal cutaneous fistula in the flank may be present (11,12). Laboratory tests include leukocytosis, anemia, and increased elevated sedimentation rate in the majority of patients. Urine cultures are usually positive at the time of diagnoses. The most common pathogens are *Escherichia coli*, *Proteus mirabilis*, and rarely *Staphylococcus aureus*, *Pseudomonas*, and *Klebsiella*. Although the urine cultures may be negative, cultures of renal tissue at surgery are often positive for these pathogens. The US pattern of XGPN corresponds to that of a solid mass with inhomogeneous echoes. US can show enlargement of the entire kidney with multiple hypoechoic areas representing hydronephrosis and/or calyceal dilatation with parenchymal destruction, as well as calculi. US may also help to differentiate the two forms of XPGN as focal and diffuse: in the diffuse form, generalized renal enlargement with multiple hypoechoic areas representing calyceal dilatation and parenchymal destruction is seen; in the focal form, a localized hypoechoic mass, often misdiagnosed as renal tumor, may be found (11–13). CT scan has been shown as one of the best preoperative diagnostic tests for the evaluation and confirmation of XGPN. Features that have been considered characteristic (but not pathognomonic) for diffuse XGPN are renal enlargement, perinephric fat strand, thickening of Gerota's fascia, and water density rounded areas in renal parenchyma representing dilated calyces and abscess cavities with pus and debris, described as "bear paw sign". CT may also reveal an obstructing urinary stone (mostly they are staghorn calculus) in the renal collecting system and absence of excretion of contrast medium, showing loss of function of the affected kidney, in 80% of patients. There may also be enlargement of the hilar and para-aortic lymph nodes. In the focal form, CT usually shows a well-defined localized intra-renal mass with fluid-like attenuation (11–14).

Several reports have described a possible role of MR in the diagnostic evaluation of patients with suspicious XGPN; in particular, Cakmakci et al. (12) have shown that in the focal form of XGPN the mass has slightly low signal intensity on T2-weighted (T2W) images and is isointense with the renal parenchyma on T1-weighted (T1W) images. These findings suggest a fluid with very high protein content. The different signal intensity of the solid component of XGPN on T1W images, compared with the renal parenchyma, depends on the amount of xanthoma cells involved in the granulomatous process. The T2W sequences are very useful for accurate differentiation between XGPN from tumors. Although MR imaging (MRI) is inferior to CT in demonstrating renal calcifications and ureteral stones, contrast-enhanced MRI can easily demonstrate infiltration of the inflammatory mass into adjacent tissue structures and better demonstrates the anatomical relationship of the XGPN on coronal and sagittal planes, as well as the fat component within the mass and the compressed renal parenchyma (13).

The differential diagnosis of XGPN include neoplastic diseases such as clear-cell carcinoma, lymphoma, leukemia, Wilms' tumor, neuroblastoma, and inflammatory processes (renal or peri-renal abscess,

pyonephrosis, renal tuberculosis, focal and diffuse nephritis, and fungal infection) (11,12).

The treatment of choice for diffuse XGPN, which is the most frequent form, is surgery and consists of nephrectomy with resection of all other involved tissues, with or without antibiotic therapy. Drainage of perirenal or renal abscess with adjunctive antibiotic treatment is strongly recommended before definitive surgery, to decrease the complications in the diffuse form of the disease. In the localized form of the disease, segmental resection of the affected kidney is effective. Partial nephrectomy is also recommended in extremely rare bilateral cases (11–16).

Macroscopic appearance of XGPN include an enlarged kidney with a thickened capsule, yellow nodules with or without central necrosis in the renal parenchyma, while the renal pelvis may be dilated and filled with stones, debris, or purulent fluid. Microscopic pathological examination of the yellow areas shows a large number of lipid-laden macrophages (foam cells) with extensive areas of inflammation and fibrosis (11,12). Misinterpretation of "foam cells" as "clear cells" consistent with renal adenocarcinoma, is the most important diagnostic challenge at histology.

In conclusion, the unusual findings of this case report suggest a careful evaluation of patients with a renal cystic mass, especially in case after blunt abdominal trauma, that can be misdiagnosed with a renal cell tumor. A combined CT and MR evaluation together with laboratory and clinical findings are mandatory for a correct differential diagnosis of this rare renal entity.

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