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Case Report

A Melkersson Rosenthal Syndrome Case with No Histological Granuloma

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Abstract

Melkersson Rosenthal syndrome is a unique syndrome that often falls off the differential diagnosis list. Its cause is unclear and the suggested hypotheses are multiple. This is a case of a 29 year old Sudanese female with a classical triad of Melkersson Rosenthal syndrome; with upper lip swelling, fissured tongue and facial palsy. It is rare for the syndrome to appear with the classical triad with a percentage of 25% of the cases. It all started a year ago, with a painless non-pitting swelling and later she developed facial palsy. Although histopathological investigation did not reveal any granulomatous formation, the swelling regressed on prescription of prednisolone and the facial palsy gradually disappeared. The presence of granuloma or the triad is not compulsory for the diagnosis of the syndrome.

Keywords: Facial palsy, Lip swelling, Fissured tongue

Introduction

Facial swelling has been manifested on different parts of the face; intraoral and extraoral: eyelids, cheek, palate, gingiva and upper and lower lips. The disease is usually reccurrent with bouts of increase and decrease [1]. The clinical presentation can be monosymptomatic or oligosymptomatic, with or without histological findings of granuloma [2-4]. The second manifestation of the disease is facial palsy. MRS was first described in 1928 by Ernst Gustaf Melkersson, and then in 1931 Curt Rosenthal added fissured tongue as the third main feature to the syndrome complex [5-9] (Figure 1). Facial nerve involvement completes the triad. Its presentation is various; presenting on one side

Figure 1: Shows lip swelling and facial palsy.

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(either involving certain branches [1] or all branches [10]), on both sides alternatingly [11], at the same time or none at all [12].

A Case Report

A 29 years old Sudanese female complained of bouts of upper lip swelling that lasted for a year. Medical attention was sought from the department of dermatology for five months with no improvement. During the five months of follow up she had had a complete blood count and ESR and the findings were normal. Aftamed® oral gel, Alfacort® cream and Antihistamine injections were prescribed but with no effect.

On the day she presented to the clinic, the swelling of the upper lip was evident along with fissured tongue and facial palsy on the left side. The swelling was diffuse involving all of the upper lip, more on the left side. It wasn't tender to palpation, fluctuant in consistency and no changes to the overlying skin except for redness. A week before she was examined, facial palsy was noticed by the patient. Associated symptoms such as tinnitus, epiphora, change of taste and pruritus on the left corner of the upper lip and left lower eyelid - that were relieved with Alfacort® - were amongst the positive findings (Figure 2,3).

A biopsy of the upper lip was taken on the same day that revealed oral mucosa with orthohyperkeratosis, hyperplastic oral epithelium and inflamed connective tissue. No granulomas were seen in the section.

The patient was put on Prednislone 5mg, 70mg per day and tapering was managed weekly. The swelling decreased and so did the facial palsy (Figure 3).

Discussion

In spite of the fact that the syndrome presented with the complete triad, granuloma histologically was absent.

A granuloma may form or disappear within days to weeks. On the day the biopsy was taken could have been the day it disappeared, explaining its absence from the biopsy [1]. In the presence of the triad and absence of the histopathological finding of a granuloma, diagnosis of the syndrome can still be made although doubts have evolved on the appropriateness of the name "granulomatous cheilitis [2-4] (Figure 4) ".

The chronicity of the swelling, the multiple Antihistamine injections and the fact that there was no change about the patient's lifestyle (no change in her personal care products or cosmetics or address and no history of an insect bite) makes the possibility of the diagnosis to be an angioedema, weak.

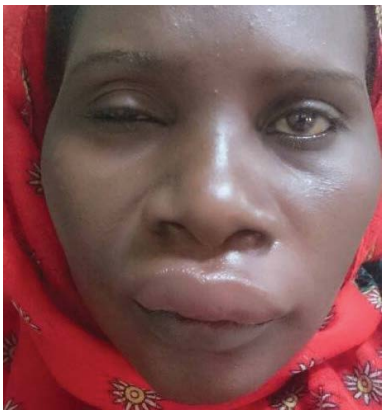

Figure 2: shows lower motor neuron facial palsy on right side and fissured tongue.

Figure 3: shows the patienton her first follow up, improvement is evident.

Figures 4: shows patient's complete recovery.

A chest x-ray has been done to exclude sarcoidosis and it wasn't suggestive.

Fortunately, the disease responds well to corticosteroids (topical, systemic or intralesional). A combination of corticosteroids and minocycline, NSAIDs and thalidomide or single drug as per this case [1,6-8]. In this case, topical steroids (Alfacort®) had no effect. Intralesional steroids were not tried. However, oral steroids showed remarkable improvement within the first week, and gradually disappeared like most of the similar cases [1,3-7,10-12] (Figure 5).

Oral steroids were prescribed over a course of 2 months and

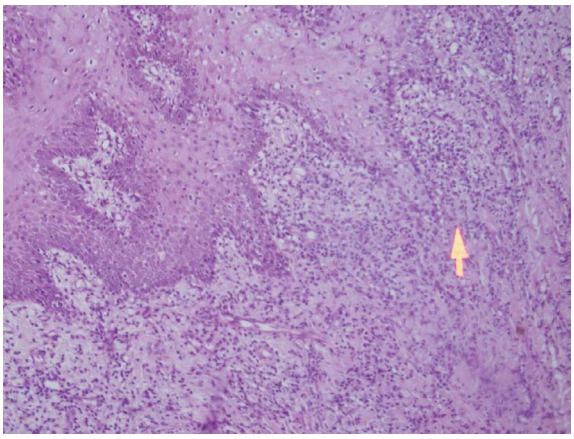

Figure 5: Chronic non-specific inflammation with epithelial hyperplasia.

the patient was followed up and tapering was done. The swelling disappeared with no new complaint. Facial palsy has a tendency to recur of 10% [2,7]. Fortunately, in this case no recurrence was detected after treatment was stopped for 36 weeks (9 months).

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