

Xanthogranulomatous Pyelonephritis: A case Report

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Abstract

Xanthogranulomatous Pyelonephritis (XPN) that was firstly described by Schlagenhauser in 1916, is a rare chronic pyelonephritis. We describe an interesting case of XPN in which patient was treated only with antibiotics.

An 80-year-old woman was hospitalized for fever and asthenia. No significant symptoms or signs were present at the admission. First instance ultrasound scan suggested the diagnosis of XPN, promptly confirmed by CT. Antibiotic therapy was successfully administered after urine analysis and culture.

Introduction

Xanthogranulomatous Pyelonephritis was firstly described by Schlagenhauser [1] in 1916. In recent decades it has been more described [2-3] also thanks to a deeper disease knowledge and a better performance of diagnostic imaging.

XPN is a rare chronic pyelonephritis that affects both men and women, even though women are more often affected and children are not spared [3]. It is almost always unilateral and its common feature is the kidney destruction. Moreover, it can simulate a neoplastic disease (that's why it's called "The Great Imitator") [4] or a specific inflammation.

Affected subjects have a history of recurrent urinary infections, associated with urinary tract abnormalities such as lithiasis, dilation of renal calyces and thinning of the renal cortical portion and vesicoureteral reflux. In addition, kidney is enlarged and perirenal fibrosis and adhesion to adjacent retroperitoneal structures are frequent.

Symptoms are often non-specific. Some patients complain fever, abdominal or flank pain, malaise or anorexia. In XPN, it is usually possible to palpate a one-sided mass.

Urinalysis and urine culture can give us additional informations. The most common pathogens are *Proteus mirabilis* and *Escherichia coli*, reported in 50%-75% of cases [5-7] but it is also possible to find *Klebsiella* species, *Pseudomonas* species, and gram-positive cocci⁹ (especially *Staphylococcus aureus*). In one series, 33% of patients exhibited mixed infections [7].

Many cases of XPN have been described with sterile urine but cultures of blood and/or renal tissue were positive. It has been suggested that sterile urine cultures may be the result of previous unsuccessful antibiotic therapy or, perhaps, spontaneous elimination of pathogens from the urine but not from the renal parenchyma [7]. Urinary sediment is usually sparse, but kidney epithelial cells, granular casts, and sometimes leukocytes may be present. Proteinuria is almost always found. From a histological point of view, giant cells, lipid-laden macrophages and cholesterol accumulations are often described [8].

Diagnostic imaging, ultrasounds, CT computed tomography and MRI (magnetic resonance) are fundamental for diagnosis [2,9] and characterization of the lesion. Therapy is usually based on antibiotics [2]; in the most severe cases total nephrectomy may be necessary, aiming to remove the compromised tissue, while a less aggressive approach may be considered in some cases for the focal form.

Case report

We describe an interesting case of XPN in which patient was treated only with antibiotics. There are only a few cases of XPN that have been successfully treated with medical therapy alone. [6]

In January 2021 an 80-year-old woman was hospitalized at the Nephrology and Dialysis Unit of the "P. Giaccone" University Hospital in Palermo. She had fever (38° C). Her mean blood pressure was about 130/80 mmHg, HR 75 bpm. She also complained asthenia and general malaise. She referred no lower back pain; none worthy of mentioning sign was found on the general physical examination, nor mass being appreciated on deep palpation of the abdomen.

The patient had a history of end-stage chronic kidney disease in hemodialysis treatment since 2019, singular nephrectomy for previous left renal lithiasis. She also referred a personal history of arterial hypertension, type 2 diabetes mellitus complicated by multi-district atherosclerosis and recurrent urinary tract infections (UTI).

During the hospitalization, first-instance ultrasound scanning was performed: it showed enlarged kidney size, with dilated calyces, diffuse thinning of the cortex and mold stones associated with pyelic dilatation thus suggesting a compatible picture of xantogranulomatous pyelonephritis. (see figure 1)

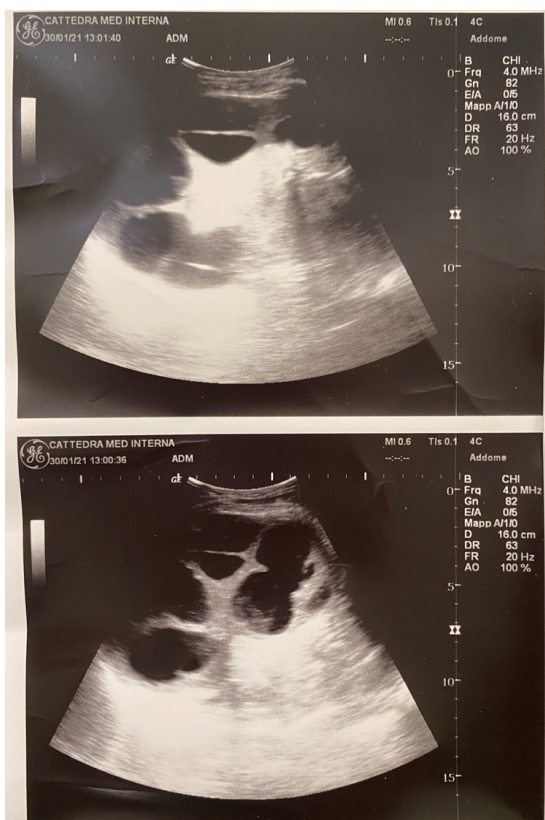


Figure 1: Left kidney in place, of increased size (DL > 13cm). Hypo-represented parenchyma with widespread thinning of the cortical area associated with conspicuous dilation of the pyramids. Presence of mold lithiasis and slight ureteropyelic dilatation.

She also underwent to chest-abdomen CT which confirmed and strenghtened the renal picture showing diffuse calyceal dilatation with cortical thinning and multiple multilocular cystic formations (fig. 2) and in the chest there was also a specific bronchopneumonic focus.

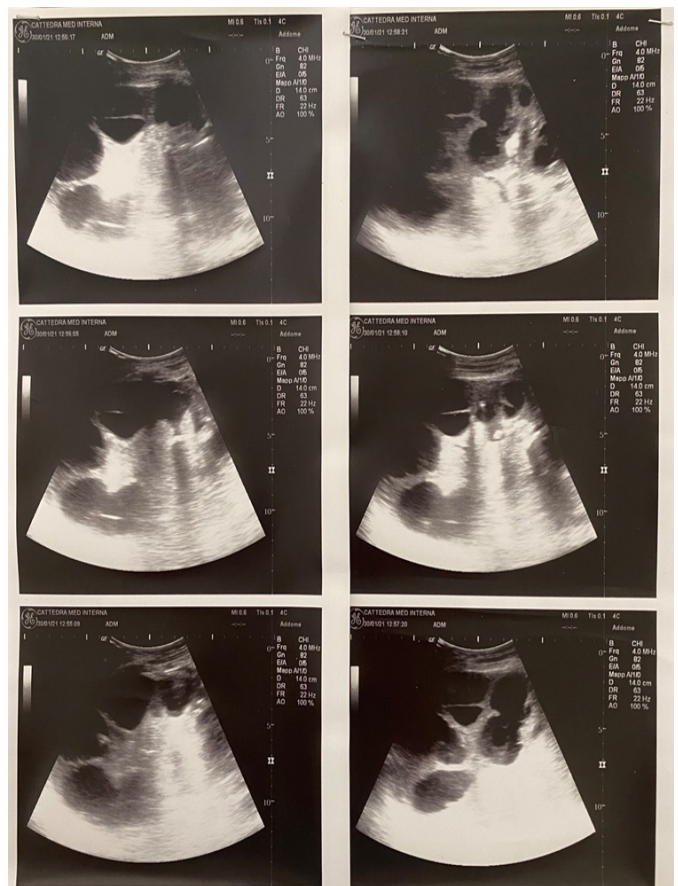


Figure 2: Mold lithiasis with enlargement of the left renal parenchyma, which has liquid density, thinning of the cortex to be referred in the first hypothesis to left xantugranulomatous pyelonephritis.

Blood tests showed a WBC count of 14820/mmc (neutrophils 88.3%), Hb of 9.6 g /dl, a C reactive protein (CRP) of 168.34 and a procalcitonin 61.5 ug /L. The chemical-physical examination showed: urine with a cloudy appearance and straw yellow color, proteins (200 mg /dl), hemoglobin 0.2 mg /dl, leukocyte esterase 500 Leu/uL with absent nitrites and urinary sediment carpet of leukocytes and red blood cells. We also performed urine culture (fig.3) and antibiogram which tested positive for Klebsiella Pneumoniae 100,000 CFU / ml.

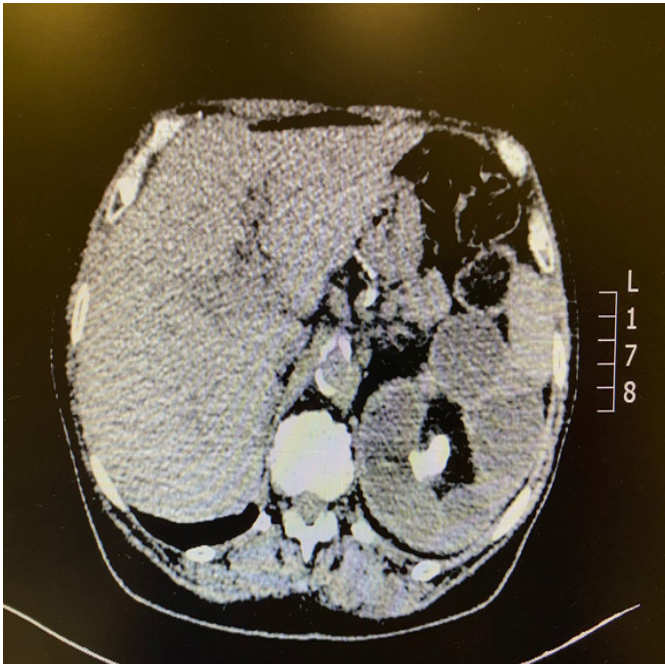


Fig. 3. Urine culture with an antibiogram

Although renal biopsy should be made to confirm the diagnosis, we chose to avoid such an invasive exam due to the age and frailty of our patient so that we decided to have a conservative approach.

She was treated with intravenous therapy with meropenem. After 7 days, we observed an improvement in clinical symptoms and a full remission of the inflammation indexes.

Discussion

XPN is a rare disease. Imaging plays a fundamental role in diagnosis and thanks to advances in this field, there has been a greater incidence of diagnosed cases.

The patient we wrote about referred non specific symptoms, as described by Leoni et al [10] in a case series. Her Hb levels were reduced, as observed by Dwivedi [11] in his experience.

Ultrasonounds give us a non invasive way to confirm, as a first instance, the presence or absence of a kidney. Ultrasonic visualization of an abnormal kidney allows to evaluate renal size, shape, and parenchymal homogeneity as well as assessment of focal versus diffuse intra- renal disease [12]. Finally, an obstructing extrarenal lesion causing subsequent xanthogranulomatous pyelonephritis may also be considered and investigated.

Differential diagnosis with neoplastic or other kidney inflammatory diseases is also fundamental. For example distinction of xanthogranulomatous pyelonephritis from pyonephrosis could be very difficult. By means of

ultrasonounds, very large, irregularly defined cystic lesions have been found to suggest the diagnosis of pyonephrosis [12]. This is in contrast to the diffuse renal involvement and enlargement without change in parenchymal homogeneity seen in this patient. The increased incidence of pericalyceal or staghorn calculus in xanthogranulomatous pyelonephritis compared with hydronephrosis is significant. By means of ultrasonounds this is demonstrable as increased calyceal and pericalyceal echoes. Moreover, the ultrasonic picture of hydronephrosis usually consists of cysts within or radiating outward from the pelvis.

Recently Saadi [13] described a rare presentation of Xanthogranulomatous pyelonephritis appearing as a Bosniak type-3 hemorrhagic cystic mass at ultrasonounds and CT scanning. Only after surgical treatment and subsequent histological examination, diagnosis of XPN was confirmed.

Xanthogranulomatous pyelonephritis may simulate renal carcinoma. On ultrasound examination the relatively homogeneous, normal appearing parenchymal pattern differs from the frequently observed more localized pattern of altered homogeneity usually found in renal carcinoma.

Ultrasonounds can give us additional informations suggesting the diagnosis of xanthogranulomatous pyelonephritis in an often confusing clinical and radiographic setting thus permitting increased preoperative detection of this disease.

Nephrectomy, with or without adjunctive antibiotic therapy, has been the first choice treatment for XPN and is actually retained curative [9]. Hughes [14] described a case of focal XPN diagnosed by percutaneous biopsy, for which a 3-week course of netilmicin was curative Rasoulpour [7] reported a case of focal XPN diagnosed by surgical renal biopsy and successfully treated with iv cefazolin (administered for 2 weeks) followed by cephalexin (orally for 8 weeks). Surgical treatment, after antibiotic therapy, has been successfully chosen also by Alvaro and coworkers in their recent experience [15].

Preferring conservative medical therapy for patient with a good residual renal function is widely appreciated. It seems reasonable, on our opinion, starting antibiotic therapy in cases of focal XPN when a significant residual function in the affected kidney is confirmed, especially in children. Success of medical therapy could be easily evaluated. Surgical treatment should be considered if patient's conditions not improve or worsen.

Conclusion

XPN is a rare entity that could be confirmed only by pathology.

In this case, we avoided biopsy in an old, frail patient. Bearing in mind the informations given by ultrasounds and CT, we administered intravenous antibiotic therapy obtaining a full remission.

Statement of Ethics

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Conflict of Interest Statement

The authors have no conflicts of interest to declare.

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Author Contributions

All the listed authors equally contributed to the paper.

Data Availability Statement

Clinical data and exams of the patient are available.

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