



## Case report

## Xanthogranulomatous pyelonephritis in an eight year old male child: A case report and review of the literature

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## ARTICLE INFO

## Keywords:

Xanthogranulomatous pyelonephritis

Nephrectomy

Case report

## ABSTRACT

**Introduction and importance:** Xanthogranulomatous pyelonephritis is an extremely rare but known form of chronic pyelonephritis resulting from prolonged suppuration of the kidney. Pre-operatively, it may mimic renal tuberculosis or neoplastic lesions including renal cell carcinoma due to its vague clinical presentation, equivocal laboratory and radiological investigations. Due to its rarity and academic interest, herein we report such a rare case we recently encountered in our clinical practice.

**Case presentation:** An-eight-year old male child patient resented to our hospital with three months history of abdominal distension associated with progressive left flank pain. Preoperative investigations including CT-scan were suggestive of nephroblastoma with differential diagnosis of clear cell sarcoma. Radical nephrectomy was performed and histopathology of the specimen confirmed the diagnosis of Xanthogranulomatous pyelonephritis. The patient fared well postoperatively and he had no symptoms in the subsequent follow up visits.

**Clinical discussion:** Xanthogranulomatous pyelonephritis is a rare, severe and atypical form of chronic pyelonephritis due to infection (*E. coli*, *Proteus*) or stones. The disease may resemble renal cell carcinoma preoperatively. Thus, high index of suspicion is necessary for preoperative diagnosis.

**Conclusion:** Preoperative diagnosis of Xanthogranulomatous pyelonephritis may be a daunting task related to the rarity of its presentation. Thus, scrupulous histopathological evaluation is essential for the definitive diagnosis. Radical nephrectomy is the mainstay treatment of choice especially in diffuse cases.

## 1. Introduction

Xanthogranulomatous pyelonephritis (XP) is a rare and severe variant of chronic pyelonephritis constituting less than 1% of chronic pyelonephritis [1]. It is frequently associated with urinary tract obstruction or nephrolithiasis [2]. Pre-operatively, it may mimic renal tuberculosis or renal carcinoma due to its vague clinical presentation, equivocal laboratory and radiological investigations [1–3]. The confirmatory diagnosis of this entity is based on histopathological examination and surgery remains the treatment of choice in almost all cases. We report a case of XP occurring in an eight year old male child with albinism. This work has been reported in line with the SCARE 2020 criteria

[4].

## 2. Presentation of case

8-year-old African male child presented to our facility with three months history of malaise, progressive left flank pain and painful micturition. The flank pain had gradual onset, dull in nature and was radiating to the back. The informant denied history of fever, loss of appetite, vomiting, diarrhea or constipation. She further denied history of allergy to drugs or food. The boy had history of kidney stones and received treatment for urinary tract infection several times with antibiotics. The boy was a third born in six children peasant family with

**Abbreviations:** ALP, alkaline phosphatase (ALP); ALT, alanine aminotransferase; BP, blood pressure; CT scan, computerized tomography scan; ESR, erythrocyte sedimentation rate; IVC, inferior vena cava; H&E, hematoxylin and eosin staining; HPF, high power field; KUB USS, kidneys, ureters & bladder ultrasound; PR, pulse rate; SO<sub>2</sub>, oxygen saturation; WBC, white blood cell; XP, xanthogranulomatous pyelonephritis.

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<https://doi.org/10.1016/j.ijscr.2021.105943>

Received 2 April 2021; Received in revised form 26 April 2021; Accepted 26 April 2021

Available online 30 April 2021

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history of albinism.

On physical examination, there was asymmetrical lower abdominal distension more on the left side which was associated with tenderness on percussion. Vital signs were:- Blood Pressure-106/69 mmHg, Pulse Rate = 98 beats per minute,  $SO_2 = 96\%$ , temperature: 37.8 °C. The patient had skin albinism; he was alert, non-dyspneic, non-pallor, non-jaundiced, non-cyanosed, without palpable lymph nodes. Review of other systems was essentially un-remarkable except for poor vision related to albinism.

Laboratory tests highlighted hemoglobin of 8.4 g/dL (11-15 g/dL), raised ESR of 28 mm/h (0-19 mm/1 h), normal leukocytes count  $6.13 \times 10^9/L$  (4.00–11.00)/L, Serum creatinine 94 (62-106  $\mu\text{mol/L}$ ), AST 40.60u/L (2.00–40.00 U/L), ALT 11 U/L (2.00–41.00 U/L). Urinalysis revealed 10–15 WBC/HPF, however there was no bacterial growth in urine culture and sensitivity test. Other urinalysis parameters were essentially normal. Chest x-ray was normal. Kidneys, Ureters & Bladder Ultrasound (KUB USS) showed enlarged and tumorous left kidney measuring about  $13 \times 9$  cm; suggestive of Nephroblastoma.

CT-scan of the abdomen (Fig. 1) demonstrated a complex mass in the left kidney with a hypo-density fluid component with peripheral hyperdense calcifications. The mass was completely enclosed by the renal capsule. The right kidney was essentially normal. Multiple retroperitoneal lymphadenopathy was visualized. Spleen appeared enlarged with however normal attenuation and homogeneous enhancement. The rest of the abdomen and visualized part of the chest were unremarkable. The CT-scan was suggestive of clear cell sarcoma of the left kidney with calcification with differential diagnoses of (1) congenital mesoblasticnephroma and (2) cystic nephroblastoma.

With the pre-operative diagnosis of malignant neoplasm of the kidney, the patient was scheduled for the left radical nephrectomy by a team of specialist urologists. The patient underwent exploration through Chevron incision. Intra-operatively, an enlarged left kidney with both solid and cystic components was found with multiple enlarged para-aortic lymph nodes. However, liver, spleen and mesentery were normal. Left nephrectomy and para aortic lymph nodes resection were



**Fig. 1.** CT-scan of the abdomen demonstrating a complex mass in the left kidney with a hypo-density fluid component and peripheral hyper-dense calcifications.

performed and all were submitted for histopathological analysis. The patient was kept on ceftriaxone, metrodizole, pethidine non-steroid anti-inflammatory drugs and intravenous fluids. The patient fared well post-operatively and seven days later he was discharged. Subsequent follow up visits were uneventful and about two months later; the boy was completely recovered and resumed his previous normal life.

Gross examination of the nephrectomy specimen revealed enlarged and irregular kidney measuring  $12 \times 10 \times 9$  cm. Cut section revealed multiple yellow nodules around calyces, (Fig. 2). Areas with solid mass with infiltrative appearance were associated. Histopathology of sections from the specimen highlighted the replacement of renal parenchyma with foamy histiocytes, presence of multinucleated giant cells and mixed inflammatory cells (Fig. 3A–B). Diagnosis of XP was arrived.

### 3. Discussion

Xanthogranulomatous pyelonephritis (XP) is a rare entity that constitutes less than 1% of chronic pyelonephritis. It is a severe and atypical form of chronic inflammatory condition of the kidney that is increasingly well recognized [1]. The disease process is characterized by the destruction and replacement of renal parenchyma by lipid-laden macrophages (xanthoma cells), [1–3]. Like it was the case in our patient, XP is generally associated with urinary tract infection and obstructing renal calculi, leading to diffuse or focal kidney destruction [3]. It starts within the pelvis and calyces and subsequently spreads into renal parenchyma; if uncontrolled; it spreads to adjacent tissue and destroys it [5–8].

XP mainly occurs in adults with preponderance in females of younger age group [8]. It is usually associated with renal stones in nearly 2/3 of cases and concomitant infection with *E. coli*, *Proteus mirabilis*, *Klebsiella spp*, *Staphylococcus aureus*, *Enterococcus spp*, *Pseudomonas spp*, *Streptococcus spp*, including anaerobic organisms [2,10]. Bilateral lesions although rare, have a fatal outcome [8]. Usually, the disease process affects the whole the kidney; focal forms are rare [9]. The former is often misdiagnosed as a life-threatening abscess, while the latter is sometimes referred to as the tumor-like form of XP because the clinical findings are easily confused with those of a renal tumor [6–8].

Patients with XP may present with various non-specific symptoms which may include fever, flank pain, and weight loss, malaise, anorexia and constipation. Other uncommon presentations include urinary symptoms like dysuria, frequent urination and hematuria or rarely discharging cutaneous fistula originating at the level of involved kidney [6]. Physical examination usually reveals an ill-defined, palpable and tender flank mass. However, some patients may present with hypertension or hepatomegaly [3,7]. Our patient had relatively fewer symptoms. Previous studies have reported leukocytosis and elevated C-reactive protein, erythrocyte sedimentation rate, or liver enzyme [6–8].

None of the laboratory or radiological investigations is confirmatory and diseases like renal tuberculosis and renal cell carcinoma may mimic XP. CT-scan has been considered to be the most useful imaging modality for identifying XP [9]. The radiologic findings may include an enlarged kidney with poor or absent function and with simple or staghorn calculi [7]. The renal parenchyma may be replaced by multiple hypodense, egg-shaped areas representing dilated calices and abscess cavities filled with debris and pus. With the high predilection of spread to contiguous tissue, especially to the peri- and paranephric spaces, psoas muscle, spleen, and formation of nephro-colonic and nephrocutaneous fistulae. Thus, CT-scan is essential in evaluation of disease extension [10]. In our case, preoperatively we suspected malignant renal tumor as it was suggested by the CT-scan. The definitive diagnosis was made post-operatively based on histopathology analysis.

Although the exact pathogenesis of XP is still not known, a number of predisposing factors have been implicated. These include recurrent urinary tract infection, genitourinary obstruction, and nephrolithiasis [7]. Previous studies have also reported XP patients with diabetes mellitus, transplanted kidney receiving hemodialysis for chronic renal failure [7]. Positive urine culture was reported in some patients [10]. In



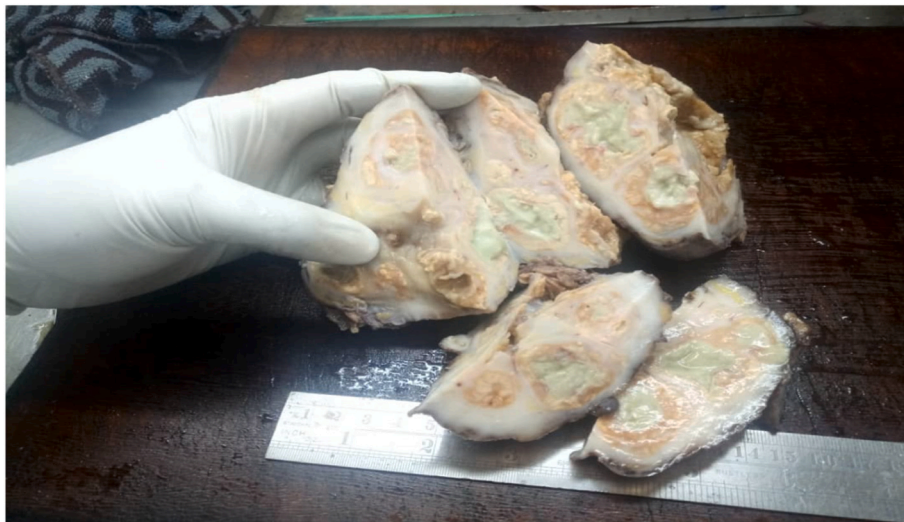


Fig. 2. Cut section of the nephrectomy specimen highlighting multiple yellow nodules around calyces.

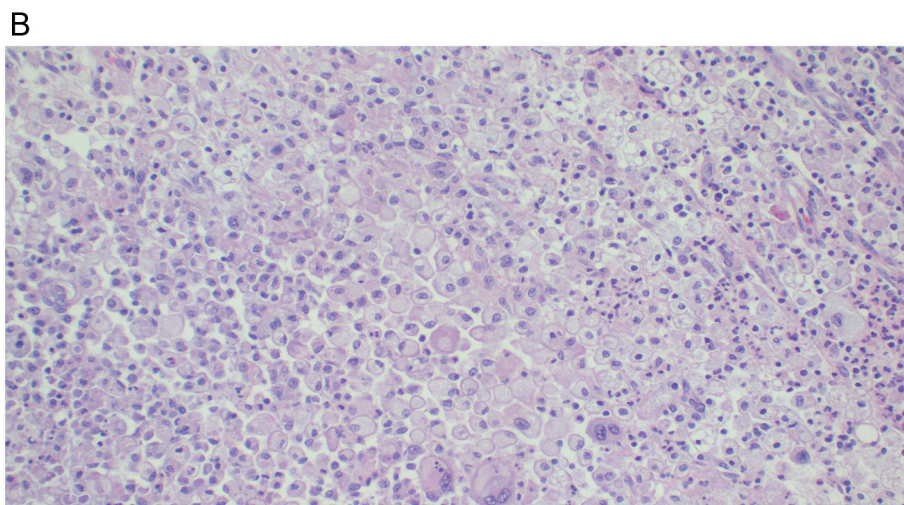
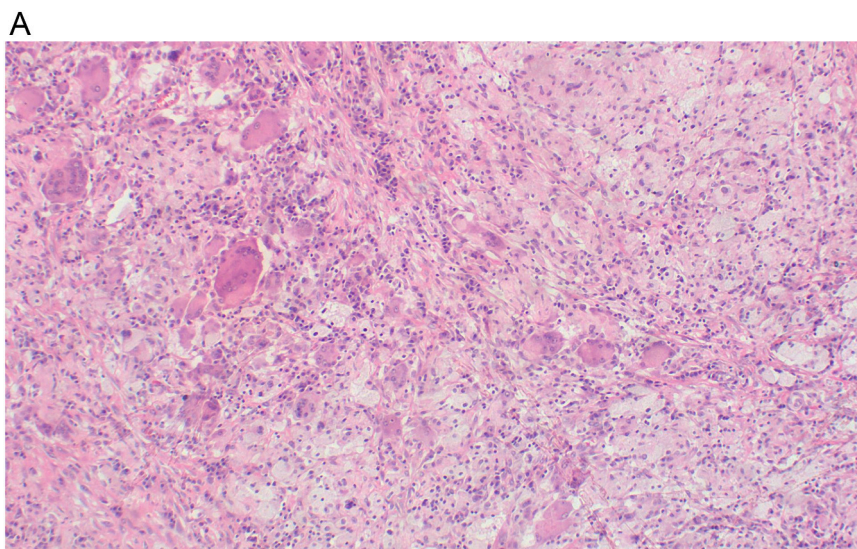


Fig. 3. A: Histopathology of the kidney specimen highlighting replacement of renal parenchyma with foamy histiocytes, occasional multinucleated giant cells and inflammatory cells; H&E staining 100× original magnification.

B: Photomicroscopy of XP demonstrating the presence of numerous foamy histiocytes; H&E staining 200× original magnifications.

our case, there was no bacterial growth in urine probably because of previous history of antibiotics use.

The disease is generally limited to the affected kidney, but spread to adjacent tissues has also been seen [10]. According to the extent of involvement of the adjacent tissue, Malek and Elder have classified this disease into three stages. Stage I: nephric, disease is confined to renal parenchyma only. Stage II: nephric and perinephric, disease process involves renal parenchyma along with perinephric fat. Stage III: nephric and perinephric, disease is extending into adjacent structure or diffuse entropertoneum. In stage III, psoas abscess and the formation of nephrocolonic, nephrocutaneous fistulae have been described [10]. Our patient had stage III as he had also para aortic lymph node enlargement and inflamed surrounding tissues.

Early diagnosis and prompt treatment play a crucial role in minimizing the morbidity and mortality rates from XP. The definitive diagnosis of XP is achieved only by histopathological examination which demonstrates the presence of xanthoma cells, as well as other inflammatory cells, including plasma cells, leukocytes, and histiocyte [5,6]. Antibiotics usually do not resolve the problem thus; nephrectomy remains the treatment of choice [8]. Nephron-sparing surgery is useful in localized XP, but diffuse XP will require radical nephrectomy with resection of all other involved tissue in most cases [11].

#### 4. Conclusion

XP is a rare cause of chronic pyelonephritis and preoperative diagnosis may be challenging related to the rarity of its presentation; thus emphasizing the importance of histopathology evaluation. Radical nephrectomy is the mainstay treatment of choice especially in diffuse cases.

#### Sources of funding

The work did not receive fund from any source.

#### Ethics approval

There was exemption of ethical clearance.

#### Consent

Written informed consent was obtained from the patient's legal guardian for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

#### Authors' contributions

AM conceptualized the study and prepared the first manuscript. OJM, BNN and FB reviewed the patients' medical records; planned and

performed surgery. AM and AP performed histopathological analysis and prepared microscopic images. All authors read and approved the final manuscript.

#### Research registration

N/A

#### Guarantor

Dr. Alex Mremi is the Guarantor of this work.

#### Provenance and peer review

Not commissioned, externally peer-reviewed.

#### Declaration of competing interest

All authors have declared that no competing interests exist.

#### Acknowledgements

The authors would like to thank the patient's guardian for allowing us to use the patient's medical information for academic purposes.

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