

CASE REPORT

A rapid unfavorable penile calciphylaxis case followed by total penectomy

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Email: ovanhooetghem@hotmail.com**Abstract**

When the diagnosis of penile calciphylaxis is suggested, the evolution of the disease is rapidly unfavorable; in this case, a rapid medical treatment must be established to obtain an improvement of the disease to avoid penectomy.

KEYWORDS

amputation, calciphylaxis, hemodialysis, penile, renal insufficient, treatment

1 | INTRODUCTION

Calciphylaxis, also called calcific uremic arteriopathy, affects 0.4%-5% of dialyzed end-stage renal-insufficient patients.¹ Calciphylaxis is a rare and serious disease that results from calcium deposits in the media of small- and medium-sized arteries and causes necrotic and painful skin ulcers. Damage to the penis is unusual because of its high vascularization.

2 | OBSERVATION

A 62-year-old patient, who was hemodialyzed for two years because of end-stage diabetic nephropathy, underwent two years ago transmetatarsal amputation of the right foot and amputation of two toes of the left foot. Daily treatment included insulin and lanthanum carbonate. The patient was not undergoing antivitamin K (AVK) treatment. He had chronic ulcerated wounds on the lower limbs and was treated for painful whitish macules of the glans, with erythematous and a pruritic lining (Figure 1). Candidosis was clinically diagnosed and treated with local antimycotic agents without any clinical amelioration. A balanic mucosa biopsy was therefore performed leading to the diagnosis of lichen planus with

initiation of treatment by topical steroid cream. The patient consulted three months later in our dermatological department. The whole balanic mucosa had a whitish cardboard appearance, was subjectively painless, and showed a necrotic zone around the urinary meatus (Figure 2). The clinical diagnosis of penile calciphylaxis was performed. In addition, the last three phalanges of the right hand also showed early digital necrosis. Biological results revealed hyperphosphotemia (0.71 mmol/L), normal calcium levels (2.20 mmol/L), and hyperparathyroidism with a parathormone level of 181 pg/mL. Computed tomography angiography of the trunk and of the right upper limb revealed parietal calcification of the aorta and of the radial and cubital arteries. Doppler echography of the right upper limb revealed calcification of the antibrachial arterial crossroad with a lack of flux at the level of the interosseus artery due to parietal calcification. All of these results were in favor of our diagnostic hypothesis of calciphylaxis.

In view of this rapidly unfavorable clinical course and the advanced stage of the lesions, a full penectomy was performed and the rhythm of dialysis sessions was intensified. Histological analysis of the surgical specimens showed calcification of the small- and medium-sized arteries, essentially at the level of the corpus cavernosum, and of the dorsal part of the penis with necrosis of the glans, suggesting calciphylaxis

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FIGURE 1 Early lesion of the glans with whitish macules and inflammatory appearance



FIGURE 2 Three months later, widespread painfully whitish cardboard appearance

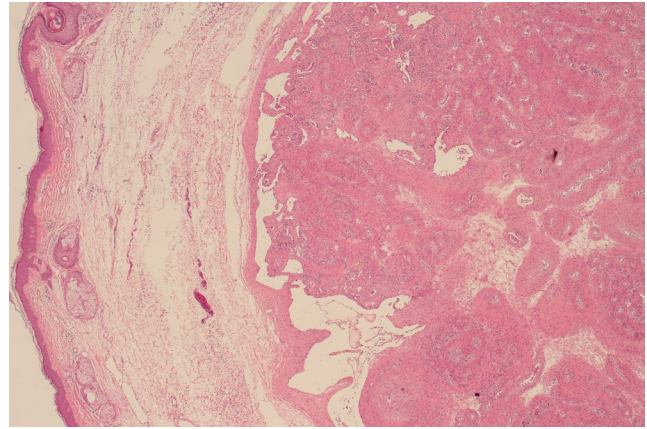


FIGURE 3 Calcium deposits in the media of small and medium arteries. Intima thickening. Reduction in vascular lumen

(Figure 3). After 4 weeks, the patient is stabilized, allowing a standardization of phosphocalcic product, the wound is healed, and dialysis was continued 3 times a week.

3 | DISCUSSION

Calciphylaxis, also called calcific uremic arteriopathy, is defined as the deposits of calcium in the media layer of small- and medium-sized vessels that can evolve into necrotic and painful skin ulcers. It affects 0,4%-5% of dialyzed end-stage kidney disease patients.¹ The etiological factors of calciphylaxis are abnormalities in the calcium-phosphate products (ie, elevated calcium-phosphate product levels and secondary hyperparathyroidism). The other risk factors are AVK intake, the presence of diabetes mellitus or obesity, female sex, and Caucasian origin.^{2,3} Two forms of calciphylaxis are described: a proximal form that affects the trunk and the limbs above the elbows and knees and a distal form that evolves the distal ends of the limbs. The latter form has a better prognosis but exposes the patient to the risk of amputation.^{4,5} The differential diagnosis of lower-limb calciphylaxis is necrotic angiodermatitis.¹ Penile calciphylaxis rarely occurs due to the rich vascularization of the penis by three interconnected arteries.⁶ To date, less than 80 cases of penile calciphylaxis have been described in the literature but the prevalence of the pathology is probably underestimated. Penile calciphylaxis is an exclusion diagnosis that is based on the combination of clinical, biological, and histological elements.⁷ Clinically, the patient presents with ulcerated and painful lesions that are resistant to treatment and appear in the context of end-stage kidney disease and often dialysis. Calcium-phosphate product is generally elevated, which can lead to secondary hyperparathyroidism. However, a calcium-phosphate product level within the normal range does not exclude the diagnosis.⁴ Histologically, deep biopsy can provide information that is useful for diagnosis. The von

Kossa staining highlights calcium deposits, but their presence is not specific and could depend on the biopsy site and medical history of the patient.⁷ However, a recent comparative study demonstrated that the presence of more or fewer interstitial vascular calcifications in hemodialyzed patients has good sensitivity and specificity for calciphylaxis and does not seem to be related to dialysis.⁸ The calcifications are thin and predominantly vascular. However, performing a deep skin biopsy in cases of penile calciphylaxis remains yet controversial because it could activate the evolution of the pathology and favor penile necrosis.^{9,10} This highlights the importance of noninvasive imaging methods to visualize the calcifications of the vessels characteristic of calciphylaxis. Although no specific medical imaging method is optimal for the diagnosis of calciphylaxis (radiography, bone scintigraphy, scanning, and mammography), the collaboration with radiologists enables earlier diagnosis.⁹ MRI is the best examination to explore the corpora cavernosa and to set clear limits between healthy and necrotic tissues.¹¹ Due to the low incidence of penile calciphylaxis, there is no literature consensus concerning patient care,¹² which must be multidisciplinary and medical-surgical. At the stage of glans necrosis, surgery is indicated (partial or total amputation).⁶ However, surgery is more controversial because of infection risks, scarring difficulties in these weakened patients, precipitation, and the potential decompensation of calciphylaxis.^{6,13} Medical treatment requires local wound care, pain management, and preservation of the phosphate-calcium balance. Prevention includes avoiding excessive intake of calcium or active vitamin D. Parenteral administration of sodium thiosulfate, a calcium ion chelator that enables solubilization and elimination, in the form of calcium thiosulfate via urine or dialysis improves prognosis, although studies on a larger scale are still necessary.¹ Lanthanum carbonate, a phosphate chelator, is also part of the therapeutic arsenal. In cases of calciphylaxis with hyperparathyroidism, the serum level of parathormone must be normalized. Cinacalcet is an alternative to parathyroidectomy, which is sometimes required.^{14,15} Hyperbaric oxygen therapy has proven efficient by increasing the oxygen supply in suffering tissues but requires specific materials and is still difficult to administer.⁶ The prognosis for calciphylaxis remains severe with significant morbidity and a mortality rate of 64%, mainly from sepsis.¹⁶ In the present case, it is not possible to state that the biopsy was an aggravating factor determining the evolution toward necrosis of the glans. We can, however, conclude, based on the review of the literature and the case of our patient, that any surgical procedure should be avoided as much as possible. The unusual localization and lack of conjunction between clinical, histological, biological, and radiological elements resulted in a delay in diagnosis for our patient, underlining the importance of multidisciplinary management.

4 | CONCLUSION

Penile calciphylaxis is a rare but serious complication. It must be recognized by specialists and detected at an early stage. The practitioner should suggest a diagnosis of penile calciphylaxis in the presence of any ulcerated, chronic, or painful lesion of the glans in patients suffering from end-stage kidney disease, particularly in those with diabetes and those on dialysis. Diagnosis is based on a combination of clinical and biological signs and requires medical imaging, particularly MRI and radiography. Penile biopsy is to be avoided when possible, and biopsy alone cannot provide a diagnosis of calciphylaxis. When the diagnosis of penile calciphylaxis is suggested, the evolution of the disease is rapidly unfavorable, but in this case a rapid medical treatment with sodium thiosulfate or lanthanum carbonate and increased dialysis must be established to obtain an improvement of the disease. Penectomy will be reserved to the last option if the disease is not controlled and progresses to necrosis.

ETHICS STATEMENT

Patient give written informed consent to publish the case report and this consent is available on request.

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Not available.

CONFLICTS OF INTEREST

All authors certified that they have no conflicts of interest, including specific financial interests and relationships and affiliations relevant to the subject of this manuscript.

AUTHOR CONTRIBUTIONS

The content of the paper "A Rapid unfavorable penile calciphylaxis case followed by total penectomy" by CR-G, IT, ML, and OV has not been published or submitted for publication elsewhere. All authors: have read and approved the manuscript. In keeping with the latest guidelines of the International Committee of Medical Journal Editors, each author's contribution to the paper is to be quantified.

DATA AVAILABILITY STATEMENT

Data are available on request.

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