Case Report

Calciphylaxis with facial involvement

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Introduction

Calciphylaxis presenting as progressive painful necrosis of the skin and subcutaneous tissue in patients of chronic renal failure is increasingly recognized owing to the greater awareness of the condition [1–11]. Pathogenesis remains uncertain and specific therapy is debatable. We report a case with facial involvement, which to our knowledge has not been documented before. This patient subsequently died of septicemia and spontaneous gastrointestinal haemorrhage. Diagnostic investigations like endoscopy and mesenteric angiogram, though planned, could not be carried out due to the poor general condition of the patient.

Case

A 58-year-old non-diabetic Caucasian man with chronic renal insufficiency of unknown duration was under renal follow-up after he was found to have impaired renal function during routine investigations following a myocardial infarction. The renal failure was diagnosed after renal biopsy to be due to mesangio-proliferative glomerulonephritis secondary to IgA nephropathy. He was treated with conventional maintenance immunosuppressive therapy that had to be stopped subsequently as his renal function continued to decline. Haemodialysis was initiated on approaching end-stage renal failure using a left brachio-cephalic arterio-venous fistula. Following a number of access-related thrombotic episodes he was commenced on warfarin. Thrombophilia screen was negative. Extensive vascular calcification was noted on abdominal radiograph. His compliance with diet and phosphate binders (calcium carbonate) was doubtful as serial blood results showed elevated serum phosphate of >2 mmol/l and calcium–phosphate product (Ca × P) of >5 mmol/l. While he had been on dialysis for eight months, violaceous discolourations were noticed over the dorsum of his hands, face (Figure 1), scalp, and feet. These evolved into shallow, well-demarcated, non-healing ulcers. Over the next four months he developed a large ulcer on the anterior abdominal wall. Deep ulcers were present on the thighs and the calves. Due to inadequate healing of these lesions he was admitted for further intervention with a multidisciplinary approach, including participation by the plastic surgeons.

At the time of admission he weighed 118 kg and his height was 1.8 m. His blood pressure was 130/80 mmHg without antihypertensive medication. The leg and abdominal ulcers were infected. Laboratory investigations revealed corrected calcium of 2.02 (N 2.2–2.6 mmol/l), serum phosphate of 2.59 (N 0.8–1.4 mmol/l), Ca × P of 5.9 (N 4.2–5.6 mmol/l), serum PTH of 248 (N 9.4–65 pg/ml), serum albumin of 34 (N 36–50 g/l), haemoglobin of 13.6 (N 13.1–16.6 g/dl), and random glucose of 5.9 mmol/l. His serum cholesterol was 9.1 (N < 5.2 mmol/l) and triglycerides 2.3 (N < 1.7 mmol/l).

Over the next month during his stay in the hospital he was provided with local wound care. Debridement and grafting of the necrotic skin areas were carried out as required. However, the ulcers did not show any sign of healing. The patient, by now, was quite unwell with worsening sepsis that was resistant to local wound care and systemic antibiotics. Thereafter, he developed an acute gastrointestinal haemorrhage. Disseminated intravascular coagulation was ruled out by the presence of normal prothrombin time, activated partial thromboplastin time and normal fibrinogen (clauss) levels. Despite the medical management he became increasingly disoriented and succumbed to his disease.

Discussion

High Ca × P, usually > 5.6 mmol/l, secondary to hyperparathyroidism of chronic renal failure, results in the deposition of crystals containing calcium phosphate in soft tissues and the tunica media of small to medium
sized arteries and arterioles. This is known as metastatic calcification and can lead to ischaemic tissue necrosis [1–4]. White race, female sex, type II diabetes mellitus, obesity, trauma, subcutaneous injections [5], protein C & S deficiencies, steroids and immunosuppressive therapy [6,7] have all been implicated. Our patient had several of the above predisposing factors. He was Caucasian with morbid obesity and had history of treatment with steroids and azathioprine.

Levin et al. [8] estimated the incidence of calciphylaxis as approximately one case per hundred haemodialysis patients per year. Angelis et al. [9] reported a prevalence of 4.1% in their dialysis unit.

Proximally localized lesions on shoulders, trunk, buttocks, and thighs have a poor prognosis mainly due to the larger bulk of subcutaneous tissue that can become necrotic and gangrenous. Distal lesions involving the forearms, calves, hands, fingers, feet, toes (acral sites), and genitalia have a better survival outcome [1–3, 6, 7]. Facial involvement, considered as distally localized, is rare and has not been documented in the listing of 377 cases on the database in the Medline search. In an extensive review by Hafner et al. [3], 63% of the patients with proximal lesions died as compared to 23% with distal localization. This explains the rapid downhill course in our patient who was relatively well until the development of the lesions on the proximal sites.

The role of parathyroidectomy is controversial [1–3,6,7]. Hafner et al. [1,3] demonstrated 70% survival benefit in parathyroidectomized patients before the onset of sepsis as compared with 43% of those without.

The role of hyperbaric oxygen therapy to improve the hypoxic conditions within the ulcers [10] and use of low molecular weight heparin needs further evaluation [11].

Prompt recognition of the condition is often delayed as calciphylaxis mimics other more common conditions like vasculitis, warfarin necrosis (it was considered a possibility in our patient), dermatomyositis, ischaemic changes due to cholesterol emboli, cryoglobulinaemia, etc. [1,2,4,6,7]. Lack of diagnostic investigations contributes to the delay as well. Skin biopsy shows calcification and atrophy of medial muscle in the arterial wall that is specific but not pathognomonic.

Early intervention to prevent the development of secondary hyperparathyroidism with an aim to prevent high Ca×P in patients suffering from chronic renal failure, greater awareness of the condition, and possible parathyroidectomy before the onset of sepsis may halt the progression of the disease and may prevent severe morbidity and mortality.

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References