

NECROTIZING FASCIITIS OF THE RIGHT FLANK MIMICKING AXANTHOGRANULOMATOUS PYELONEPHRITIS

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ABSTRACT

Objective: Xanthogranulomatous pyelonephritis (XGP) is a rare and serious chronic inflammation of the kidney, manifesting as a granulomatous inflammation of the kidney tissue. It can be life-threatening if not recognized and managed appropriately with antibiotics and surgery. Since there are no single pathognomonic clinical or radiological features, diagnosis requires combining the results of several clinical signs and imaging characteristics. However, determining the disease is oftentimes difficult as other conditions may mimic its characteristics, such as renal cancer, mesoblastic nephroma, renal abscess, and necrotizing fasciitis. **Case(s) Presentation:** A 45-year-old man was admitted to the emergency department of Soetomo General-Academic Hospital with right flank gangrene and fever for one week. **Discussion:** The patient was admitted to the hospital with sepsis and poor laboratory result. Computed Tomography (CT) scan results showed nephrolithiasis. The patient was diagnosed with Xanthogranulomatous pyelonephritis and underwent necrotomy in the Emergency Operating theatre. **Conclusion:** During the operation, necrotizing fasciitis was found in the right flank region instead. The external oblique muscle was intact without any tunneling into the retroperitoneal space. The postoperative care was joined by the Plastic Surgery department.

Keywords: Necrotizing fasciitis, flank, kidney, Xanthogranulomatous pyelonephritis.

ABSTRAK

Tujuan: Xanthogranulomatous pyelonephritis (XGP) adalah penyakit langka dimana terdapat inflamasi kronis dengan manifestasi inflamasi granulomatosa pada jaringan ginjal. Kasus ini dapat mengancam jiwa apabila tidak dikenali dan ditangani dengan antibiotik dan pembedahan. Tidak ada gambaran klinis serta hasil pencitraan yang patognomonis dari penyakit ini, sehingga penegakkan diagnosis memerlukan kombinasi dari beberapa hasil temuan klinis dan pencitraan. Penegakkan diagnosis seringkali sulit karena kelainan lain dapat menyerupai karakteristik penyakit ini, seperti tumor ren, nefroma mesoblastik, abses ren, dan fasciitis nekrotikans. **Presentasi Kasus:** Seorang laki-laki berusia 45 tahun datang ke instalasi gawat darurat (IGD) Rumah Sakit Dr. Soetomo dengan gangren pada pinggang kanan disertai demam selama satu minggu. **Diskusi:** Pasien dirawat di rumah sakit dengan sepsis dan hasil laboratoris yang buruk. Pemeriksaan computed tomography (CT) menunjukkan nefrolitiasis. Pasien didagnosis dengan XGP dan menjalani nekrotomi di ruang operasi IGD. **Simpulan:** Selama operasi, jaringan nekrotik hanya ditemukan di regio pinggang kanan. Otot oblikus eksternal tidak mengalami nekrosis. Penanganan pasca operasi dilakukan bersama tim dari Bedah Plastik.

Kata Kunci: Fasciitis nekrotikans, pinggang, ginjal, Xanthogranulomatous pyelonephritis.

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INTRODUCTION

This Xanthogranulomatous pyelonephritis (XGP) is a variant of chronic pyelonephritis with an incidence of less than one percent.¹ It involves renal parenchymal destruction, causing the replacement of normal renal tissue with granulomatous tissue, containing xanthomatous cells.² Since its first discovery in 1916, the case has been reported

numerous times in case reports and several case series.³ However, its true epidemiology and etiology are still unknown. Nevertheless, It is most associated with immunosuppressed patients with urinary tract infections, stones, and obstructive nephropathy.⁴ There are two forms of XGP, a diffuse and a local form. The diffuse form is often mistaken for an abscess, whereas the local form is often misdiagnosed as a renal tumor.

Based on previous reports, it often mimics the characteristics of other diseases, such as kidney cancer, kidney abscess, and necrotizing fasciitis.⁵ Necrotizing fasciitis, also known as flesh-eating disease, is an infection causing soft tissue necrosis. This condition possessed sudden onset and expanded quickly. The symptoms that can occur include purplish-red skin, severe pain, fever, and vomiting. Necrotizing fasciitis commonly occurs in the lower extremity and perineal area.⁶ Risk factors for necrotizing fasciitis are immunodeficiency syndromes, including diabetes, cancer, obesity, alcoholism, IV drug use, and peripheral artery disease.⁷ It possesses similar risk factors and characteristics with XGP; thus, a misdiagnosis may occur. We report a 45-year-old male who was admitted to the hospital with necrotizing fasciitis of the right flank mimicking an XGP.

CASE(S) PRESENTATION

A 45-year-old male was admitted to the emergency department (ER) with complaints of right flank pain for a year before admission. The pain was felt intermittently. The patient also complained of fever for a week. Nausea was present, but there was no vomiting. There was a history of stone expulsion

10 years ago with the size of corn seed. There were no lower urinary tract symptoms. Diabetes mellitus and hypertension were denied. He never had any history of surgery. The patient was in a weak general condition with stable hemodynamic, but he was anemic. An abscess was found and there was a necrotic area in the right flank. There was costovertebral angle tenderness in right and left flank. There was no mass in both flanks upon palpation. External genitalia examination was normal. Digital rectal examination revealed a normal sphincter tone, smooth mucosa, no mass, normal prostate, normal bulbocavernosus reflex, and there was no blood.

Laboratory examination revealed a hemoglobin (Hb) level of 7.9 g/dL, 17.200/uL number of leucocytes, and a thrombocyte level of 390.000/uL. There was a decrease in renal function with a BUN level of 86 mg/dL and a creatinine serum of 2.63 mg/dL. The patient also suffered from hypoalbuminemia, with a 1.65 g/dL albumin and an increase in C-reactive protein (CRP) level of 293.51. The patient was in a metabolic acidosis condition, with a 7.25 pH, 7.1 mmol/L HCO_3^- , and a base excess (BE) of -19.3. Urinalysis revealed a 6.0 pH, > 100 hpf leucocyte, and a > 100 hpf erythrocyte. The laboratory risk indicator for necrotizing fasciitis



Figure 1. The Patient's Clinical Appearance Upon Admission from the superior (left) and lateral (right) views.

(LRINEC) score was 9, indicating a high risk for necrotizing fasciitis.⁸ Multiple imaging examinations were carried out, plain abdominal X-Ray, ultrasonography (USG), and Computed Tomography (CT) Scan. Plain abdominal X-Ray in figure 2 revealed multiple radiopaque shadows with

the size of 2,2 x 2.2 cm in the right lumbar vertebra 3-4 region, and the size of 3.73 x 2.23 in the left lumbar vertebra 2-3 region.

Due to the high level of creatinine, a contrast-enhanced abdominal CT-Scan could not be performed. The results of a non-contrast abdominal



Figure 2. Plain abdominal X-Ray result of the patient, showed multiple radioopaque shadows USG examination in figure 3 showed bilateral staghorn stones, right hydronephrosis, right psoas major abscess, left severe hydronephrosis, left renal cortex thinning, and a normal bladder.

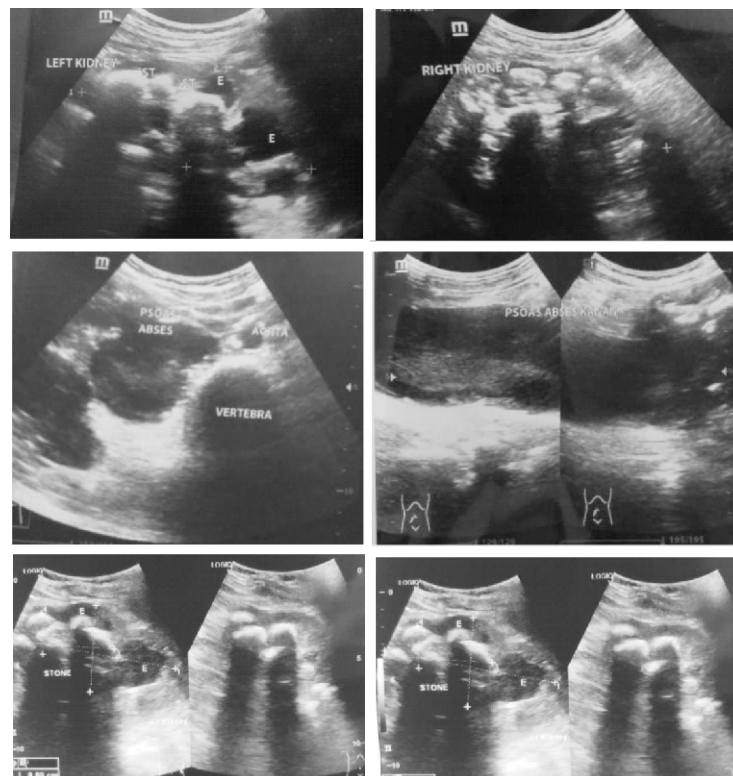


Figure 3. USG results show right and left staghorn stones, right moderate hydronephrosis, right psoas abscess, left severe hydronephrosis, thinning of the left kidney cortex.

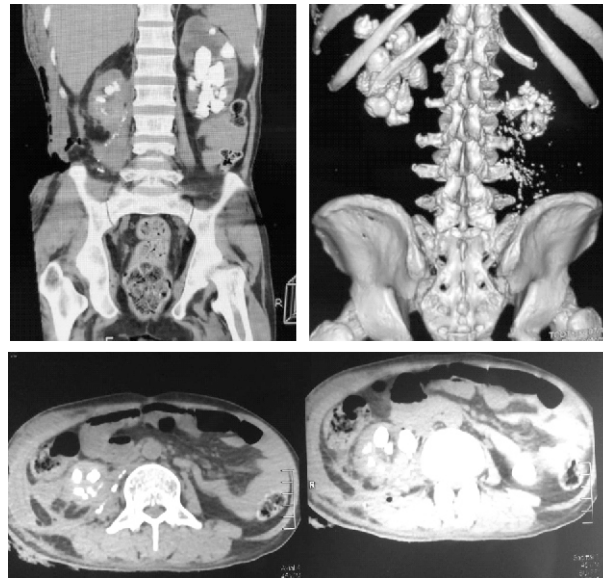


Figure 4. USG results show right and left staghorn stones, right moderate hydronephrosis, right psoas abscess, left severe hydronephrosis, thinning of the left kidney cortex.

CT scan revealed severe bilateral hydronephrosis with cortex thinning, bilateral staghorn stones, right psoas major abscess extending to the right pelvic cavity infiltrating the iliac, internal oblique, and quadratus lumborum muscles with calcification, fat stranding, and gas-forming, as shown in Figure 4.

The patient was administered 1 g of ceftriaxone twice a day and a 500 mg metronidazole infusion of 500 mg three times a day as the broad-spectrum antibiotic regimen. Oxygen support with a nasal flow of 3 lpm was given. The patient was scheduled on a high calorie high protein diet of 2100 Kcal per day. The patient was rehydrated with 0.9% NaCl, Kalbamin, and Aminofluid with a 2:1:1 ratio. Natrium bicarbonate of 50 meq was given twice a day. The patient was given two bags of PRC transfusion daily for a day until his Hemoglobin level was normal. The patient was planned for surgical debridement. Surgical debridement was performed, and necrotic tissue was found in the skin and subcutaneous layer to the right flank Scarpa and lumbar fascia. Fifty ml of pus was aspirated and tested for bacterial culture. The right external oblique muscle was intact, with no association between the right flank abscess and retroperitoneal cavity. Necrotomy was performed on the affected tissue. The field of operation was sterilized using povidone-iodine and hydrogen peroxide, as well as NaCl 0.9% for rinsing. Post-operative care in this patient was performed routinely and multidisciplinary care was performed with the plastic surgery department.

DISCUSSION

Necrotizing fasciitis is a disease mostly affecting the extremities, anogenital region, and torso due to a severe and extensive bacterial infection of the soft tissue, causing necrosis of the affected tissue.⁹ Over the past few centuries, the term has developed from hospital gangrene to necrotizing infection. In 1952, the term necrotizing fasciitis was introduced and is still used today.¹⁰ Local signs of necrotizing fasciitis include erythema, extensive edema, and necrosis. The signs are usually accompanied by a disproportionate pain beyond the margins of the apparent skin area. In some cases, hypoesthesia and anesthesia may occur instead.¹¹ Severe systemic consequences, such as fever, delirium, shock, and multi-organ failure may happen due to sepsis.¹²

Even though its incidence is very low based on the current literature, its potentially life-threatening manifestations should not be taken lightly. As previously mentioned, its occurrence in the right flank area may mimic XGP, also a chronic inflammation caused by a bacterial infection. The process was characterized by a phlegmon infection starting from the renal pelvis extending to the medulla and cortex of the kidney, causing damage which will be later replaced with lipid-laden macrophages. The infection process then extended to the tissue outside the kidney.¹³

In this patient, gangrene was found in the right flank. This condition was preceded by fever one week before admission. The incidence of XGP was around 1% from all renal infections, four times higher in women compared to men and most commonly present in five and six decades of life. Although most of the XGP cases were unilateral, XGP could present bilaterally and mostly fatal.¹⁴ The focal form of this disease was around 12.5% of all XGP cases with similar clinical appearance, laboratory result, and radiological appearance.¹⁵ During the surgical procedure, we discovered that the necrotic and abscess were limited to the superficial flank area without tunneling to the kidney, indicating that this was not an XGP, but necrotizing fasciitis of the flank area.

Necrotizing fasciitis is a subset of aggressive skin and soft tissue infections, limited to the skin and muscle fascia layers. However, the infectious process can spread rapidly to the perifascial planes, causing a secondary infection of the overlying and underlying tissues.¹⁶⁻¹⁸ To successfully treat necrotizing fasciitis, there are two important main factors, clinical awareness, and proper management. In the early stages of the disease, distinguishing necrotizing and non-necrotizing infections can be performed based on the classification consisting of clinical symptoms, used for assisting in a rapid diagnosis.¹⁹

The classification includes tenderness to palpation beyond the apparent area of involvement and disproportionate pain. Apart from pain, erythema; swelling; and increased temperature due to capillary leakage can be seen.²⁰ One of the possible methods of clinical diagnosis is the bedside finger test. The test is performed by making an incision to the deep fascia and using the index finger to palpate the fascia. Upon palpation, lack of tissue resistance or bleeding and foul-smelling pus indicate necrotizing fasciitis.²¹⁻²²

A scoring system using LRINEC can also be used.¹⁸ Imaging modalities such as USG, X-Ray, and CT-Scan can also be used. However, USG is preferable to prevent any delays. On the other hand, CT scan can show characteristics that are strongly associated with the disease. CT scan can also show more detailed information regarding the proliferation of the infection extending beyond the cutaneous signs.⁸ However, despite these modalities, surgery is necessary to rule out all doubts of a definite diagnosis. It is essential to not delay surgical debridement and antibiotic administration.²³ The

main principle of successful therapy is a radical and early debridement of all necrotic tissue. Treatment delay causes a high mortality rate.²⁴ Unclear presentations before the definitive diagnosis can be confirmed during the surgery, as with the case in this patient.⁹

CONCLUSION

Necrotizing fasciitis in the flank area may mimic the characteristics of other diseases, especially XGP. Clinical and radiologic results may not be able to completely distinguish the affected area in order to make a definite diagnosis. An awareness starting from the first encounter with the patient to the time of management is necessary to fully develop a beneficial strategy for the patient in reducing the risk of morbidity and mortality.

REFERENCES

1. Stoica I, et al. Xanthogranulomatous pyelonephritis in a paediatric cohort (1963–2016): Outcomes from a large single-center series. *Journal of Pediatric Urology*. 2018; 4(2): 169-e1.
2. Ichauoui H, et al. Xanthogranulomatous pyelonephritis in adults: clinical, biological, radiological and therapeutic main findings in diffuse and focal forms. About 42 cases. *La Tunisie medicale*. 2018; 96(8–9): 495–500.
3. Kuo C, et al. Xanthogranulomatous pyelonephritis: critical analysis of 30 patients. *International Urology and Nephrology*. 2011; 43(1): 15–22.
4. Kundu R. et al. Clinicopathological spectrum of xanthogranulomatous pyelonephritis. *Indian journal of nephrology*. 2019; 29(2): 111.
5. Wu S T. Bear paw sign: classic presentation of xanthogranulomatous pyelonephritis. *QJM: An International Journal of Medicine*. 2019; 112(6): 461–462.
6. Desta R and Yee J. Necrotizing fasciitis. *Journal of Education and Teaching in Emergency Medicine*. 2020; 5(2).
7. Wallace H A and Perera T B. Necrotizing fasciitis. *StatPearls [Internet]*. 2020
8. Bechar J, et al. Laboratory risk indicator for necrotising fasciitis (LRINEC) score for the assessment of early necrotising fasciitis: a systematic review of the literature. *The Annals of The Royal College of Surgeons of England*. 2017; 99(5): 341–346.
9. Leiblein M. et al. Necrotizing fasciitis: treatment concepts and clinical results. *European Journal of Trauma and Emergency Surgery*. 2018; 44(2): 279–290.

10. Hakkarainen T W, et al. Necrotizing soft tissue infections: review and current concepts in treatment, systems of care, and outcomes. *Current problems in surgery*. 2014; 51(8): 344.
11. Kiat H J, Natalie Y H E, and Fatimah L. Necrotizing fasciitis: how reliable are the cutaneous signs?. *Journal of emergencies, trauma, and shock*. 2017; 10(4): 205.
12. Hysong A A, et al. Necrotizing fasciitis: pillaging the acute phase response. *JBJS*. 2020; 102(6): 526–537.
13. Garrido-Abad P, et al. Bear paw sign: xanthogranulomatous pyelonephritis. *Journal of radiology case reports*. 2018; 12(11): 18.
14. Prasanni W D D and Malalasekera A P. Xanthogranulomatous pyelonephritis. *Sri Lanka Journal of Urology*. 2019; 13.
15. Jha S K and Aeddula N R. Pyelonephritis Xanthogranulomatous. *StatPearls [Internet]*. 2020.
16. Kim YH, et al. Managing necrotising fasciitis to reduce mortality and increase limb salvage. *Journal of Wound Care*. 2018; 27(Sup9a): S20–S27.
17. Lange JH and Cegolon L. Comment on: Early clinical manifestations of vibrio necrotising fasciitis. *Singapore Medical Journal*. 2018; 59(8): 449.
18. Heijkoop B, Parker N and Spernat D. Fournier's gangrene: not as lethal as previously thought? A case series. *ANZ Journal Of Surgery*. 2019; 89(4): 350–352.
19. Wang Y, Wong C and Tay Y. Staging of necrotizing fasciitis based on the evolving cutaneous features. *International Journal of Dermatology*. 2007; 46(10): 1036–1041.
20. Misiakos E P, et al. Current concepts in the management of necrotizing fasciitis. *Frontiers in surgery*. 2014; 1: 36.
21. Ryssel H, et al. Necrotizing fasciitis of the extremities: 34 cases at a single centre over the past 5 years. *Archives of Orthopaedic and Trauma Surgery*. 2010; 130(12): 1515–1522.
22. Machado NO. Necrotizing fasciitis: The importance of early diagnosis, prompt surgical debridement and adjuvant therapy. *North Am J Med Sci*. 2011; 3(3): 107.
23. Hietbrink F, et al. Triple diagnostics for early detection of ambivalent necrotizing fasciitis. *World Journal of Emergency Surgery*. 2016; 11(1): 1–7.
24. Burnham JP, Kirby JP and Kollef MH. Diagnosis and management of skin and soft tissue infections in the intensive care unit: a review. *Intensive care medicine*. 2016; 42(12): 1899–1911.