

Introduction

Systemic sclerosis is a disease with multisystem organ involvement diagnosed primarily from clinical findings with support from serologic abnormalities. Further breakdown and classification of this systemic disease include diffuse cutaneous (dcSSc) and limited cutaneous (lcSSc). LcSSc includes skin fibrosis and features of CREST syndrome; calcinosis cutis, Raynaud phenomenon, esophageal dysmobility, sclerodactyly, and telangiectasia.

Calcinosis cutis occurs in about 15% of patients with dcSSc and 44% of patients with lcSSc and can be intracutaneous or subcutaneous. Calcium deposits may lead to severe pain and functional deficits in the distal extremities, most commonly in the hand but also in the lower legs and feet. Vascular compromise, and therefore decreased cutaneous circulation, along with skin fibrosis leads to thin, fragile skin susceptible to wound development.

Conservative treatments include medications such as calcium channel blockers, methotrexate, bisphosphonates, and warfarin, all with variable success rates. Surgical excision is typically required in severe or widespread cases, typically resulting in success of pain relief and return to normal function.

Case Presentation

A 69 year old female with past medical history of systemic scleroderma and rheumatoid arthritis, managed with oral steroids and immunosuppressive therapy, presented with a right lower extremity leg wound initially in January of 2017 after suffering trauma to the area. Infection and worsening of the wound led to surgical debridement, resulting in the discovery of subcutaneous calcified tissue throughout the entire right anterior lower leg. The patient healed uneventfully with local wound care but returned a year later in January of 2018 with similar symptoms and wounds to bilateral lower extremities. Surgical intervention was again performed with a vast amount of calcification removed from the subcutaneous tissue. At this time, a correlation was noted between the development of calcified soft tissue and decrease in the patient's oral steroid medications.

Multidisciplinary approach was initiated with coordination between rheumatology and internal medicine to determine a threshold dose of steroid to prevent further recurrence of calcinosis cutis.

Discussion

The patient underwent a third surgical wound debridement in March of 2018, 2 months after adjustment to oral medications and steroid therapy. Minimal calcified soft tissue was encountered.

Bi-weekly follow up with local wound care and allograft application was performed following the last surgical intervention with vast improvement in decreasing the size and depth of the patient's bilateral lower extremity wounds. Her threshold dose of oral steroid has been maintained, and supplementary topical steroid, Solu-cortef, is applied to each wound with dressing changes.

Topical wound therapy also included various topical collagen products and allografts. Multi-layer compression wraps were utilized to control lower extremity edema.

To date, the patient continues to improve in pain level and return to normal function and daily activities, and she has not suffered from any recurrence of calcinosis cutis to the lower extremities.



Figures from May 10, 2018 (left to right) : Right anterior leg, Left Achilles, Left lateral leg



Figures from June 14, 2018 (Left to Right) : Right anterior leg, Left Achilles, Left lateral leg

Conclusion

Calcinosis cutis is a complication of systemic scleroderma, typically seen in the later stages of diagnosis. Soft tissue and skin quality is affected by numerous complications of the disease including decreased circulation, edema, fibrosis, and development of subcutaneous or intracutaneous calcification. In later stages, the disease and its complications can be painful and debilitating, necessitating dual surgical and medical treatment. Along with surgical excision or curettage comes long-term wound care follow up as well as systemic medication adjustment in a multidisciplinary approach. With recognition of the catalyst of initial calcification development, patients may be spared relapse and maintain a better quality of life.



Figures from October 25, 2018:

Left : Right anterior leg
Middle : Left Achilles
Right : Left medial leg

Resources

Davis, MDP, Anderson, LK, Wetter DA, Balin SJ. Calcinosis Cutis Occurring in Association with Autoimmune Connective Tissue Disease; The Mayo Clinic Experience with 78 Patients. Arch Dermatol Apr 2012 (4): 462-455.

LeRoy, EC, Medsger TA Jr. Criteria for the classification of early systemic sclerosis. J Rheumatology. 2001; 28:1573-1576.

Nalebuff EA, Axford JS, Curtis MR. Surgery in Patients with Systemic Sclerosis (Scleroderma) of the Hand. Jul 2018. www.uptodate.com

Peytrignet, S, et al. Disability, Fatigue, Pain, and Their Associates in Early Diffuse Cutaneous Systemic Sclerosis: the European Scleroderma Observational Study. Rheumatology 2018; 57: 370-381

Valenzuela A, et al. Calcinosis Associated with Digital Ulcers and Osteoporosis in Patients with Systemic Sclerosis: A Scleroderma Clinical Trials Consortium Study. Semin Arthritis Rheum. 2016 December; 46(3) 344-349

Varga, J, Axford JS, Curtis MR. Diagnosis and Differential Diagnosis of Systemic Sclerosis (Scleroderma) in Adults. Jul 2018. www.uptodate.com

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