Miescher's cheilitis granulomatosa a form of Melkersson-Rosenthal syndrome
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Abstract
Melkersson-Rosenthal syndrome (MRS) is a complex neuromucocutaneous disorder characterized by localized orofacial oedema and cranial nerve dysfunction, frequently associated with minor signs, including furrowed tongue. Complete forms are rare whereas mono- and oligosymptomatic variants are more common. This paper reports a case of 30 year old female patient with Miescher's cheilitis granulomatosa as a monosymptomatic form of the Melkersson-Rosenthal syndrome which was treated by systemic steroid therapy with complete recovery.

Key Words: Melkerson Rosenthal Syndrome; Meischer Cheilitis; Granulomatous Cheilitis; Steroid therapy.

Introduction
Melkersson-Rosenthal syndrome (MRS) is a complex neuromucocutaneous disorder characterized by localized orofacial oedema and cranial nerve dysfunction, frequently associated with minor signs, including furrowed tongue. Complete forms are rare whereas mono- and oligosymptomatic variants are more common. (1, 2) Melkersson syndrome was described in 1928 as peripheral facial palsy and swelling of the lips. (3) Rosenthal, in 1930, included the presence of a fissured tongue, completing the triad which defines the syndrome. This paper reports a case of 30 year old female patient with Miescher's cheilitis granulomatosa as a monosymptomatic form of the Melkersson-Rosenthal syndrome which was treated by systemic steroid therapy with complete recovery.

Case Report
We present a case of a 30 year old female patient who reported to our dental wing with a chief complaint of swelling in her upper lip for last one year and in the lower lip from the past 15 days. The swelling was consistent in size without any aggravating or relieving factors. It was not associated with any history of trauma. The patient revealed that she had a similar swelling 2 years back which had subsided on its own and hence she did not seek any medical intervention but this time the swelling did not subside and hence reported here for treatment.

On general physical examination, the patient appeared moderately built and nourished with the other vital signs were within the normal limits. Extra-oral examination revealed diffuse swelling was present on both the lips which were soft, compressible, non-tender, non-fluctuant; on palpation (Fig 1) Intra-oral examination did not reveal any significant abnormality. On the basis of the history and clinical examination, a provisional diagnosis of oro-facial granulomatosis was made. The patient was advised to undergo labial mucosa biopsy, the histopathology report of which revealed scattered aggregates of non-caseating granulomatous inflammation, consisting of lymphocytes and epitheloid histiocytes with or without multinucleated giant cells. (Fig 2) Final diagnosis of monosymptomatic MRS was given after the anamnesis, the clinical evidence, the anatomo-pathologic study and the negative laboratory tests. The patient was administered prednisolone (10 mg) twice daily for 3 weeks with levoceptrizine for 7 days along with multivitamins and iron supplements. The corticosteroid was tapered gradually. The swelling regressed following our treatment and the patient was asymptomatic after 3 weeks of treatment. (Fig 3) Patient was re-evaluated each week for the next 6 weeks for any reoccurrence.
Discussion
Melkerson Rosenthal Syndrome is a systemic disorder that has variable cause and granulomatous nature. It involves nerves, mucous membrane and skin, especially the oro-facial region. Meischer in 1945 described chelitis granulomatosa in patients suffering from Melkersson Rosenthal syndrome. The presence of two or one of the manifestations mentioned above, with granulomatous chelitis in the biopsy, is sufficient to make the diagnosis of oligosymptomatic or monosymptomatic form of MRS. The etiology of MRS remains unknown. There is no scientific evidence that the origin of this disease could be of infective, allergic or hereditary nature.

Miescher’s chelitis granulomatosa is the most important cause of chronic lip oedema. The second sign of the triad characterizing MRS is represented by facial nerve paralysis. It may be unilateral, bilateral, partial or complete. This paralysis is due to granulomatous infiltration of the nerve and its sheath or due to its compression by tissue edema as the nerve passes through the facial canal within the temporal bone. The third sign of MRS is the presence of a fissured tongue or “lingua plicata”. It can be associated with a burning sensation and dysgeusia (it affects 2/3 of the anterior part of the tongue).

Early diagnosis is important as it provides better prognosis. The first goal of management should be identification of the initiating cause. As the etiology of MRS is unknown, treatment of granulomatous chelitis remains symptomatic. A great number of therapeutic schemes employ topical, intralesional, or systemic steroids with symptom regression in 50-80% and recurrence in 60-75% of the cases. Patients with a moderate form of granulomatous chelitis show more benefit from the administration of steroids. Surgery is reserved for patients whose chelitis does not respond to steroid therapy or who present a reasonable face deformation. In order to prevent recurrence of the disease, intralesional injections of triamcinolone 0.1% are necessary for a period of 2 to 6 months after the operation.

Conclusions
MRS should be taken into consideration in every patient with recurrent or persistent swelling of the upper lip. Therapy is only symptomatic and recurrences are frequent.

References

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