AMERICAN JOURNAL OF MEDICAL AND CLINICAL SCIENCES, 2022
VOL 7, NO. 3, PAGE 1 – 4

CASE REPORT

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Xanthogranulomatous Pyelonephritis, a Great Imitator: Case Report

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ABSTRACT

Xanthogranulomatous pyelonephritis (XPN) is an atypical form of chronic pyelonephritis, characterized by the destruction of the renal parenchyma and its replacement with a chronic infiltrate of lipid-laden macrophages [1]. The clinical presentation is nonspecific and variable. Consider clear cell carcinoma, sarcomatoid renal cell carcinoma, renal tuberculosis, renal lymphoma, renal or psoas abscess, actinomycosis, renal cystic disease, and emphysematous pyelonephritis [2,3]. We present the case of a 59-year-old female patient, with a history of recurrent urinary tract infections, who came to our center due to severe anemia (5gr/dl), weight loss of 10 kg in 2 months, night sweats and weakness, concomitant sudden-onset lumbar pain and hematuria, after multiple studies, a diagnosis of PNX was made, which was confirmed by anatomopathological study.

ARTICLE HISTORY

Received 07 Jul 2022
Accepted 17 Aug 2022
Published 22 Aug 2022

KEYWORDS

Xanthogranulomatous Pyelonephritis, Urinary tract infections, Anemia.

Introduction

XPN was described by Schlagenhaufer in 1916 and by Putshar in 1934 who observed, in renal anatomopathological studies of patients with chronic pyelonephritis, the presence of foamy histiocytes in the inflammatory infiltrate that altered normal renal histology [4]. It is classified into 3 stages: stage I, the lesion is confined to the kidney; stage II, there is infiltration of Gerota's capsule; and stage III, extends to the perinephric space and retroperitoneal structures [1].

It generally presents in adults, being more common in young women [5] and is associated in 2/3 of cases with infected kidney stones; the most common pathogens are Proteus mirabillis, Escherichia coli, Pseudomonas, Klebsiella and Staphylococcus [1,6]. It is common for it to involve one kidney, although bilateral involvement is possible [5].

The clinical presentation is variable, occasionally it can cause a mass with tumor characteristics, which requires a differential diagnosis with different pathologies such as clear cell carcinoma, sarcomatoid renal cell carcinoma, renal tuberculosis, renal lymphoma, renal or psoas abscess, actinomycosis, cystic disease kidney disease and emphysematous pyelonephritis [2,3].

Computed Tomography (CT) is the imaging technique of choice since it allows determining the magnitude of renal parenchymal involvement, the degree of extrarenal extension, and its association with neoplasms [5].

Confirmation diagnosis is histological, characterized by poor corticomedullary differentiation on cut, at the microscopic level it shows an acute inflammatory infiltration with a predominance of polymorphonuclear cells with intracytoplasmic lipid-laden macrophages (foamy macrophages or xanthogranulomatous cells) and multinucleated giant cells, a characteristic histopathological image that name the entity [7,8].

Case Report

A 59-year-old female patient with a history of recurrent urinary tract infections came to our care center due to a 2-month evolution of 10kg weight loss, night sweats and weakness. On physical examination, the patient was febrile, tachycardic, with accentuated mucosal cutaneous pallor and orthostatic hypotension, abdominal palpation revealed the presence of a mass in the left flank of approximately 10x9 cm and positive left renal fist percussion.

Paraclinical examinations were performed at admission (Tables 1-5) with the presence of severe hypochromic microcytic anemia with a ferrokinetic profile consistent with anemia due to chronic disease, leukocytosis with a shift to the left, hypoalbuminemia, elevated serum acute phase reactants (C reactive protein, platelets and fibrinogen), liver abnormalities (elevated alkaline phosphatase, direct bilirubin, and AST), leukocyturia, and hematuria.

Abdomen and pelvis contrast-enhanced CT revealed the presence of a large 4 x 0.9 cm staghorn stone that obstructed

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the calyces and renal pelvis of the left kidney, as well as alteration of its renal architecture, cortical thinning and calyceal dilation (grade IV hydronephrosis). In the right kidney, two stones located at the level of the calyces, 1 x 1cm each with grade II hydronephrosis were found. The rest of the abdominal structures were without alterations (Figure 1).

Five units of globular concentrate were administered, reaching hemoglobin levels of 10 g/dl, and empirical antibiotic treatment with Meropenem 1 gram every 8 hours intravenously was started, pending the result of the urine culture, which subsequently turned out to be positive for Extended Spectrum Beta Lactamase producing Escherichia coli.

In view of the findings in the radiological studies and the poor response to antibiotic treatment, it was decided to place a double JJ catheter in the right kidney and a total nephrectomy of the left kidney, procedure was performed without complications (Figure 3 and 3.1).

The patient's postoperative course was in good in general with paraclinical improvement (Table 6). The anatomopathological report of the left kidney reported ''Distortion of normal renal architecture, with extensive predominantly acute inflammatory infiltrate, parenchymal fibrosis, necrosis, and glomerular atrophy extending to the capsule, presence of foamy macrophages in renal parenchyma'' considering the final diagnosis of clinical stage II XPN.

The patient complete antibiotic treatment for 21 days and in view of her clinical and paraclinical improvement she was discharged, currently she remains asymptomatic with periodic outpatient controls.

Table 1: Admission laboratories Blood count.

RBC	**2790000 cel/mm³ (4100000 - 5100000)**
WBC	8600 cel/mm³ (4400 - 10000)
Eosinophils	2%
Basophils	0%
Neutrophils	**81%**
Lymphocytes	10%
Monocytes	7%
Hemoglobin	**6.8 gr/dl** (12.3 – 15.3)
Hematocrit	**21,3%** (37 - 42)
MCV	**76.6 fl** (80 - 100)
MCH	**24,5 pg** (27 - 32)
MCHC	31,9% (32 - 37)
Platelet count	**402000 cel/mm³** (1770000 - 393000)

Table 2: Admission laboratories – Coagulation profile.

Fibrinogen	**463.3 mg/dl (200 - 400)**
TTP	26,9 seg (0.0 – 39.0)
PT	**15,7 seg (12.0 – 15.0)**
INR	**1,27 (0.8 – 1.20)**
Bleeding time	2 seg (0.00 - 5)

Table 3: Admission laboratories – Peripheral blood smear.

Red blood cells	**Decrease in its elements, with the presence of hypochromia +1, microcytosis 2+, anisocytosis 2+**
White blood cells	Elements within the established parameter, with normal morphology.
Platelet	Elements in slightly increased quantity, with normal morphology.

Table 4: Admision laboratories -Blood biochemistry.

Creatinine	**1,31 mg/dl (0.5 – 1.20)**
Lactic dehydrogenase (LDH)	86 U/L
Glucose	118 mg/dl
Urea	**58 mg/dl** (11 - 50)
BUN/Creatinine	**20,6**
T3	0.84 ng/ml (0.80 – 2.10)
T4	6.83 ug/dL (4.8 – 12.7)
TSH	3.64 uUI/ml (0.27 – 4.8)
Ferrokinetic profile:	
- Iron	94,1 ug/dL (37 - 145)
- Transferrin	**84 mg/L (200 - 364)**
- Transferrin saturation	88.19 %
- Ferritin	**265.4 ng/ml (15 – 150)**
Liver profile:	
Albumin	**1.97 g/dL (3.50 - 5)**
Total proteins	**4.84 g/dL (6.4 – 8.3)**
Alkaline phosphatase	**198 U/L (35-105)**
TGO	**63 U/L (0 - 40)**
TGP	27 U/L (0 - 38)
Total bilirrubin	1.16 U/L (0.00 – 1.20)
Direct bilirrubin	**1.04 U/L (0.00 – 0.3)**
Indirect bilirrubin	0.12 U/L (0.00 0.90)
C Reactive protein	**15,47 mg/dl**
Procalcitonine	0.46 ng/mL
Sodium	141 mEq/L
Potassium	4.38 mEq/L
Chlorine	109.2 mEq/L

Table 5: Admision laboratorios.

Simple urine test	**Yellow color, Cloudy appearance, Epithelial cells 1-2 x field, Leukocytes in urine: more than 100 x field. Red blood cells in urine: 15 x field. Germs: few.**
Urine culture	GRAM stain: Gram-negative bacilli. Cell count: more than 100,000 x field. Positive extended-spectrum beta-lactamases. *E coli* sensitive to Piperacillin Tazobactam, Cefoperazone Sulbactam, Ertapenem, Meropenem and Imipenem were isolated.

Table 6: Discharge laboratory.

RBC	**4340000 cel/mm³**
WBC	5250 cel/mm³
Neutrophils	77%
Lymphocytes	15%
Hemoglobin	11.7 gr/dl
Hematocrit	37.2%
MCV	81.8 fl
MCH	27 pg
MCHC	33%
Platelet count	228000 cel/mm³
Creatinine	1,16 mg/dL

Urea	46 mg/dL
BUN/Creatinine	18.5
C Reactive protein	4 mg/dl
Total proteins	6.05 g/dL
Albumin	2.9 g/dL

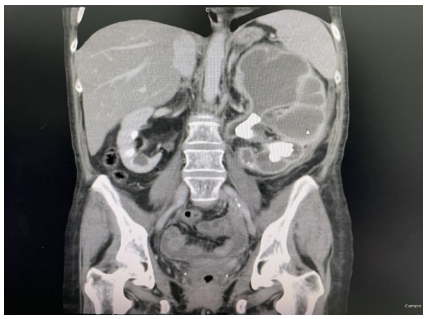

Figure 1: CT of the abdomen without contrast, coronal section. Left kidney with the presence of a staghorn stone and grade 4 hydronephrosis. Right kidney with 2 stones at the level of the pyelocalyceal junction, grade 2 hydronephrosis.

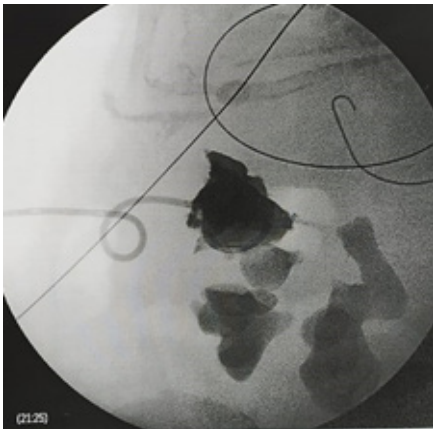

Figure 2: JJ catheter placement in the right ureter.

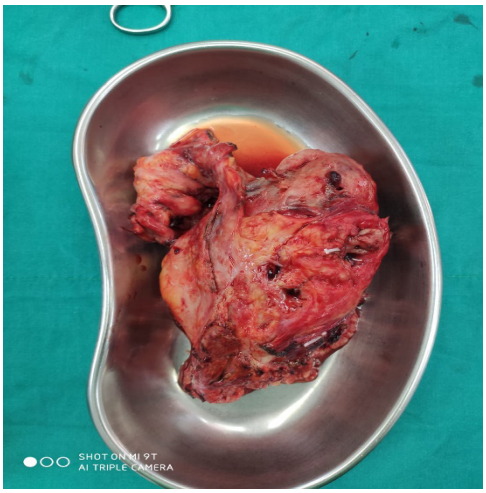

Figure 3: Macroscopic pathology. Enlarged kidney, with severe perirenal inflammatory process.

Discussion

XPN is a disease with a very low incidence in population, representing less than 1% of chronic pyelonephritis [5]. It is considered a great imitator, easily confused with renal neoplasms or other origin. The preoperative diagnosis requires the fusion of clinical and pathological findings, as well as imaging studies or biopsy [1].

The symptoms are nonspecific, they include fever of unknown origin or prolonged febrile syndrome, abdominal pain, weight loss, weakness due to severe anemia, anorexia, and constipation; less frequently the presentation includes dysuria and hematuria [1,9].

In an observational, descriptive, and retrospective study carried out in the city of Córdoba (Argentina), 10 patients diagnosed with XPN were studied. The initial diagnostic suspicion was uropyonephrosis in 70% of cases and cancer in 30%, 60% had a history of recurrent UTIs. The symptoms manifested by patients at admission in all cases (100%) were: lumbar and abdominal pain, weight loss, pallor and asthenia, 80% of the cases presented high fever and palpable abdominal mass, 100% of the cases presented anemia and 50% hematuria. In all cases (100%) the involvement was unilateral and 57% presented staghorn stones [5], findings that do not differ from the clinical presentation of our patient.

In agreement with other publications [5-7] the paraclinical tests showed altered liver function, with hypoalbuminemia, decreased total proteins, slight prolongation of PT, elevation of alkaline phosphatase, TGO and direct bilirubin, which returned to normal once the pathology was treated.

Although antibiotic treatment alone is not capable of solving the problem, it is useful in controlling the infectious process and prevents septic complications. For this therapy to be effective, it is of vital importance to have the bacterial agents well identified through a urine culture with its respective antibiogram. However, the final treatment is always surgical, performing total or partial nephrectomy as appropriate [2,5,7].

Considering the above, it can be stated that clinical and paraclinical findings of patients with XPN are not sufficient for its evaluation and diagnosis, since most of the symptoms are non-specific and imaging studies are not usually characteristic, always requiring a pathology study to confirm this disease.

Conclusion

XPN should be included in the diagnostic possibilities when studying a patient with an abdominal mass associated with nonspecific symptoms such as weight loss, prolonged febrile syndrome, or asthenia.

Anemia, leukocytosis, hematuria, and pyuria are the most frequent laboratory findings in the majority of published case series [2,5,7].

The definitive diagnosis (Gold Standard) is histopathological, however the imaging study of choice when suspected is computed tomography, useful for determining the extension of the infectious process, especially when it involves near organs.

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