Not all Calciphylaxis is due to Renal Disease – A Rare Presentation of "Non-Uremic" Calciphylaxis

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Introduction

Calciphylaxis, known as calcific uremic arteriopathy (CUA), is classically found in patients with end stage renal disease and advanced chronic kidney disease. This condition has a high morbidity and mortality rate as it causes abnormal calcium deposition in vessels resulting in vascular thrombosis and tissue infarction. "Non-uremic" Calciphylaxis is rare and often overlooked as a diagnostic consideration in patients with normal functioning kidneys. We present a patient with non-uremic calciphylaxis and co-morbid illnesses including anti-phospholipid antibody syndrome (APLS) on anticoagulation with Coumadin

Case Presentation

A 37-year-old Caucasian female with a past medical history of APLS, diabetes mellitus type 2, depression, and chronic opioid use was admitted for pain management for a recurrent abdominal wound that recently underwent debridement. The patient had a Jejunostomy feeding tube (J-Tube) insertion six months prior. Shortly after the procedure, she underwent a surgical wound debridement of her abdomen (Image 1), lower back, and pelvis. She had regular follow- up with wound care. Her pain was difficult to control despite daily opioid use. She had also been placed on Coumadin for antiphospholipid syndrome. Prior to admission, a biopsy of the abdominal wound showed subcutaneous medium-sized blood vessels with medial circumferential and near occlusive calcification and soft tissue calcification consistent with calciphylaxis.

On admission, her serum creatinine was noted to be 1.50 mg/dL from her baseline of 1.0 mg/dL. Her mild acute kidney injury and electrolyte disturbances quickly resolved with tube feeds and intravenous fluids. Nephrology and dermatology were consulted. The patient was started on treatment with sodium thiosulfate infusion after port placement as well as medication for pain control. She was discharged after changing the wound VAC on her abdomen and maintained on sodium thiosulfate post hospital-discharge. She was instructed to follow up with dermatology, nephrology, palliative care, and wound care.

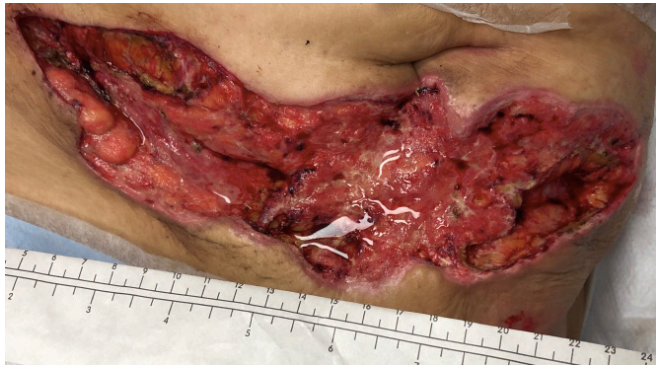

Image 1: Soft tissue debridement from the abdomen consisting of irregularly shaped fragment of soft tissue with presumed overlying skin measuring 24x7.5 cm showed portion of extensively necrotic skin and subcutis with sloughed epidermis. Also noted subcutaneous medium-sized blood vessels with medial circumferential and near occlusive calcifications and soft tissue calcifications consistent with calciphylaxis

Conclusion

Calciphylaxis needs to remain in the differential when evaluating deep and superficial lesions even in patients with normal kidney function. Among the few reports of nonuremic calciphylaxis, cases cited were primarily among Caucasian women reporting various co-morbid illnesses including malignancy, hyperparathyroidism, protein C and S deficiencies, mineral abnormalities, diabetes, and autoimmune conditions. The pathogenesis of calciphylaxis is likely due to a contribution of many factors. In our patient, we suspect that antiphospholipid antibody syndrome could have contributed to the development of calciphylaxis. In addition, Coumadin anticoagulation increased the likelihood developing of warfarin-induced skin necrosis leading to calciphylaxis. While the incidence of nonuremic calciphylaxis is low, the high patient mortality rate highlights the necessity for better understanding the pathophysiology of this rare condition so that we can better improve our treatment for it.

References

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