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Penile calciphylaxis diagnosis and treatment challenges a case report

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ABSTRACT

INTRODUCTION: Painful tip penile ischemic lesion that varies from ulceration to dry gangrene which is calcified in a patient with ESRD on chronic dialysis is a seriously complicated disease due to microvascular disease of subcutaneous and adipose tissue.
CASE PRESENTATION: 72 gentleman who is on chronic dialysis for the last 8 years because of ESRD, In which he developed many vascular disease and amputation done for him presented with spreading black painful areas at the tip of the glans for which conservative treatment took place for about month.
DISCUSSION: The diagnosis and management of this rare disease still unclear. Diagnosis mostly clinical, treatment conservative versus surgical.
CONCLUSION: Controversies of for penile Calciphylaxis diagnosis and treatment for its rarity, high mortality rate, and as its part of systematic disease treatment till know individualized according to patient status and extent of the necrotic area.

1. Introduction

Penile calciphylaxis is part of a systematic disease affecting mainly patients with end-stage renal disease on dialysis (ESRD) [1,2]. Calciphylaxis incidence is estimated to be 1–4% in chronic hemodialysis patients [3,4]. One of the rare areas to be affected by calciphylaxis is the penis [4] which could be explained by a penile rich vascular network [5]. Penile calciphylaxis is very rare with only about 50 reported cases in the literature [5–7]. We report a case of 65 years old male with penile calciphylaxis. We also provide a review of pertinent literature.

This work has been reported in line with the SCARE 2018 criteria [13].

2. Case presentation

We report a case of a 65 years old Caucasian male patient with ESRD on dialysis since 2013. In the last year, he started to develop painful infected lower limb ulceration, 4 months before admission he developed ischemia in his right leg for which trans metatarsal amputation was performed. He started to complain of a painful ulcer at the tip of the penis in February 2020 for which he was treated with dressing and antibiotics. The ulcer failed to heal and spread within 2 weeks to cover the whole glans. The pain became more severe and black eschar start to appear at the glans penis.

One month later he presented to our center complaining mainly of agonizing pain. Therefore, the decision was made to admit the patient as a case of advanced penile calciphylaxis. On physical examination, there was a black eschar covering the whole glans, which was extremely hard, and tender. There was also dryness of the penile shaft skin with infected oozing edges of the black lesion (Fig. 1).

His laboratory results showed low albumin (2 g/dL), CRP (58.9 mg/L), Hemoglobin (5.13 g/dL), WBC (6.8 * 103 /dL), serum creatinine (4.9 mg/dL), Phosphorus (5.15 mg/dL), calcium (8.8 mg/dL) and PTH (26.45 pg/mL). Pelvic x-ray show bbhed extensive vascular calcifications (Fig. 2).

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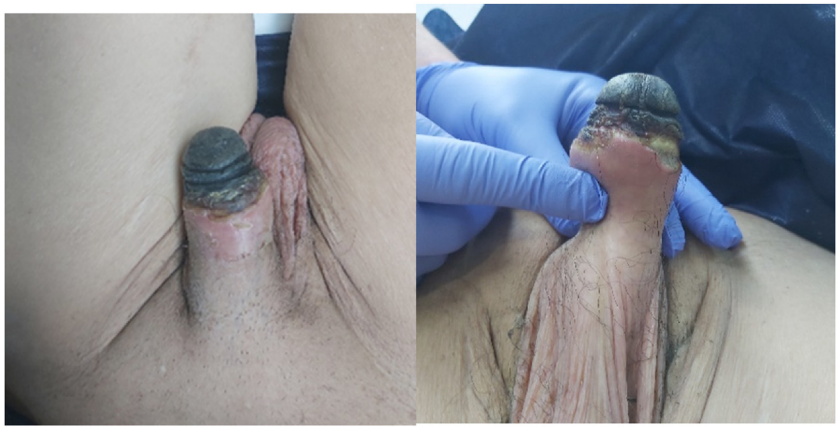

Fig. 1. Black eschar at glans penis with infected edges.

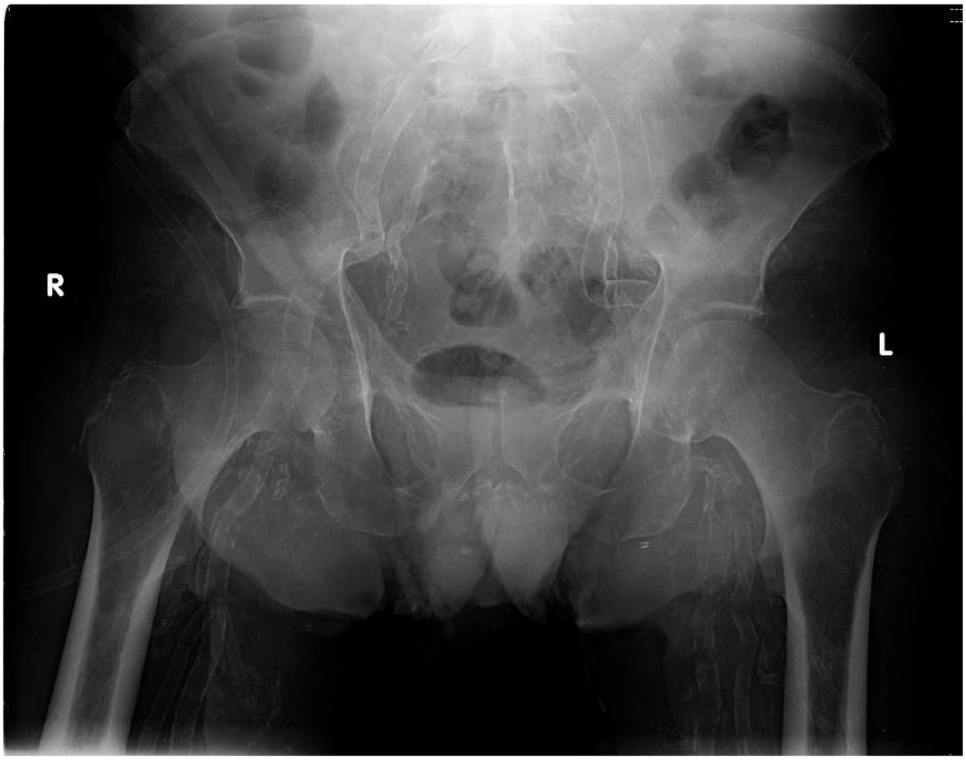

Fig. 2. Pelvic x-ray with extensive vascular calcifications.

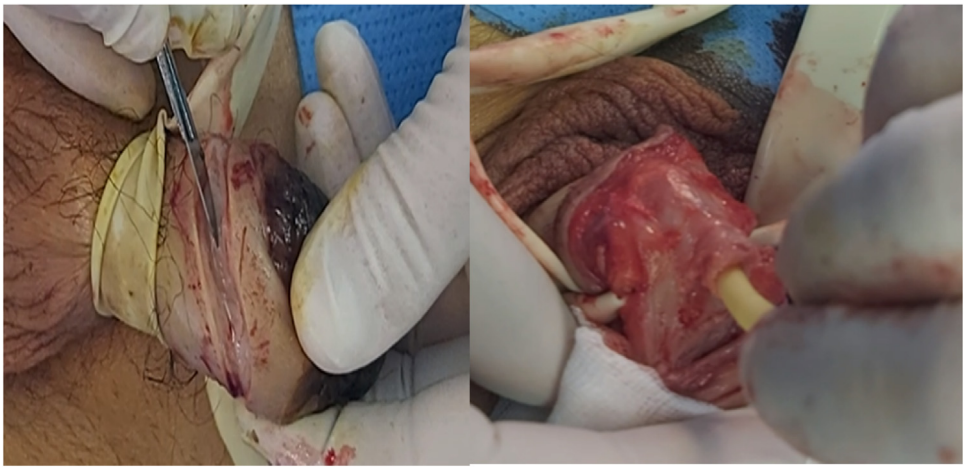

Fig. 3. a) 1 cm safety margin from the infected area, b) adequate urethral length.

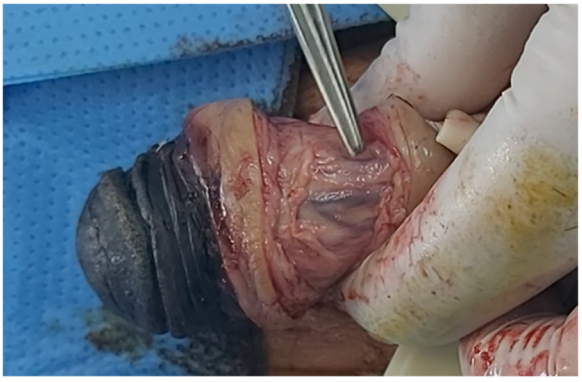

Fig. 4. Poor blood supply and thrombosis of superficial dorsal vein.

After stabilization of the patient general condition and blood transfusion for his anemia, the patient was offered a partial penectomy to relieve the excruciating pain. Under general anesthesia, a circumferential incision was made to about one cm from the edge of the infected area down to the healthy viable skin tissue (Fig. 3a). Partial penectomy was done with the preservation of adequate length of the penile urethra for anastomosis (Fig. 3b).

Interestingly, what was noticed during penectomy is the poor blood supply, hardness of the tissue during dissection, and thrombosis of the superficial dorsal vein (Fig. 4).

Pathology confirmed the diagnosis of penile calciphylaxis with evidence of intimal calcification and thrombosis (Fig. 5).

The patient recovered well, pain – which was the patient's main concern – subsided immediately after surgery. The wound healed completely in 2 weeks. At 6 months follow-up, the patient was doing good with complete healing of the penis (Fig. 6).

3. Discussion

Penile calciphylaxis is a very rare condition with around 50 cases reported in the literature [5–7]. In most of these cases, it is associated with systemic calciphylaxis [8]. Calciphylaxis is a rare and severe disease that manifests with painful skin ulceration and necrosis [9]. This life-threatening condition manifests in very painful nonhealing ischemic lesions due to underlying occlusion of micro vessels in the dermis, subcutaneous, and adipose tissue [1,2]. The described mortality rate of calciphylaxis, in general, is about 64% predominantly resulting from sepsis [5,10]. It carries a very poor prognosis with less than 1-year survival rate [1]. Challenges in dealing with penile calciphylaxis start from the complexity of its pathophysiology which is multifactorial and poorly understood [3,4], its rarity [6,7], the systematic nature of this disease [1], and the high mortality which reaches 69% in the first 6 months [7] which

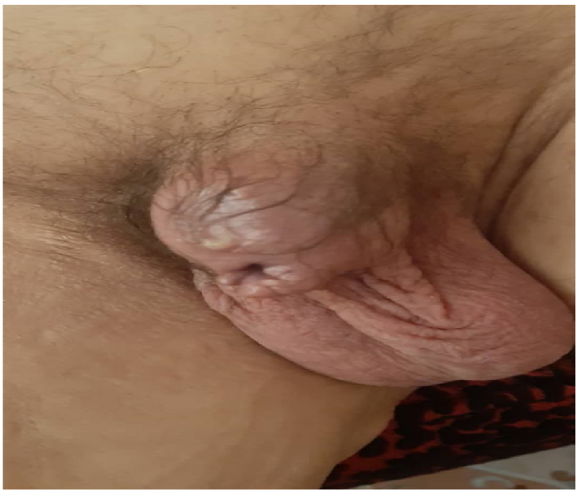

Fig. 6. The penis at 6 months follow up.

make data in the literature about penile calciphylaxis limited to the few case reports published [5–7].

Many laboratory and radiological measurements failed to give a solid background for the diagnosis [1,8,9]. Penile Doppler ultrasound can be performed to assess the penile vascularity to exclude gangrene. CT is sensitive to check for calcifications, and MRI to assess for spreading of ischemia [11]. Thus, until the time being published research about penile calciphylaxis rely mainly on clinical diagnosis [6,7,9]. Clinical diagnosis should be considered in an ESRD patient presented with agonizing pain and erythematous tender penile lesion which might be covered with black eschar [7,9]. Early diagnosis is essential to prevent delay or improper treatment as it might be aggravated by the introduction of corticoids if misdiagnosed as balanitis [6]. Although biopsy of the lesion is the gold standard for definitive diagnosis [8], it carries the risk of sepsis and poor wound healing [4,8,9]. Hence, there are controversies about performing a biopsy in the standard workup and it is reserved for equivocal cases [9].

The differential diagnosis of penile calciphylaxis includes sexually transmitted diseases or other infections, primary neoplasms such as squamous cell carcinoma, trauma, fixed drug eruption, cutaneous Crohn's disease, pyoderma gangrenosum, erosive lichen planus, and contact dermatitis [12]. Treatment of penile calciphylaxis is challenging because of the systemic nature of the disease [2,5,9], and the unclear pathophysiology [2,9]. All the available data is based on case reports [5–8]. The main described problems are infection, poor healing, and agonizing pain [2,5,8,9].

There is no consensus regarding optimal management. The described treatment in literature is multidisciplinary and indi-

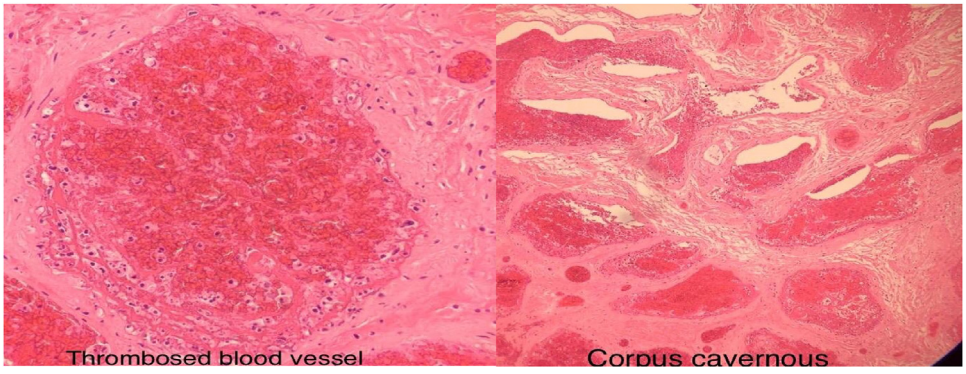

Fig. 5. Thrombosed blood vessels.

vidualized, conservative initially with controlling electrolyte disturbances mainly ca, phosphate and PTH, correction of nutritional status, analgesic and wound care of sepsis to partial versus total penectomy in refractory, progressive and intolerable pain cases [3,7,9,10]. For medical treatment; sodium thiosulfate was used due to its anti-oxidative and vasodilatory properties [2], and bisphosphonates were also described which is a potent inhibitor of calcium hydroxyapatite formation and its anti-inflammatory effects may play a role in decreasing vascular smooth muscle calcification [2]. The benefit of surgery remains controversial and individualized. The survival was not statically significant whether local wound care verse penectomy was done [5,7].

4. Conclusion

Although penile calciphylaxis is rare, we must keep this rare entity in our differential diagnosis in ESRD patients on dialysis presenting with penile ulceration. Our case emphasizes that careful clinical assessment helps early diagnosis and reduces improper management due to the unawareness of the disease. The definitive management for our patient was partial penectomy and the patient was doing well at his 6 months follow up. Nevertheless, the role of surgical versus conservative management remains controversial and larger volumes of patients are needed to reach a consensus.

Declaration of Competing Interest

No conflict of interest.

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Ethical approval

This research did not need ethical approval.

Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

Author's contribution

MORAD, BANI-HANI; study design, data collection, data analysis and writing the paper.
Sager Nawafleh: writing the paper.
Mohammad Al-zubi: reviewing the paper.
Hassan Alkhatatbeh: writing the paper.
Y Altal: writing the paper.
Mohammed Yahia Sarhan: writing the paper.
Salah Tewfik Daradkeh: writing the paper.
Sulieman Alriyalat: writing the paper.
Sakher Tahaineh: writing the paper and editing.

Registration of research studies

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Guarantor

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