Delayed diagnosis of xanthogranulomatous pyelonephritis in a quadriplegic patient with uncontrolled cutaneous fistula

A case report

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Abstract
Rationale: Xanthogranulomatous pyelonephritis (XGP) is a chronic destructive granulomatous inflammation that is characterized by urinary tract obstruction and invasion of the renal parenchyma. Although rare, XGP can lead to fatal complications, including perinephric inflammation, psoas abscess, and cutaneous fistula.

Patient concerns: A quadriplegic patient initially presented to the hospital with a chronic open wound and cutaneous fistula.

Diagnoses: Abdominal computed tomography revealed a renal obstructing stone and enlarged right kidney with a perinephric fluid collection that communicated with the cutaneous fistula.

Interventions: The patient underwent a right nephrectomy at the department of urology.

Outcomes: Two months after surgery, the patient was clinically well with no discharging fistula.

Lessons: The XGP accompanied by complications requires an immediate evaluation and early diagnosis. In this case, the diagnosis was delayed because the state of quadriplegia rendered no symptoms of XGP.

Abbreviations: CT = computed tomography, XGP = xanthogranulomatous pyelonephritis.

Keywords: cutaneous fistulas, psoas abscess, quadriplegia, xanthogranulomatous pyelonephritis

1. Introduction

Xanthogranulomatous pyelonephritis (XGP) is a chronic destructive granulomatous inflammation that is characterized by urinary tract obstruction, urinary calculi, and invasion of the renal parenchyma. Although rare, XGP can be found in 2 forms: the focal form and diffuse form. The diffuse form of XGP is rare but manifests extrarenal complications.[1,2] Although rare, if not treated immediately, the diffuse form of XGP can lead to fatal conditions, including perinephric inflammation, psoas abscess, nephrocutaneous fistula, and renocolic fistula.[3–5] The point of XGP is an accurate diagnosis and immediate treatment of XGP is essential.[2]

In the previously reported cases, there were specific symptoms, such as flank pain and urinary symptoms. Therefore, early diagnosis and treatment were performed immediately. In the present case, however, the diagnosis was delayed due to the state of quadriplegia, which rendered no symptoms of XGP. This paper reports a rare case of XGP with a condition making a diagnosis difficult.

2. Case

A 56-year-old male with a history of newly detected type 2 diabetes mellitus, staghorn stone in the right kidney with hydronephrosis (grade IV), and a C-spine injury (C 3–4) resulting in a quadriplegia state for 40 years visited our out-patient clinic. At the initial presentation, he had no systemic symptoms. He had been in a cystocatheter insertion state for 15 years due to the urinary symptoms. Therefore, early diagnosis and treatment were performed immediately. In the present case, however, the diagnosis was delayed due to the state of quadriplegia, which rendered no symptoms of XGP. This paper reports a rare case of XGP with a condition making a diagnosis difficult.

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Incision and drainage (Fig. 2) were initially performed and the pus was sent for culture, which revealed Pseudomonas aeruginosa. Therefore, the antibacterial medication was changed from cefoxitin to cefazidime and aminoglycoside. During incision and drainage, a cutaneous fistula was found at the base of the open wound, and purulent discharge was observed oozing from the fistula. The pharmacologic therapy and massive surgical irrigations were maintained. Despite the septic conditions...
improving, the pus-like discharge persisted. Abdominal computed tomography (CT) was then performed to determine the origin of the pus-like discharge. A CT scan (Fig. 3) with intravenous and oral contrast revealed a right-sided renal obstructing stone and enlarged right kidney with multiple low-attenuation cystic lesions, showing “bear paw” like signs, and a perinephric fluid collection that communicated with the cutaneous fistula and right psoas muscle. After confirming a non-functional right kidney due to XGP, the patient underwent a right nephrectomy at the department of urology. The resected kidney was 11 × 7.2 × 4.8 cm in size and the surface had degenerated to a deep brownish color. The renal pelvis was filled with multiple deep brown-colored dystrophic calcification, greatly expanding the renal pelvis and atrophying the renal parenchyma (Fig. 4). After surgery, the patient’s condition improved showing no evidence of pus-like discharge. Two months after surgery, the patient was clinically well with no discharging fistula.

This study was approved by the institutional review board of the Chonbuk National University Hospital. Informed consent was given by the patients.

3. Discussion

XGP is a rare histological subset that accounts for only 0.6% of histologically documented cases of chronic pyelonephritis. The reported incidence of XGP is highest among middle-aged women. The 2 important factors in the pathogenesis of XGP are urinary tract obstruction and infection. Therefore, in 70% of XGP patients, the causes of the urinary obstruction, such as staghorn calculi or renal calculi, are found in many sites of the urinary tract. XGP are found most commonly in middle-aged women, and these patients generally show symptoms of fever, flank pain, malaise, and weight loss.

XGP has 2 morphological subtypes. The focal form is confined to one segment but the greater part of the kidney remains intact. This form usually does not require surgical approaches, such as a nephrostomy or nephrectomy. In the diffuse form, the whole kidney is affected and destroyed. Without treatment, this is a life-threatening condition.

Figure 1. Chronic open wound.

Figure 2. After incision and drainage, cutaneous fistula is shown.

Figure 3. Preoperative CT. XGP extend pararenal space is shown with “bear paw” sign.

Figure 4. Macroscopic appearance of right kidney. Enlarged right kidney with renal obstructing stone.
threatening disease and in most cases, antibiotics treatment
followed by a surgical approach is essential.[1,2]
Lipid-containing foam cells (xanthoma cells) observed on
the renal biopsy specimen establishes a definite diagnosis of XGP,
but noninvasive diagnostic aids, such as a CT scan, are also
available.[1] Accordingly, XGP accompanied by complications
requires an immediate evaluation and early diagnosis with a
biopsy, culture, and imaging studies. The therapeutic approach to
XGP is based on the extent of the disease. Furthermore, diffuse
or advanced stage XGP complicated by a psoas abscess and
nephrocutaneous fistula should be treated with antibiotics,
nephrostomy, and subsequent nephrectomy, whereas an early-
diagnosed focal or segmental XGP can be treated preoperatively
with antibiotics.[1,2] Symptomatic management along with broad
spectrum antibiotics before the surgical treatment and during
postsurgical care has been associated with the successful
management and better prognosis of the disease.[1,6,7]
In previously reported cases, the evaluation was not delayed
because the patients had the classical symptoms of pyelonephritis,
such as flank pain, and urinary symptoms at the time of the
diagnosis.[8] In the present case, the chief complaint of the patient
was a chronic cutaneous fistula. The patient’s particular state of
quadriplegia rendered no symptoms of XGP, such as flank pain
or urinary symptoms. Therefore, the evaluation was delayed and
made only after repeated incision and drainage; the surgical
irrigations did not relieve the symptoms in the patient. Without a
further evaluation, the patient may have transitioned to an
aggravated state due to the undiagnosed disease.
In many XGP cases, the symptoms are nonspecific, making a
diagnosis difficult, often resulting in a misdiagnosis of tumorous
disease of the kidney.[9,10] The significance of this case to a
surgeon is that chronic wounds require a further evaluation to
identify and treat the underlying cause rather than the mere
maintenance of the wound with a surgical approach and
dressings.

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