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Case Report

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Limited Cutaneous Systemic Sclerosis Presenting with Corneal Melt: A Complication of Uncontrolled Disease

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Abstract

Limited cutaneous systemic sclerosis is an autoimmune condition with an abnormal proliferation in connective tissue proliferation. Corneal melt is caused by breakdown of corneal collagen which may be caused by various etiologies including inflammatory, infectious, and trauma have been linked to corneal melt. Corneal melt is rarely caused by autoimmune conditions including rheumatoid arthritis, vasculitis, systemic lupus erythematosus, relapsing polychondritis, and systemic sclerosis. We present an 83-year-old female hispanic patient with history of peripheral vascular disease who presented with corneal melt. Patient's clinical history as well as labs, specifically anti-centromere antibody were consistent with limited cutaneous systemic sclerosis. Patient was started on treatment with Rituximab for the management of corneal melt associated with systemic sclerosis, which resulted in improvement of the corneal melt as well as symptoms of systemic sclerosis. Careful attention needs to be placed on the etiology of the corneal melt in order to prevent disease progression. The key to treatment of corneal melt is treatment of the underlying cause. Appropriate treatment and identification of corneal melt is paramount to prevention of permanent visual loss.

Keywords

Corneal Melt, Limited cutaneous systemic sclerosis, Sclerosis, Connective tissue proliferation, Raynaud's

Introduction

Corneal melt is caused by corneal collagen breakdown that may lead to serious ocular complications such as irregular astigmatism, decreased vision which may lead to blindness. Corneal melt may be caused from autoimmune conditions such as rheumatoid arthritis, systemic lupus erythematous, and very rarely seen with Sjogren's disease and systemic sclerosis [1]. Systemic sclerosis is a chronic disease caused by abnormal and widespread proliferation of connective tissue leading to multisystem involvement. Limited systemic sclerosis is a type of sclerosis that is associated with distal puffiness of fingers and skin sclerosis, along with symptoms such as of calcinosis cutis, Raynaud's phenomenon, esophageal dysmotility, sclerodactyly, and telangiectasia [2]. Corneal melt may be caused by various causes including infection, trauma, infection, malignancy, autoimmune causes, or paraneoplastic syndromes. Complete history as well as laboratory workup needs to be done to identify the cause of corneal melt. Paraneoplastic syndromes may

manifest with corneal melt, therefore in patients presenting with corneal melt it is imperative to rule out malignancy as the cause.

Case presentation

We present the case of an 83-year-old hispanic female with past medical history of peripheral artery disease and deep venous thrombosis presenting for evaluation of corneal melt. The patient was in her usual state of health until a corneal melt was detected which led to corneal ulceration of the right eye requiring grafting. Despite treatment with oral steroids for three months the patient experienced melting of the cornea through the graft. The patient did not have any prior history of trauma or infection. Paraneoplastic syndrome was ruled out as she was up to date with all age-appropriate cancer screening and did not have any clinical evidence to suggest malignancy. The patient was referred to rheumatology by ophthalmology as there was concern for an underlying autoimmune condition as the etiology of the corneal melt. Prior to referral to rheumatology workup for autoimmune conditions was not started prior to initial corneal grafting. Patient admitted to Raynaud's phenomenon, telangiectasias, eye pain, and puffiness of fingers. She denied any gastroesophageal reflux disease, dysphagia, joint pain, dry eyes, or dry mouth.

On physical exam vitals were within normal limits. Patient had decreased vision of the right eye with conjunctival erythema. Her vision in the right eye was 20/160 which was decreased prior to grafting. Skin tightening was noted distal to the proximal interphalangeal joints with nonpitting puffiness of the fingers, additionally no synovitis was noted (Figure 1). Patient had normal complete blood count and comprehensive metabolic panel. She was found to have high titer antinuclear antibody 1:320, normal values are less than 1:80. Anti-centromere level, detected by enzyme-linked immunoassay, was greater than 240 U/mL, normal anti-centromere level is less than 7 U/mL. Erythrocyte sedimentation rate and C-reactive Protein, which signify systemic inflammation were normal. Given patients significantly elevated anti-centromere antibody in setting of clinical findings of limited cutaneous systemic sclerosis, treatment with Rituximab infusion was initiated to treat this underlying autoimmune condition in order to improve the corneal melt. Treatment with Ritxumab was initiated with a scheduled cycle of every 8 weeks. This patient received three infusions of rituximab with improvement of corneal melt, decreased inflammatory activity of the eye, and no further melting of the corneal graft.

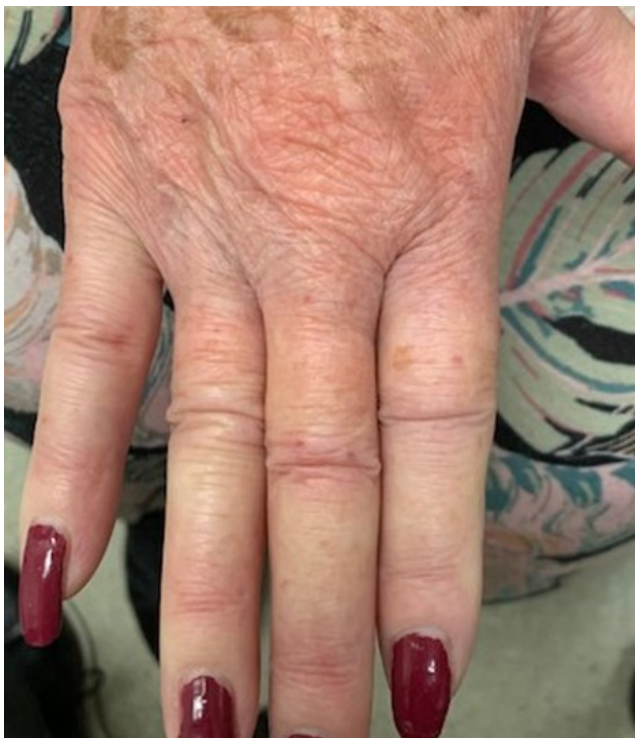

Figure 1: Tightening and thickening of the skin distal to the proximal interphalangeal joints.

Discussion

Corneal melt may be cause by autoimmune, infectious, chemical burns, or trauma. The proposed pathophysiology of corneal melt secondary to autoimmune conditions is the abnormal and progressive destruction of the corneal collagen, due to the significant inflammation of the corneal extracellular matrix [3]. This process is likely due to the increased inflammatory cytokines which cause corneal cells to produce an increased number of proteolytic enzymes which degrade the corneal extracellular matrix. Several studies have shown the corneal melting is influenced by excessive tissue degradative proteases, particularly matrix metalloproteinases [3, 4]. The usual presentation of corneal melt was monocular visual disturbance with pain, middle age, with equal distribution in females and males [3].

In autoimmune causes of corneal melt, treatment of the underlying autoimmune condition is key. Treatment of corneal melt is with systemic immunosuppressive therapy, specifically with biologic therapy [5]. Biologic therapy is a type of treatment that uses substances from living organisms. Most patients with corneal melt will require a surgical approach such as grafting along with pharmacological treatment in inflammatory conditions to suppress the immune system [3, 5]. Rituximab is not first line treatment in patients with limited cutaneous systemic sclerosis and is typically only initiated when there is organ involvement. First line treatment for systemic sclerosis is supportive treatment, followed by specific agents targeting the involved organ. In this case the patient was started on Rituximab given the corneal melt.

This case highlights the importance of identifying underlying etiology of disease manifestations in order to initiate proper treatment. There does not appear to be any published cases of corneal melt associated with systemic sclerosis. In this case, other etiologies of corneal melt such as trauma, infection, or malignancy were ruled out. The patient's autoimmune condition causing the corneal melt, systemic sclerosis, was treated with systemic immunosuppressive therapy. Timely treatment of corneal melt is critical, as this condition progresses rapidly and when left untreated may lead to permanent visual loss.

Disclosure statement

The authors have no conflict of interest of acknowledgements.

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