Penile necrosis and calciphylaxis

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ABSTRACT

Penile necrosis is an uncommon condition associated, in most cases, with calciphylaxis, also known as calcific uremic arteriolopathy. It is a progressive disease found in approximately 1%-4% of patients with end stage renal disease. Patients with penile calciphylaxis present a mortality rate of up to 70%, with life expectancy of two-and-a-half months following the diagnosis. We report a severe calciphylaxis case that had to be submitted to a penectomy but survived the penile event.

Introduction

Penile necrosis is an uncommon condition associated, in most cases, with calciphylaxis, also known as calcific uremic arteriolopathy. It is characterized by calcification and fibrosis of the intima of medium sized and small arteries, resulting in ischemia and necrosis of the skin and of the subcutaneous tissue. It is a progressive disease found in approximately 1%-4% of patients with end stage renal disease, and it is connected with high calcium and phosphate circulating levels, notwithstanding reports of cases with normal serum levels.

Penile calciphylaxis has a bad prognosis and patients as a rule suffer an intense penile pain, due to the ischemia caused by calcification and fibrosis of the penile arteries. Extreme forms may require penectomy. We report a severe calciphylaxis case in a man that had to be submitted to a penectomy but survived the penile event.

Case report

Fifty-eight-year old patient, complaining of penile pain and edema, associated with foul smell for about 2 weeks. He used topical treatment with ointments, with no improvement to the condition. He has chronic renal disease, undergoing hemodialysis for over 4 years, hypertension and type 2 diabetes. Five years ago he had an ischemic stroke event, shows intermittent claudication for short distances and, now and then, chest pain events. He has secondary hyperparathyroidism, regularly using cinacalcet, isosorbide, NPH insulin, gabapentin and captopril. He smokes and drinks.

At the first examination wet necrosis and hyperemia were found all over the penis (Fig. 1). Debridement of the penile skin was first performed, but evolved to necrosis of the entire penile shaft in 5 days, when he underwent total penectomy (Fig. 2) with confection of perineal urethrostomy (Fig. 3). He was treated with ceftriaxone and metronidazole and improved gradually, and eventually was discharged 5 weeks later. He is currently monitored in different clinical treatment groups, and is clinically controlled. The anatomopathological exam revealed presence of an ischemic necrosis, with Monckeberg atherosclerosis and acute inflammatory infiltrate.

Discussion

Penile necrosis is a rare condition, given a large network of collateral vessels at the origin of the lower limbs. Most patients with penile necrosis are aged between 40 and 60 years, with a history of atherosclerosis, diabetes mellitus, intravascular calcification, chronic renal disease requiring dialysis, obesity or high blood pressure. Treatment of such comorbidities, helps preventing penile necrosis, in addition to improving the patient’s quality of life.

Patients with penile calciphylaxis present a mortality rate of up to 70%, with life expectancy of two and a half months following the diagnosis; our patient developed well clinically, currently has good quality of life.
healing of wounds, one year after surgery. In this case, necrosis occurs due to total obstruction of the artery lumen on account of the arterial wall that leads to acute ischemia and posterior formation of necrotic tissue. Our patient additionally presented necrosis of the tunica media, also known as Monckeberg arteriosclerosis, in which there is focal calcification and even bone formation inside the arterial wall, and segments of the artery may turn into a calcified and rigid tube, with no lumen obstruction.

Diagnosis may be difficult before necrosis is installed but it must be considered in patients with end stage chronic renal disease and a painful penile lesion. High values of serum calcium, phosphorus and parathyroid hormone can be useful for diagnosis, although some patients may have normal hormone and ion levels. Biopsy can be useful for the final diagnosis, although there are recommendations that routine biopsy should be avoided for implying risk of infection and development of the injury to wet gangrene.

In a suspicious case, Doppler ultrasound, computed tomography and magnetic resonance should be performed. It is recommended that the Doppler ultrasound be the first exam to be requested for better assessment of the vessels permeability and penile blood flow. A CT scan can assess the dimension of vascular calcification in soft tissues, at the infection site, in the necrotic area and in ischemic areas, whereas magnetic resonance images can better specify the edges of the necrotic tissue. In our patient’s case, as skin debridement developed to necrosis of the shaft of the penis, supplementary tests were unnecessary.

Before penile necrosis develops, a conservative approach can be adopted aiming at topical treatment of the wound, pain control and normalization of the patient’s altered lab results, i.e., reduction of the calcium and phosphate serum concentrations. Sodium thiosulfate should also be used as it appears to be a good alternative, of proven efficacy in a significant number of patients with calciphylaxis. Although the mechanisms through which this drug operates are not fully understood yet, it is known to remove calcium from the vessels through chelation, probably also mobilizing the calcium ions of the blood stream. Thus, sodium thiosulfate can either delay or disrupt development of additional calciphylaxis lesions, preventing formation of calcium crystals as well as a deeper involvement of the blood vessels. The use of phosphate blockers is recommended for patients undergoing hemodialysis and presenting hyperphosphatemia, whereas in patients with high levels of parathyroid hormone the use of cinacalcet is advisable, which had been given to our patient.

Conclusion

The prevention of calciphylaxis should be thought in all patients with end stage renal disease because the risk of penile necrosis is great and penectomy can often not be avoided, as happened with our patient.

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