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ABSTRACT

The rare, non-caseating, granulomatous disease known as Melkersson-Rosenthal syndrome (MRS) can be diagnostically difficult especially when not presenting as the characteristic triad of facial paralysis, facial edema, and fissured tongue.¹ We present a case of a 65-year-old female with a 15-year history of recurrent bilateral upper eyelid swelling previously unsuccessfully treated for a presumed lid allergy and rosacea. Following imaging and histopathology results, she has now been diagnosed with Melkersson-Rosenthal syndrome. Since this disease is typically refractory to treatment, she is being followed and treated with cosmetic eyelid skin reduction if necessary during inactive stages of the disease.

Key Words: Melkerson-Rosenthal syndrome, non-caseating granuloma, eyelid swelling, eyelid inflammation.

INTRODUCTION

Melkersson-Rosenthal Syndrome (MRS) is a rare condition of unknown etiology. We present a case of this disease to demonstrate the need to include this in the differential diagnoses of eyelid edema.

CASE REPORT

A 65 year-old Caucasian female with no family history and consanguinity, presented with a 15-year history of non-pitting, non-pruritic, recurrent bilateral upper eyelid swelling worse on the right side. Seven years prior, she was given a diagnosis of rosacea and lid allergies. She has been treated unsuccessfully with oral anti-allergy medications, topical antibiotics and steroid ointments. For the last year, she has had a complaint of a mildly pruritic facial rash that worsen in sunlight.

Examination demonstrated a mild right mechanical upper ptosis and bilateral upper lid swelling more prominent on the right side (Figure 1). Her skin was brawny in color with no signs of pain, itchiness or sensory/motor defect. No significant laboratory findings were present. Her head-orbit CT was only significant for right eye preseptal soft tissue swelling (Figure 2). After excision biopsy, the histopathology showed dermal edema, ectatic vessels, and granuloma formation near dilated vessels. In addition, scattered interstitial and perivascular lymphocytes were present as well as intravascular histiocytes which stained with CD68 (Figures 3 and 4).

Due to the disease process and specifically the histopathology, her diagnosis is Melkersson-Rosenthal Syndrome. The current treatment will be observation for the underlying disease state. She will be offered upper lid skin reduction if the disease progresses.

FIGURE 1: Clinical photography of a 65-year-old Caucasian female demonstrating predominately right-sided upper eyelid non-pitting swelling and mechanical ptosis secondary to Melkersson-Rosenthal syndrome.
DISCUSSION

MRS is a rare chronic condition that has relapsing-remitting course involving painless, non-pitting orofacial swelling. It may exist as a triad of symptoms including orofacial edema, relapsing facial palsy, and fissured tongue; however, this triad need not exist for the diagnosis to be made. Typically, the remainder of the ophthalmic examination is normal. The differential diagnosis includes thyroid-associated orbitopathy, angioedema, sarcoidosis, preseptal cellulitis, lymphoma, lymhphedema, acne rosacea, and blepharochalasis. Diagnosis is usually confirmed with histopathology which is characteristically a granulomatous angiitis with perivascular inflammatory cells and variable dermal edema. Although, granulomas may be absent in the early stages of this disease. Our case study demonstrated the representative dermal edema and granuloma formation. Therapy for this syndrome typically consists of corticosteroids; however, the condition is frequently refractory to any treatment.

In this case report, it took 15 years of symptoms before the correct diagnosis was made; however, in that time she did receive treatment with steroid ointments, which is the mainstay of treatment, without success. Because of this, we decided to take the conservative approach and proceed with observation alone at this time. If, due to recurrences, the edema is visually significant or cosmetically unacceptable, then we will proceed with upper eyelid skin reduction during an inactive phase of the condition.

MRS should be considered amongst the differential diagnosis of recurrent eyelid swelling and it can only rule out with histopathology once this condition is suspected.

REFERENCES