Severe Digital Calcinosis in CREST Syndrome

Shakeel Anjum*, Donough Howard and Paul O’Connell
Department of Rheumatology, Beaumont Hospital, Ireland

A 65 year old lady with diagnosis of CREST for last 10 years presented with progressive right sided unilateral digital calcos\-nosis particularly worse in her thumb, middle and little fingers. She also had areas of inflammation around calcinosis. This resulted in significant disability as she was right-handed. She was treated with colchicine which resulted in improvement of inflammation and pain.

Calcinosis is common manifestation of CREST Syndrome. These calcium deposits are located in soft tissues, without causing direct joint involvement, and may be small or large and usually complicated by skin ulceration and superimposed infection. The main pathophysiological mechanism for the development of calcinosis in scleroderma is tissue hypoxia [1].

The decrease in perfusion is accompanied by inflammatory cell activity and macrophage activation as well as an imbalance between various mediators that cause increased calcium influx to cells [1]. Patients with extensive calcinosis exhibit significant impairment of functional capacity due to flexion contractures of the adjacent joints. Regarding the treatment of calcinosis, no treatment is postulated to be more effective than another. Therapeutic alternatives include: warfarin, diltiazem, colchicine, probenecid, bisphosphonates, minocycline, aspirin, corticosteroid intrale\-sional injections, aluminum hydroxide, IVIG, iontophoresis and ultrasound [1,2]. Some authors suggest initiating therapy with diltiazem and even combining drugs [2]. Importantly, the severity of calcinosis is not related to the severity of the underlying disease [2]. Ulcerated calcinosis should be properly observed due to the possibility of bacterial superinfection. Surgical resolution may be considered in cases of extensive deposits of calcium that cause flexion contractures or impaired joint function but remembering that these injuries tend to become infected and are difficult to heal. For smaller localized lesions, CO2 laser and extracorporeal shock wave techniques can be useful [1,2].

REFERENCES
