Disorders of the parathyroid glands, calcium metabolism and bone

CALCIPHYLAXIS SECONDARY TO VITAMIN D DEFICIENCY

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Introduction: Calciphylaxis is an uncommon disorder characterised by small-vessel calcification leading to skin ischaemia and skin and soft tissue necrosis. It most commonly occurs in the setting of end-stage renal failure, but has also been reported in patients with primary and secondary hyperparathyroidism, cirrhosis of the liver, cholangiocarcinoma, and breast cancer. We report a case of calciphylaxis in a patient without renal failure, who had secondary hyperparathyroidism as a result of Vitamin D deficiency, and responded well to oral replacement of Vitamin D, without the need for parathyroidectomy.

Case report: A 77-year-old woman presented with a painful, non-healing ulcer over her calf, developing over 6 weeks. Relevant past medical history consisted of ischaemic heart disease, diabetes mellitus, and atrial fibrillation. The patient was on warfarin for thrombo-prophylaxis. Examination revealed an irregular ulcer above the lateral malleolus, with well-defined edges, measuring 10cm x 5cm. Initial review led to a differential diagnosis of vasculitic ulcer with possible superimposed infection. Biochemical tests were negative for vasculitis, and the patient therefore underwent a biopsy of the ulcer to reach a conclusive diagnosis. The histology showed appearances consistent with calciphylaxis. We checked her PTH levels and found them to be elevated (13.8 pmmol/L), with normal Ca\textsuperscript{2+} and PO\textsubscript{4}\textsuperscript{3-} levels. Further assessments showed her to be deficient in vitamin D. Although an isotope scan revealed a solitary left sided parathyroid adenoma, we felt that a parathyroidectomy would not be without its own risks, given the patient’s co-morbidities. We therefore opted to treat the vitamin D deficiency first, and the patient responded well to oral supplementation, with resolution of the ulcer and good symptom relief.

Discussion: The pathogenesis of calciphylaxis is not well understood, but is likely to be the end result of several factors acting together. Abnormalities of calcium homeostasis, coagulation defects, and defects in inhibitors of mineralisation have all been implicated. Patients at risk of calciphylaxis include white race (especially women), morbid obesity, warfarin administration, recent severe weight loss, low serum albumin, and an elevated Ca x PO\textsuperscript{4} product. Our patient probably developed calciphylaxis as a result of secondary hyperparathyroidism caused by Vitamin D deficiency. It is also possible that warfarin acted as an additional factor in contributing to her calcium imbalance. Most studies and case reports in this field till date, recommend total parathyroidectomy as the treatment for this condition, especially in the face of elevated PTH. Other recommendations include appropriate wound care and maintenance of hygiene, use of antibiotics where needed. The patient was successfully discharged home once her ulcers healed with normalisation of PTH, and regular follow-ups have revealed no recurrence of the ulcer.