



Treatment of Calcinosis Cutis Secondary to Juvenile Dermatomyositis with Intralesional Sodium Thiosulfate Injections

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KEYWORDS: dermatomyositis, STS, calcinosis cutis

Learning Objectives

1. Calcinosis cutis (CC) as a complication of juvenile dermatomyositis (JDM)
2. Sequelae of CC
3. Sodium thiosulfate (STS) injections for treatment of severe, refractory CC

Case Description

A 23-year-old female with a 16-year history of JDM categorized by elevated creatinine kinase (2400units/L), lactate dehydrogenase (404units/L), AST (145units/L), ALT (102units/L) and aldolase (25units/L); myositis, dysphagia, lipodystrophy, Gottron's papules and dystrophic calcinosis cutis (CC), was referred for management of subcutaneous calcification of the hands and elbow (**Figure 1A**). Her JDM was well controlled on abatacept (750mg IV qmonth), colchicine (0.6mg PO tid), hydroxychloroquine (200mg PO bid), immune globulin intravenous (IVIG) (2g/kg per month) and

methylprednisolone (500mg IV qmonth). Calcinosis was the most troubling manifestation of her JDM at the time which developed 10 years post diagnosis and progressed rapidly to involve much of her body, especially joints. Pain from these nodules proved difficult to manage.

Over a 6-year period, treatment with methotrexate, rituximab, IV pamidronate, diltiazem, mycophenolate mofetil, tacrolimus, topical creams and intralesional steroid injections failed. As her condition worsened, she developed depression with suicidal ideation, and her quality of life diminished.

Discussion

CC is the deposition of insoluble calcium crystals in the skin and subcutis^{1,2}. It is often precipitated by underlying autoimmune connective tissue diseases (ACTDs) like JDM, where CC can be severe³.

Sequelae of CC include pain, ulceration, functionality loss and infection.^{1,2,3} While aggressive management of ACTDs has improved symptom development, treatment remains



Figure 1. A (top panel) Initial presentation of hands and right elbow (05/07/2020); firm yellow nodules with induration and ulceration.

B (bottom panel) After four rounds of intralesional STS injections to the hands and three to the elbow (06/19/2020); decreased size and number of calcified nodules.

challenging as no current guidelines exist^{1,2,3}. Options include calcium channel blockers, colchicine, IVIG, bisphosphonates, methotrexate, rituximab, cyclophosphamide, surgical interventions and STS^{1,3}.

STS is an inorganic salt with antioxidant, reducing and chelating properties used to treat cyanide poisoning and calcium-related disorders^{4,7}. It has many proposed mechanisms including formation of soluble calcium thiosulfate and stimulation of vasodilation via antioxidant and reducing properties, restoring endothelial function^{4,5,7}. Although originally delivered topically and intravenously, diffuse distribution of lesions and poor side effect profiles initiated intralesional injections¹⁻⁴. This improves accessibility of the target, lowers complication risk and systemic effects.⁴ Limitations include inability to treat lesions deep to soft tissue and local pain or infection^{4,6}. There are no contraindications for STS injections, but side effects include hypotension, headache and disorientation⁸.

Five mL of 250 mg/mL of STS mixed in normal saline were injected into lesions on the right and left middle metacarpophalangeal joints and right index finger distal interphalangeal joint. Two weeks later, the treated areas softened and decreased in size. The same protocol was repeated with three additional injections to the elbow. In total, four rounds of STS were injected into the hands and three into the elbow in two-week intervals. Nodules improved with decreases in size, pain and rigidity, improving range of motion and joint mobility (**Figure 1B**). Our patient was happy with the results but did experience pain for 48 hours post-injection. While this treatment significantly improved CC symptoms, other JDM manifestations remained unchanged.

Mounting evidence describing successful response to intralesional STS in individual CC patients should be an impetus for controlled studies on the efficacy of this treatment. Through its multifactorial mechanism, STS reduces calcium deposition, alleviates pain and promotes healing. This quick outpatient treatment can provide significant relief and improve quality of life.

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Author Contributions

All authors have given approval to the final version of the manuscript.

Funding Sources

All authors received no financial support for the research, authorship, and/or publication of this article.

Disclosures

No authors have any disclosures or conflicts of interest at this time.

Acknowledgements

None.

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