Ulcerative lesions on the extremities with black eschar

Iran J Dermatol 2011; 14: 37-39

Mohammad Shahidi-Dadras, MD
Somayeh Hejazi, MD
Azin Ayatollahi, MD
Mohammad Saeedi, MD

Skin Research Center, Shahid Beheshti Medical University, Shohada-e-Tajrish Hospital, Tehran, Iran.

Corresponding Author:
Azin Ayatollahi, MD
Skin Research Center, Shahid Beheshti Medical University, Shohada-e-Tajrish Hospital, Tehran, Iran
Email: azinay@gmail.com

Conflict of interest: None to declare

Received: October 08, 2010
Accepted: October 31, 2010

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 (Figure2).

What is your diagnosis?

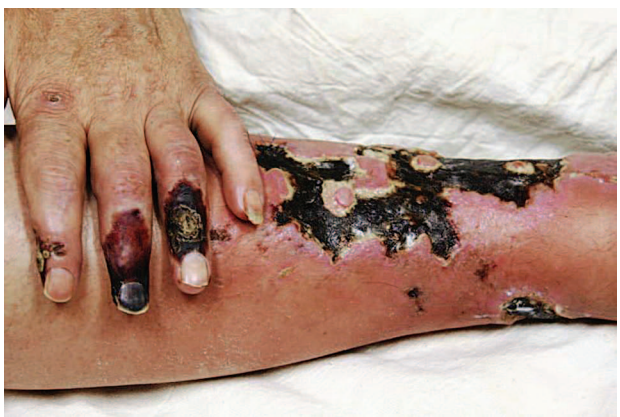

Figure 1. Large multiple ulcers with surrounding erythema and induration, some with dark eschar, were seen on his calves, thighs and left hand.

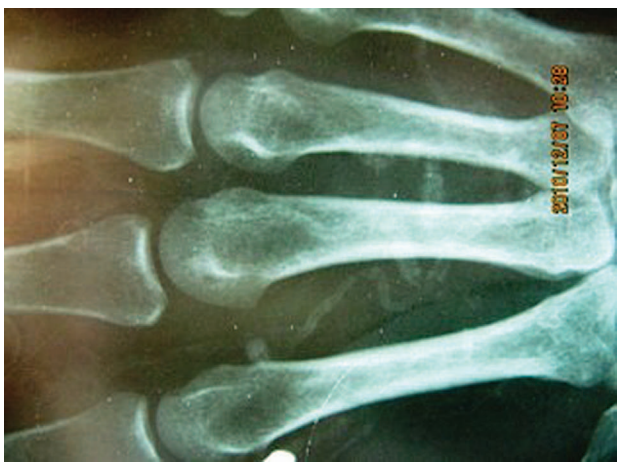

Figure 2. Hand x-ray showed opacities in the soft tissue.

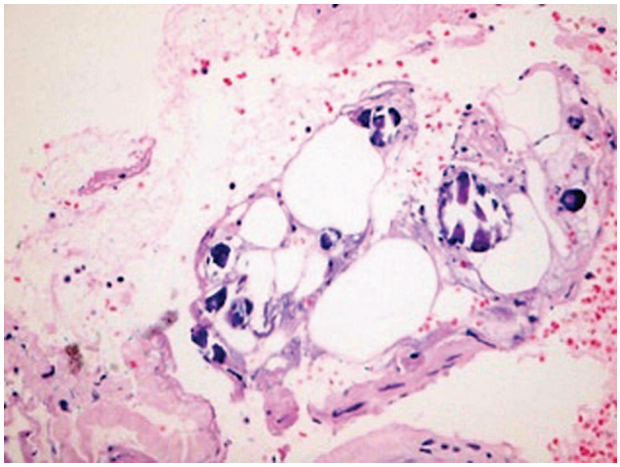

Figure 3. Histopathological view; Calcification of small and medium sized subcutaneous vessels accompanied by mixed dermal infiltration (H&E*40).

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 [1]. 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 [2-6]. Several risk factors have been introduced for calciphylaxis such as the Caucasian race, female gender, obesity, diabetes, hypoalbuminemia [7]. 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 arterioles, 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 glucocorticoides 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, calcimimetics 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 [8-12].

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

1. Kang AS, McCarthy JT, Rowland C, Farley DR, van Heerden JA. Is calciphylaxis best treated surgically or medically? Surgery 2000;128:967-72.
2. Goyal S, Huhan KM, Provost TT. Calciphylaxis in a patient without renal failure or elevated parathyroid hormone: possible aetiological role of chemotherapy. Br J Dermatol 2000;142:1087-90.
3. Golitz LE, Field JP. Metastatic calcification with skin necrosis. Arch Dermatol 1972;106; 398-402.
4. Bogdonoff M, Woods A, White J, Engel F. Hyper-

parathyrodism. Am J Med 1956;21:583-95.

5. Friedman M, Marshall-Jones P, Ross EJ. Cushing's syndrome: adrenocortical hyperactivity secondary to neoplasms arising outside the pituitary-adrenal system. Q J Med 1966;35:193-214.
6. Barri YM, Graves GS, Knochel JP. Calciphylaxis in patient with Crohns disease in the absence of end stage renal disease. Am J Kidney Dis 1997; 29:773-6.
7. Arch-Ferrer JE, Beenken SW, Rue LW, Bland KI, Diethelm AG. Therapy for calciphylaxis: An outcome analysis. Surgery 2003;134:941-5.
8. Dereure O, Leray H, Barneon G, Canaud B, Mion C, Guilhou JJ. Extensive necrotizing livedo reticularis in a patient with chronic renal failure, hyperparathyroidism and coagulation disorder: Regression after subtotal parathyroidectomy. Dermatology 1996;192:167-70.
9. Mehta RL, Scott G, Sloand JA, Francis CW. Skin necrosis associated with acquired protein C deficiency in patients with renal failure and calciphylaxis. Am J Med 1990;88:252-7.
10. Leblanc M, Roy LF, Legault L, Dufresne LR, Morin C, Thuot C.Severe skin necrosis associated with heparin in hemodialysis. Nephron 1994;68:133-7.
11. Elamin EM, McDonald AB. Calcifying panniculitis with renal failure: A new management approach. Dermatology 1996; 192:156-9.
12. Guerra G, Shah RC, Ross EA. Rapid resolution of calciphylaxis with intravenous sodium thiosulfate and continuous venovenous haemofiltration using low calcium replacement fluid: Case report. Nephrol Dial Transplant 2005;20:1260-2.