CASE REPORT

Bali Medical Journal (*Bali Med J*) 2020, Volume 9, Number 1: 163-166
P-ISSN.2089-1180, E-ISSN.2302-2914

Published by DiscoverSys

Penile necrosis due to calciphylaxis in peritoneal dyalisis patient: A case report

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ABSTRACT

Introduction: Calciphylaxis (or calcific uremic arteriolopathy) is a life-threatening condition that affects 1–4% of patients with end-stage renal disease (ESRD). Penile necrosis is one of the complications of calciphylaxis that occur in ESRD patients undergone haemodialysis. Although rarely happen, this condition is potentially life-threatening and usually has a poor prognosis. It is associated with diabetes mellitus (DM), cholesterol embolism. The treatment varies, from surgical to conservative, and is usually depends on patient's general condition.

Case presentation: A 41-years-old male, known to have ESRD and underwent hemodialysis for 1 year in 2014, but has been switched to peritoneal dialysis ever since, presented with complaints of severe pain, purulent and progressive necrosis of glans penis within the last two months, and history of long-term DM and hypertension. Physical examination showed that the patient was alert, with blood pressure of 141/80 mmHg, pulse rate of 88 bpm, respiration rate of 20 times per minute, and temperature of 36.8ºC. A yellow purulent tissue was noticed over the glans penis, foul-smelling discharge was observed from urethra and undersurface of penile lesion. Laboratory examination showed elevated calcium level of 7.3 mg/dL. Ultrasound revealed multiple calcifications at the base of penis, hypoechoic lesion, nearly circular, irregular border, likely sac with thin wall, attached to the gland penis, and filled with exudate. Doppler confirmation showed avascular area. The patient was treated with partial penectomy, debridement, and antibiotic.

Conclusion: Calciphylaxis in peritoneal dialysis patients as reported in this case was rarely found. Although the treatment is still debatable, however, we decided to impose surgical approach in this patient along with antibiotics. Despite its poor prognosis, the patient in this case showed improvement after the procedure.

Keywords: calciphylaxis, end-stage renal disease, penile necrosis.
Cite This Article: Dahril, Manurun, F., Pratama, R. 2020. Penile necrosis due to calciphylaxis in peritoneal dyalisis patient: A case report. *Bali Medical Journal* 9(1): 163-166. DOI:10.15562/bmj.v9i1.1712

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Received: 2019-12-19
Accepted: 2020-03-13
Published: 2020-04-01

INTRODUCTION

Penile necrosis as a consequence of a vasculopathy is seen in patients with diabetes with chronic renal failure on long term hemodialysis. It was first described in uremic patients by Bryant and White in 1898, though the term calciphylaxis was coined by Hans Seyle in 1962. It has been termed Calciphylaxis or Calcemic uremic arteriolopathy.[1] Calciphylaxis (or calcific uremic arteriolopathy) is a life-threatening condition that affects 1–4% of patients with end-stage renal disease on haemodialysis or those who received renal transplant. It is characterised by medial calcification and intimal fibrosis of medium and small arteries. Macroscopically, cutaneous necrosis is a characteristic clinical presentation that affects the distal extremities, buttocks, thighs and, less frequently, the penis. It is a clinical condition with a high mortality rate >60% within 6 months.[1,2]

Risk factors include female, obesity, diabetes, high phosphate concentration, medications (warfarin, systemic steroids, calcium binders, vitamin D analogues), a hypercoagulable state and hypoalbuminaemia.[3] Diagnosis is clinical: painful, non-ulcerating subcutaneous nodules or plaques, non-healing ulcers and/or necrosis, which are most commonly present in the thigh, areas of increased adiposity, and the penis. Treatment consists of dialysis, sodium thiosulfate, wound care, correction of calcium–phosphate abnormalities, cinacalcet, parathyroidectomy (if the parathyroid hormone (PTH) is elevated) and supportive care.[4]

This case is about a very rare disease that has been described in the literature. What makes our case unique is that it involves the penis and was seen in a patient on peritoneal dialysis (CAPD). Treatment is supportive. This patient was taken to the operating room for partial penectomy, penile debridement, and antibiotic therapy, then converted to haemodialysis.

In this study, we report a rare clinical case of penile and generalised calciphylaxis in a patient on peritoneal dialysis.

CASE DESCRIPTION

A 41-years-old Asian man was admitted to Zainoel Abidin hospital on May 4th 2018, known to have ESRD and was on hemodialysis for 1 year in 2014, but has been on maintenance peritoneal dialysis until now. The patient presented with chief complaints of severe pain, purulent and progressive

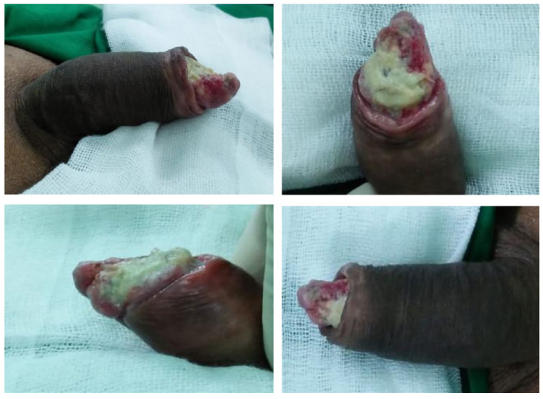

Figure 1 Gangrenous glans penis

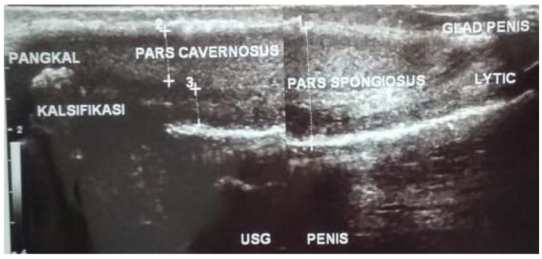

Figure 2 Ultrasonography examination showed Calcification at Base of Penis

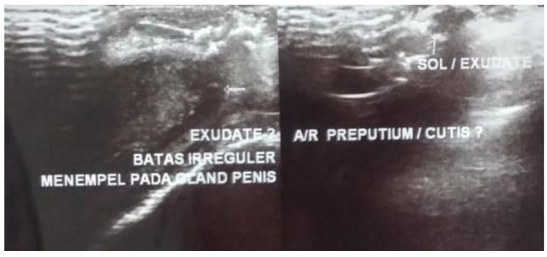

Figure 3 Ultrasonography examination showed exudate with irregular border attached to gland penis

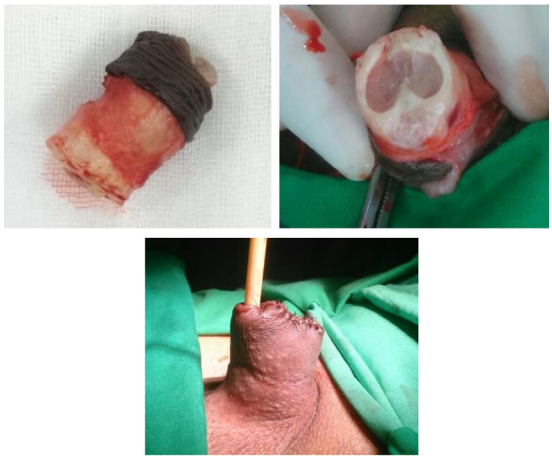

Figure 4 Partial penectomy

necrosis of glans penis of two-months duration. On review of medical history, the onset of skin lesions was 2 months prior to admission. The initial lesion was like mosquitos bite with reddish wound at first. The patient had history of long-standing diabetes mellitus and hypertension.

On physical examination, the patient was alert mental status, blood pressure from right brachial artery in supine position was 141/80 mm/Hg, normal pulse rate (88 beat per minute), normal respiration rate (20 times per minute), and normal temperature (36.8°C). Localized examination at region Penis showed a yellow purulent tissue was noticed over the glans penis, foul-smelling discharge was coming out of lesion and necrotic of penile (Figure 1). His Laboratory examination showed Anemia (9.3g/dl), leukocytosis (16.800/mm3), low calcium level (7.3mg/dl), elevated uremic (222mg/dl), elevated creatinine (13.89mg/dl), low sodium (129mg/dl), and elevated potassium (5.5mg/dl). No other significant electrolyte abnormalities were noted. Ultrasonography at regio gland penis revealed multiple calcifications at base of penis, hypoechoic lesion, nearly circular, irregular border, likely sac with thin wall, attached to the gland penis, and filled with exudate. Doppler confirmation showed avascular area. Echocardiography examination revealed intracardiac thrombus and vegetation with ejection fraction 64%, left ventricle dilatation, and left ventricle hypertrophy.

He was diagnosed with calciphylaxis based on a clinical presentation combined with a medical history of diabetes and end-stage renal disease, and calcification on USG (Figure 3). The patient was treated with partial penectomy, debridement, and antibiotic therapy. The patient underwent blood culture before discharged from hospital, and the result showed none growth organism in the blood, suspected due to given antibiotics treatment or caused by organism instead of bacteria (Figure 4).

DISCUSSION

Calciphylaxis (calcific uremic arteriolopathy) is an obliterative calcific vasculopathy in which calcification occurs in the media of small arteries of up to 600 μm in diameter, arterioles, and capillaries, with intimal fibrosis and thrombosis leading to tissue necrosis. Most commonly it occurs in haemodialysis dependent patients who have ESRD or renal transplant recipients. It affected up to 5% of dialysis-dependent patients. Risk factors include female gender, diabetes, and obesity. Other associations are raised calcium/phosphate product (CPP), high serum alkaline phosphatase level, malnutrition, and low serum albumin level. Calciphylaxis also occurs in the absence of renal failure in the context of hyperparathyroidism, malignancy, chemotherapy,

Published by DiscoverSys | *Bali Med J* 2020; 9(1): 163-166 | doi: 10.15562/bmj.v9i1.1712

corticosteroid and warfarin therapy, alcoholic liver disease, autoimmune disease, and inflammatory bowel disease.[5] CUA typically presents with ischemic necrosis predominantly involving areas of adiposity in the body such as the trunk, buttocks, or proximal extremity. However, non-adipose regions can be involved and patients can present with digital ischemia and more rarely penile gangrene as is the case with this patient.[6]

Isolated gangrene of the penis represents a localized manifestation of vascular calcification which occurs in patients with ESRD. Diabetic vasculopathy can also contribute especially with superimposed infection. By conducting a review of the medical literature, very few of these cases were found to be reported. Although pathogenesis is not clearly understood, abnormalities in mineral metabolism that predispose to vascular calcification might play a role. Histologically there is calcific infiltration of tunica media with subsequent intimal hyperplasia leading to marked luminal compression.[7,8]

Our patient has end-stage renal disease on peritoneal dialysis. Calciphylaxis is rarely described in patients on peritoneal dialysis. It is a disease mostly noted in patients on haemodialysis. Few limited studies have shown that peritoneal dialysis is by itself a risk factor, but this remains to be proven. From the laboratory finding showed calcium level was 7.3 mg/dl, while phosphorous and PTH were not examined. Our patient had two additional risk factors: he has intracardiac thrombus and vegetation where the ejection fraction was 64%, and in addition to type 2 diabetes. Long-standing diabetic microvascular disease and predisposition to atheroma in larger arteries could, in this case, have contributed to critical tissue ischaemia.

Demonstration of widespread calcification of vascular smooth muscles and fibrinous thrombi occluding vessel lumina in absence of inflammation confirms the diagnosis. If there are no contraindications, skin biopsy is recommended, which shows medial calcification of the arterioles with intimal hyperplasia leading to arterial occlusion and ischaemia. Histopathology of the excised tissue should demonstrate. Other diagnostic modalities are ultrasound scan, Computed Tomography scan, and bone scans.[9,10,11]

Radiological studies in our patient showed multiple calcifications at base of penis, hypoechoic lesion, nearly circular, irregular border, likely sac with thin wall, attached to the gland penis, and filled with exudate. Doppler confirmation showed avascular area. Those modality help to diagnose as is the case in our patient.

Treatment usually consists of a combined medical and surgical approach. The surgical management of the gangrenous penile lesions is also debated. Wood et al. recommended penectomy, while others believe that local wound care and debridement of penile lesions are sufficient. In the presence of infection, aggressive surgical debridement or penectomy is often required for any chance of survival.[6] Prompt treatment of calciphylaxis is important to reduce the risk of progressive necrosis and sepsis. Surgical treatment of penile calciphylaxis includes circumcision or partial penectomy. Adjuncts to surgical treatment include correction of the calcium–phosphate balance, and management of renal failure, infection, and nutrition. Newer treatments include hyperbaric oxygen therapy and sodium thiosulphate.[5] Regarding penile involvement, Doppler ultrasound scan showed multiple calcifications with absent flow to the glans. Penile involvement was managed surgically. Our patient had a partial penectomy, debridement and antibiotics therapy.

Early recognition of the condition together with liberal pain control and prevention and/or treatment of infection (local wound care and appropriate antibiotics) are critical to allow wound healing and decrease morbidity and mortality in these patients.CUA should be differentiated from the diseases that could have similar clinical presentations such as Warfarin skin necrosis, cryoglobulinemia, vasculitides, cellulitis, nephrogenic fibrosing dermopathy, and cholesterol embolization, which are all unlikely in the patient presented here based on history of physical examination and the laboratory results.[6]

In conclusion, calciphylaxis in ESRD is usually found in haemodialysis patients but rarely discovered in peritoneal dialysis patients. It is characterized by calcification of subcutaneous arteries and infarction of the subcutaneous cellular tissue and overlying skin. The diagnosis is usually based on clinical signs, symptoms, and calcification in radiology examination. Although the treatment is still debatable, however, we decided to treat the patient with partial penectomy, debridement, and antibiotic. Despite its poor prognosis, the patient in this case showed improvement after the procedure. Proper studies are needed to further investigate calciphylaxis in peritoneal dialysis patients.

CONFLICT OF INTEREST

The author declares no conflict of interest in this report.

FUNDING

This study was self-funded by the authors.

AUTHOR CONTRIBUTIONS

Dahril contributed to the principal investigator of this study, in charge of the content validation. Fitro Manurun responsible wrote the main part of the report. Rovy Pratama responsible for final editing and preparation of the manuscript. All author had agree and review the final manuscript for publication.

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