

Calciphylaxis and vascular calcification: clinical picture and new data

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Calciphylaxis (calcific uremic arteriopathy; CUA) is a rare and devastating condition associated with high morbidity and mortality predominantly observed in patients on dialysis or with advanced chronic kidney disease (CKD). Calciphylaxis is characterised by very painful necrotic skin ulcerations, histomorphologically characterized by severe media calcification of cutaneous arterioles, nerve sheaths and fat tissues. The differential diagnosis may include cutaneous vasculitis, pyoderma gangraenosum, diabetic ulcers and cholesterol emboli. Until recently, calciphylaxis was thought to be tightly linked to severe hyperparathyroidism, but this perception must be revised based on novel registry findings in which low parathyroid hormone levels were associated with the majority of cases. Factors including a positive calcium and phosphate balance, adynamic bone turnover, inflammation, calcification inhibitor deficiencies and vitamin K antagonism may rather play causal roles as triggers of this disease. Novel therapeutic options (e.g., cinacalcet, bisphosphonates, sodium thiosulfate, vitamin K2 or K1) are in clinical use, however, given the orphan nature of calciphylaxis no randomized controlled trials are available on these therapeutics. Based on a case report, the lecture will review the currently available knowledge on the pathophysiology and clinical management of the syndrome and present recent data from two registries.