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Morphea Affecting Bilateral Ankles

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Introduction

Morphea, also known as localized scleroderma, is an inflammatory disorder causing patches of hardened or discolored skin and subcutaneous tissue and has an incidence of about 0.4-2.7 per 100,000. The five main types of morphea include linear, plaque, generalized, deep, and bullous. More severe forms can lead to devastating functional and cosmetic impairment due to excessive collagen deposition in the deeper subcutaneous tissue. Though thought to have an autoimmune component, there are no clear immunological markers associated with morphea. Our case highlights the difficult treatment course and possible correlation between morphea and autoimmune diseases.

Clinical Presentation

65-year old female presented to the podiatry clinic on 12/21/2018 complaining of increased swelling, pain, and skin darkening to bilateral dorsal ankles and stiffening of her ankle joints for two months. Patient stated her ankles felt "raw and painful". The patient also noted that she had similar hyperpigmented plaques on her back, neck, and upper extremities.

Past Medical History: Type 2 diabetes mellitus, hypertension, hypothyroidism, arthritis, and Sjogren's syndrome.

Past Surgical History: Bunion and hammertoe corrective surgery left foot in 2014

Medications: Metformin, Hydrochlorothiazide, Levothyroxine, and Restasis.

Family History: Sister with Raynaud's syndrome

Diagnosis

Physical exam revealed the patient was neurovascularly intact. No local or generalized signs of vasculitis were noted. Patient did have pain on palpation of the anterior aspects of bilateral ankles. Localized edema 2+ was present to bilateral ankles.

Lab work performed previously by her rheumatologist revealed she was ANA+, SSA+, SCL70-. The lesion on her neck had been biopsied and showed dermal sclerosis consistent with morphea. It was then determined clinically that the skin lesions affecting her ankles were consistent with those on her neck and she was given the diagnosis of generalized morphea.

Images



Figure 1: Dorsal aspect of left foot



Figure 2: Lateral aspect of left foot



Figure 3: Medial aspect of right foot



Figure 4: Medial aspect of left foot

Treatment

Patient was initially started on the immunosuppressive drug CellCept (mycophenolate mofetil) 500mg TID in 2018 by her rheumatologist. She was then seen by a dermatologist in 2019 who added topical clobetasol, UVA1 phototherapy once a week, and physical therapy to her regimen.

Results

Over the course of two years, our patient's morphea, which presented in a generalized pattern, flared into a more bullous form. Her CellCept has been increased over time from 500 mg TID to 1500 mg bid. Then in January 2020, the patient then began complaining that the skin hardening was spreading and that she had more difficulty walking. She was admitted to the hospital to start a high-dose steroid regimen of SoluMedrol and was later discharged on prednisone and with instructions to continue her previous therapies. The patient reported some resolution of symptoms after her most recent hospital encounter. She continues to follow up with her rheumatologist, dermatologist, and podiatrist.

Conclusion

This case demonstrates the importance of a multi-disciplinary approach to this lower extremity pathology. Though the exact cause of morphea is unknown, the literature suggests there is an increased incidence in patients with concurrent autoimmune disease; Sjogren's syndrome in this case. Therefore, having the expertise from multiple disciplines available to diagnose and treat these possibly related disease was greatly beneficial. By having various specialties involved, a wide range of treatment modalities were available to help alleviate her symptoms.

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