A calcareous calamity

A 66-year-old woman had a successful renal transplant 4 years previously and was admitted to the hospital with an extremely painful purple skin abnormality of the right upper leg. During the preceding 6 months she suffered from progressive pain and weakness in all extremities, which had decreased her ability to walk. Ultrasound flow measurements of the large vessels and investigation by a neurologist had revealed no abnormalities.

On admission, general physical examination revealed no abnormalities, except a prominent dark-red/violet non-palpable skin lesion of 6 × 6 cm with a sharp margin, and surrounding livedo reticularis (Fig. 1). In both legs, arterial pulses were palpable, but capillary refilling time was prolonged.

Biopsy was taken from the centre of the skin lesion and showed a normal architecture of epidermis, but intimal proliferation with prominent calcification of a small arteriole (Fig. 2). X-rays of the extremities revealed impressive increase of calcification of large and small arteries as compared to the X-rays taken 1 year previously. Prominent vascular calcification was seen on routine mammography (Fig. 3).

Question

What is your diagnosis? (Answer on next page)
Answer to quiz case on preceding page: calciphylaxis

Routine laboratory investigation had revealed no abnormalities apart from slightly elevated serum ionized calcium and phosphate (1.38 mmol/l and 1.37 mmol/l, respectively). Serum creatinine was 200 μmol/l (unchanged compared to preceding measurements). The patient had a history of parathyroidectomy 5 years previously. Plasma PTH levels were not detectable. After cessation of calcium and vitamin D treatment, which had been given from the time of parathyroidectomy, ionized calcium and creatinine decreased to 0.59 mmol/l and 125 mmol/l, respectively. The skin biopsy was complicated by progressive local necrosis. Despite the use of several vasodilators there was no tendency towards wound healing. Several weeks later, spontaneous necrosis of the left big toe with marginal circulation of the skin of the forefoot developed. High dose oral morphine was needed to relieve pain. Treatment with hyperbaric oxygen was started. After 2 weeks of daily sessions with hyperbaric oxygen, a slight healing tendency of the ulcers was noted and the morphine dose could be reduced. The situation has gradually improved ever since.

Suggested reading:


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