

CALCIPHYLAXIS FROM NON-UREMIC ETIOLOGIES: A SYSTEMATIC REVIEW

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OBJECTIVE: Calciphylaxis or calcific uremic arteriopathy (CUA) is a well described entity in end stage kidney disease (ESKD) and renal transplant patients. However, little systematic information is available on calciphylaxis from non-uremic etiologies. This systematic review was designed to characterize etiologies, clinical features, laboratory abnormalities and prognosis of non-uremic calciphylaxis (NUC).

METHODS: We performed a systematic review of MEDLINE, Ovid, and Google for case reports and case series of NUC. Cases included met the operational definition of NUC-histopathological diagnosis of calciphylaxis in the absence of ESKD, renal transplantation or acute kidney injury requiring renal replacement therapy.

RESULTS: We found 36 cases (75% females, 63% Caucasian, age 15-82 years) of NUC reported in the literature. Primary hyperparathyroidism (n=10), malignancies (n=8), alcoholic liver disease (n=7), and connective tissue diseases (n=4) were the most common reported etiologies. Others included diabetes mellitus (n=2), vitamin D deficiency (n=1), nadroparin calcium (n=1), chemotherapy (n=1), POEMS syndrome (n=1) and severe weight loss (n=1). Preceding corticosteroid use was reported in 61% of patients. Protein C and S deficiencies were seen in 11% of patients. Approximately 1/3rd of the skin lesions were proximal, 1/3rd distal and 1/3rd were both proximal and distal. Morphology of skin lesions was similar to CUA. Only minority of patients with NUC had elevated serum phosphorous or calcium-phosphorous product. There were no clearly effective treatments and mortality rate was 52% with sepsis being the leading cause of death.

CONCLUSION: NUC is a well reported entity. Calciphylaxis should be considered while evaluating skin lesions in patients with predisposing conditions even in the absence of ESKD and renal transplantation. NUC is reported most often in Caucasian women and characteristic laboratory abnormalities that are seen in CUA may not be present. NUC is associated with high mortality and there is no known effective treatment.