Ulcerative lesions on the extremities with black eschar

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CASE

A 47-year-old Iranian man presented with a 3-month history of painful extremities lesions. The lesions began with an erythematous papule that progressively enlarged and ulcerated. His left calf was the first site of involvement. At the time we visited him, there were necrotic painful violaceous skin ulcers with a symmetrical distribution involving lower parts of distal limbs and his left thigh. He had a 10-year history of hypertension and a 5-month history of peritoneal tuberculosis. Recently, he received dialysis due to End Stage Renal Disease (ESRD). He had received medications such as Furosemide, Omeprazol, Isoniazid, Ethambutol, Rifampin and Pyrazinamide from 5 months ago. Physical examination showed a cachectic middle aged man in distress because of severe pain. His abdomen was distended, but no ascitis or organomegaly was detected. The pedal pulse was absent. Large multiple ulcers with surrounding erythema and induration, some with dark eschar, were seen on his calves, thighs and left hand (Figure 1). Some of his toes and two fingers of his left hand were gangrenous. A hand x-ray and a skin biopsy from the newest lesion were obtained. Some soft tissue opacities were seen in the hand x-ray (Figure 2).

What is your diagnosis?

Figure 1. Large multiple ulcers with surrounding erythema and induration, some with dark eschar, were seen on his calves, thighs and left hand.

Figure 2. Hand x-ray showed opacities in the soft tissue.
Diagnosis
Calciphylaxis

Microscopic findings

Microscopic study revealed a focal epidermal ulceration, parakeratosis, severe dermal infiltration, mixed inflammatory cells and granulation tissue formation, calcification of small and medium sized subcutaneous vessels accompanied by endothelial swelling and fibrin thrombi, compatible with the diagnosis of calciphylaxis (Figure 3).

Discussion
Calciphylaxis is a pathophysiological change which occurs due to deposition of calcium, resulting in painful skin lesions that rapidly progress to advanced non-healing ulcers and skin necrosis. It is an uncommon condition that occurs mostly in patients with ESRD who are on dialysis. They usually have hypercalcemia, hyperphosphatemia, and hyperparathyroidism in association with secondary hyperparathyroidism. In the case of a normal kidney function, calciphylaxis may be associated with primary hyperparathyroidism, rheumatoid arthritis, short bowel syndrome, breast cancer, Crohn’s disease, protein S deficiency, and acute renal failure in alcoholic cirrhosis. Several risk factors have been introduced for calciphylaxis such as the Caucasian race, female gender, obesity, diabetes, hypoalbuminemia. For diagnosis, clinical history and physical examination may be sufficient. The most accurate method to confirm the diagnosis is the biopsy of the ulcerated wound. Histological examination revealed medial calcification, and intimal fibrous hyperplasia of arteriolo, capillaries, and venules. X-ray of the lower extremities revealed extensive arterial calcifications within the arteries.

Treatment is mainly supportive. Elimination of the triggering factors is indicated, such as discontinuation of parenteral iron therapy, calcium, and vitamin D supplementation. According to the recent studies, some patients may benefit from systemic glucocorticoids in the early disease, before ulcerated lesions develop. Serum calcium and phosphate concentrations must be brought to the lower limit of normal as soon as possible. Conservative therapy with dietary alteration, use of non-calcium, phosphate binders and low-calcium bath dialysis should be tried first. Some patients benefit from increasing the frequency of dialysis sessions. In cases of hyperparathyroidism, calcimetics such as Cinacalcet hydrochloride may be beneficial. If response to conservative management is poor, parathyroidectomy should be considered. There are some reports of marked improvement in calciphylaxis with the use of intravenous sodium thiosulfate, which increases the solubility of calcium deposits. Careful use of antibiotics may be advantageous. Hyperbaric oxygen may be beneficial in some cases. The role of anticoagulant agents is controversial. Prophylactic use of warfarin or heparin is not indicated because precipitation of calciphylaxis may occur with random use of these medications.

Finally, we emphasize that calciphylaxis is a lethal disease with a high rate of morbidity and mortality. Our case died one month after the diagnosis of calciphylaxis, although vigorous conservative management was done.

References
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