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(CASE REPORT)

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Navigating the complexities of calciphylaxis: case study, diagnosis, and treatment options

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Abstract

Introduction: Calciphylaxis is a rare but serious complication, primarily observed in patients with chronic kidney disease, especially those undergoing hemodialysis. It presents with painful, ulcerated, and sometimes necrotic skin lesions. The disease can lead to rapid deterioration of the general condition, and early diagnosis is essential to improve survival chances. This article reports the clinical case of a 49-year-old female patient who developed calciphylaxis on her left leg.

Materials and Methods: We describe the clinical case of a patient with chronic kidney disease on hemodialysis who presented with isolated chronic ulceronecrotic skin lesions, without any other associated cutaneous or extracutaneous signs. A comprehensive blood workup and a skin biopsy were performed to support the diagnosis.

Results: Blood tests revealed elevated serum calcium levels and an increased parathyroid hormone (PTH) level. The skin biopsy showed medial calcifications and signs of vascular fibrosis, thus confirming the presence of calciphylaxis. The patient also had hyperphosphatemia, a major risk factor for the disease.

Conclusion: Calciphylaxis is a rare but severe condition, often associated with chronic kidney disease and biochemical imbalances such as hyperphosphatemia. Although treatments for calciphylaxis are still under development, early diagnosis is crucial for better patient management. Treatment includes phosphate reduction, optimization of hemodialysis, and the use of sodium thiosulfate. The findings of this article highlight the importance of a multidisciplinary approach in managing this complex pathology

Keywords: Calciphylaxis; Chronic renal failure; Skin ulcerations; Mediacalcosis; Blood calcium; Hyperparathyroidism

1. Introduction

Calcyphilaxis, or uremic calciphylaxis, is a rare condition, mainly affecting patients with chronic kidney disease treated with dialysis. It is a rapidly progressive and life-threatening disease that clinically presents with persistently painful, ulcerative, or necrotizing skin lesions in multiple parts of the body. Diagnosing and treating calciphylaxis presents significant challenges.Current treatment options rely on retrospective data and are not consistently effective.We report in this article the case of 49 old female with chronic renal failure who developed extensive calciphylaxis lesions on her left leg.

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2. Material and methods

We describe the case of a A 49-year-old woman with a medical history of hypertension, heart disease, and chronic kidney failure, receiving vitamin K antagonists and vitamin D supplementation, presented with multiple painful, ulcerated and necrotic plaques on the left leg, which had appeared three months prior to her consultation, without any other notable cutaneous or extracutaneous signs (Figures 1 and 2). A comprehensive blood panel was performed, including parathyroid hormone and cryoglobulin levels, coagulation parameters, and autoantibody screening, along with a leg X-ray and a skin biopsy.

3. Results

Laboratory results revealed a serum calcium level of 109 mg/L and an elevated parathyroid hormone level of 2,484 ng/L, while coagulation parameters and autoimmune tests were within normal limits. Radiographs showed evidence of calcifications. The skin biopsy revealed epidermal necrosis as well as lesions involving the hypodermis, the intima, and the media of small- to medium-sized vessels: medial calcification, thrombi, and a moderate lobular lymphohistiocytic infiltrate .

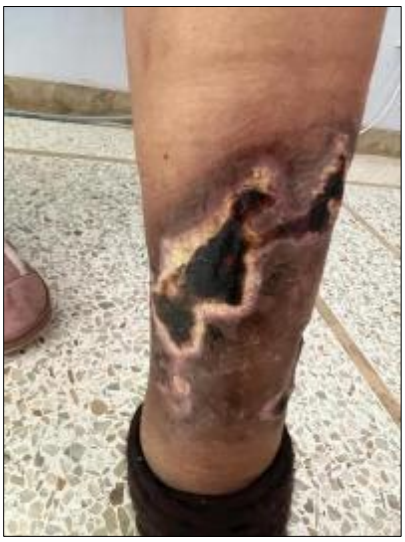

Figure 1 Ulcerated and necrotic plaques on the left leg

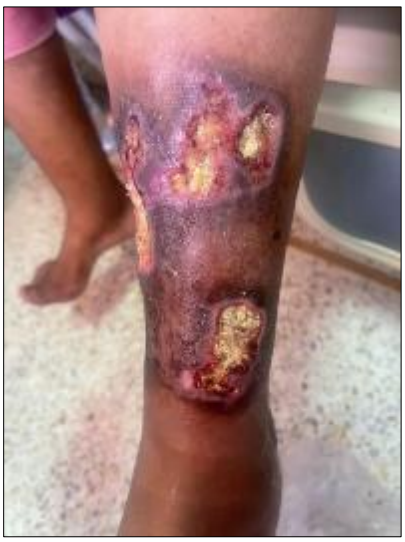

Figure 2 Clinical appearance after mechanical debridement

4. Discussion

Calciphylaxis is a poorly understood and highly severe cutaneous vascular disorder. Bryant and White first identified its association with uremia in 1898, but the term calciphylaxis was only introduced in 1962 by Hans Selye and colleagues.[1] This condition is multifactorial, fatal, and painful, with a one-year mortality rate reaching up to 50%. Despite its severity, it is extremely rare, with an incidence of less than 1%. Calciphylaxis is believed to result from an imbalance between pro-calcifying and anti-calcifying factors. The most common risk factors include hyperphosphatemia, elevated calcium-phosphate products, hyperparathyroidism, and deficiencies in vitamin D and vitamin K.[1–5]

Patients with chronic kidney disease or end-stage renal disease often develop chronic hyperparathyroidism, which leads to high-turnover bone disease, hypophosphatemia, hypercalcemia, and ectopic calcium deposition, as was the case in our patient. However, vascular calcification alone is not sufficient to cause calciphylaxis. The formation of calciphylaxis lesions requires medial calcification, intimal fibrosis of arterioles, and thrombotic occlusion. [2,3]

Diagnosing calciphylaxis can be challenging; it is primarily based on the presence of high-risk factors, characteristic skin lesions, and histopathological features. In early stages, clinical manifestations can range from chronic erythema, purpura, and livedo reticularis to painful necrotic ulcers in more advanced stages. In our case, the patient presented with painful, necrotic plaques on the left leg, which progressed to large ulcers with well-defined borders.[2,3] Lesions typically affect areas rich in adipose tissue, such as the trunk, breasts, abdominal region, flanks, buttocks, and proximal lower limbs. However, involvement of the genitalia and digits has also been reported. [2,3,5,6]

The pathognomonic histological features of calciphylaxis lesions include epidermal ulceration, focal dermal necrosis, and vascular calcification, as observed in biopsy samples. Calcium salts are easily identifiable on hematoxylin and eosin (H&E) staining due to their strong and uniform basophilia. [1–5]

When diagnosing calciphylaxis, it is essential to consider and rule out other differential diagnoses, including cholesterol embolism syndrome, warfarin- and heparin-induced skin necrosis, antiphospholipid syndrome, pyoderma gangrenosum, cryoglobulinemia, and vasculitis.

Treatment is largely based on observational studies and clinical expertise and requires a multidisciplinary approach. Management aims to prevent vascular calcification, restore blood flow, and treat wounds and pain. Preventing vascular calcification involves addressing underlying risk factors. This can be achieved by increasing phosphate removal, reducing phosphate intake, increasing the frequency of hemodialysis sessions to four or five times per week, and avoiding phosphate-rich foods. The target serum phosphate level is around 3 mg/dL. Warfarin should be discontinued in patients with calciphylaxis, and the need for continued anticoagulation with an alternative agent should be carefully evaluated. [2,4,7]

Sodium thiosulfate has emerged as one of the main therapeutic options for calciphylaxis lesion healing. It acts by chelating calcium from deposits in the skin, subcutaneous tissues, and organs, forming a compound that is more soluble than other calcium salts and can be more readily eliminated via dialysis. However, a recent systematic review and meta-analysis published in the Journal of the American Medical Association (JAMA) showed that it was not associated with significant improvement in skin lesions or survival benefits in patients with chronic kidney disease and calciphylaxis.[8]

Our patient had multiple risk factors. The therapeutic decision included discontinuing vitamin D supplementation, adding calcium chelators along with surgical debridement, optimizing dialysis parameters, and proposing a parathyroidectomy.

5. Conclusion

Calciphylaxis remains a rare complication of chronic kidney disease; however, it has a very poor prognosis. Early recognition and management of this complication can improve and extend the lives of our patients.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of ethical approval

All the procedures were carried out after the agreement of all individual participants included in the study.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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