CLINICAL CASE

Xanthogranulomatous pyelonephritis - a complex diagnosis: a case report

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Abstract Our aim was to report a case of diffuse stage III xanthogranulomatous pyelonephritis and the diagnostic difficulties found in relation to the differential diagnoses.

A 64-year-old woman had a past history of intense smoking, 30 cigarettes/day for a period of 20 years, and diabetes treated with insulin. She came to the emergency department presenting with a palpable mass in the left hemiabdomen, pain in the left renal fossa, and gross hematuria. She reported a 20-kg weight loss in the last year. Kidney ultrasound study revealed a kidney stone and a heterogeneous, vascularized mass dependent on the left kidney with echoes in its interior of 8 x 5 x 5 cm. She was admitted to the hospital for evaluation that ended in the performance of simple nephrectomy.

The term xanthogranulomatous pyelonephritis is an anatomopathologic concept that defines specific changes: foam cell infiltration of the renal parenchyma associated with very frequent and severe kidney infection; its lipid-laden macrophages or xanthomatous cells, so characteristic of this entity, have a morphologic similarity to the clear cells of renal carcinoma, leading to confusion when the tissue architecture is not observed. Surgery is generally accepted as the only logical course of action and nephrectomy is the treatment of choice.
Resumen

Objetivo: Reportar un caso de pielonefritis xantogranulomatosa difusa estadio iii y las dificultades diagnósticas encontradas en relación con sus diagnósticos diferenciales.

Presentación del caso clínico: Una paciente de 64 años, con antecedente de tabaquismo intenso de 20 años, 30 cigarrillos/día, diabética en tratamiento con insulina, acude al servicio de urgencias con presencia de masa palpable en hemiabdomen izquierdo, dolor en fosa renal izquierda y hematuria macroscópica, refiere pérdida de 20 kg en el último año; en ecografía renal se documenta lito renal y masa dependiente de riñón izquierdo, heterogénea, vascularizada con ecos en su interior de 8×5×5 cm. Se ingresó para estudio y finalmente fue sometida a nefrectomía simple.

Discusión: El término pielonefritis xantogranulomatosa es un concepto anatomopatológico que define unos cambios específicos: infiltración del parénquima renal por linfocitos espumosos, asociados con una infección renal muy frecuente y de características severas; presenta similitud morfológica de los macrófagos cargados de lípidos o células xantomatosas, tan características de esta entidad, con las células claras del carcinoma renal, que llevan frecuentemente a confusión cuando no se observa la arquitectura del tejido. Existe un acuerdo generalizado en aceptar la cirugía como única actuación lógica siendo la nefrectomía el tratamiento de elección.

Conclusión: A pesar de la alta capacidad diagnóstica de las tecnologías disponibles, la pielonefritis xantogranulomatosa, sigue siendo un gran simulador tanto a nivel de imagen como de histopatología, planteándonos problemas diagnósticos preoperatorios.

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PALABRAS CLAVES
Pielonefritis xantogranulomatosa; Tumor renal; Presentación clínica; Diagnóstico

Introduction

Xanthogranulomatous pyelonephritis is an atypical form of chronic pyelonephritis characterized by the destruction of the renal parenchyma and replaced with a chronic infiltrate of lipid-laden macrophages.1 It is a rare entity and makes up 1% of the cases of chronic pyelonephritis.1 This pathology was first described by Schlagenhaufer in 1916. It has 3 forms: diffuse, segmented, and focal;2 and is classified in 3 stages: in stage I the lesion is confined to the kidney, in stage II there is infiltration into Gerota's capsule, and in stage III it extends to the perinephric space and the retroperitoneal structures.

It is associated with nephrolithiasis, urinary tract obstruction, chronic urinary infection, and the most common pathogens are Proteus mirabilis, Escherichia coli, Pseudomonas, Klebsiella, and Staphylococcus.1 Clinical presentation is nonspecific and variable and sometimes can resemble a mass with tumor characteristics. Abscesses and renal tuberculosis must also be ruled out, making it a diagnostic and therapeutic challenge.3-5 Computed tomography is the ideal diagnostic method because not only does it enable the magnitude of the involvement to be determined, but also its extrarenal extension and association with neoplasia.5

Surgery is the treatment of choice and in many cases nephrectomy is required. The procedure is notoriously difficult due to the frequent inflammation of the renal pelvis, hilum, and adjacent structures and the consequent loss of the normal anatomic planes. Urinary fistula development is not uncommon, resulting in severe fibrosis that contributes to making surgical dissection a challenge.6-7

Case presentation

A 64-year-old woman had a past history of intense smoking for 20 years of 30 cigarettes/day and a 3-year history of diabetes with intermediate insulin treatment. She came to the emergency department due to a 72-h progression of asthenia, adynamia, and hyporexia. She did not present with fever and stated she had a weight loss of 20 kg during the past year. Physical examination revealed tegumental pallor, dehydrated mucosae, pain upon thigh flexion and adduction, and intense pain upon lifting the left leg. A mass in the left renal fossa was felt upon deep abdominal palpation.
The laboratory work-up reported leukocytes 14,210, neutrophils 79.6%, hemoglobin 10.5 mg/dl, platelets 488,000, glucose 108 mg/dl, creatinine 1.2 mg/dl, CRP 90, LDH 231, sodium 139, potassium 2.8, calcium 8.3, chloride 99, urinalysis with no alterations, and urine culture with no bacterial development.

Plain abdominal x-ray revealed a radio-opaque image suggestive of a left staghorn stone (fig. 1) and ultrasound identified a kidney stone that involved the entire pelvis and a heterogeneous, vascularized mass dependent on the left kidney with echoes in its interior measuring 8 x 5 x 5 cm, highly suggestive of renal neoplasia.

The patient was hospitalized to receive antibiotics and undergo complementary studies. Noncontrast and contrast abdominal tomography scans revealed an enlarged left kidney, heterogeneous parenchyma, loss of cortex/medulla differentiation, the presence of gas in the perirenal space, and an image suggestive of a stone in its interior involving the pelvis and all the calyces (figs. 2 and 3). With these findings and the null response to medical treatment, the decision was made to perform an exploratory lumbotomy, finding multiple firm adherences toward the peritoneum and an abundant oozing of a fetid and purulent secretion (figs. 4 and 5). Simple nephrectomy was carried out with no complications (fig. 6).

The general conditions of the patient in the postoperative period were regular to good and there were no complication data. Therefore she was released 7 days after surgery. The anatomopathologic study of the left kidney reported that the renal parenchyma was extensively compromised by a mixed, predominantly acute inflammatory process, with fibrosis of the parenchyma, sclerosis, and glomerular atrophy extending to the capsule and adjacent fatty tissue. The medullar areas showed atrophy, necrosis of the tubular epithelium, and a dense, mixed inflammatory infiltrate. There was a diffuse histiocytic granulomatous reaction with xanthogranulomatous foam cells that mainly affected the renal parenchyma and perinephric tissue. The conclusion was diffuse stage III xanthogranulomatous pyelonephritis with the finding of a fistula to the psoas muscle.

Discussion

Xanthogranulomatous pyelonephritis is an atypical variety of chronic pyelonephritis that is produced in the presence of urolithiasis and/or urinary tract obstruction in 20-60% of the cases. It is infrequent and considered a great imitator that is easily confused with renal neoplasias or those of other origin (carcinomas of the colon, retroperitoneal sarcomas, etc.).8 Preoperative diagnosis requires the combination of clinical and pathologic findings, as well as those of imaging.
Symptoms are nonspecific and include fever, flank pain, weight loss, anorexia, and constipations and less frequently manifestations include dysuria, pollakiuria, and hematuria, or a cutaneous fistula. 

Surgery is the treatment of choice and is nephrectomy with resection of the involved tissues. Opportune and studies or biopsy. Early diagnosis and the appropriate treatment play a crucial role in reducing the morbidity and mortality rates of xanthogranulomatous pyelonephritis.
adequate antibiotic administration is essential for preventing complications. Given the above, it must be taken into account that there are no pathognomonic imaging findings. When the numerous possible findings are added to an appropriate clinical setting, diagnosis is suggested, but it can only be confirmed through histopathology. Despite the high diagnostic capacity of the available technologies, xanthogranulomatous pyelonephritis continues to be a great simulator at both the imaging and histopathologic levels, posing preoperative diagnostic problems.

Conclusions

The preoperative diagnosis of xanthogranulomatous pyelonephritis requires a clinical approach that includes laboratory and imaging studies, as well as a high degree of suspicion, because it can simulate different inflammatory, infectious, and malignant entities. Despite the high diagnostic capacity of available technologies, xanthogranulomatous pyelonephritis continues to be a great simulator at the imaging and cytology levels, posing preoperative diagnostic problems. Definitive diagnosis is always histologic.

Because it is an atypical disease, xanthogranulomatous pyelonephritis should be considered among the diagnostic possibilities when studying a patient with an abdominal mass associated with nonspecific symptoms. When these are added to numerous, though also nonspecific, imaging study findings, an adequate diagnostic approximation and therapeutic plan can be made. In the present case, as described in the medical literature, total nephrectomy was performed, given the extent to which the kidney was compromised.

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Conflict of interest

The authors declare that there is no conflict of interest.

References