Miescher syndrome receives poor attention and may often remain misdiagnosed

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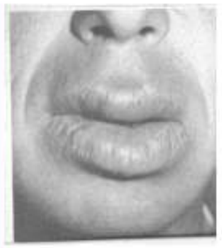
PURPOSE

Miescher syndrome, Cheiliti granulomatosa (CG) is a inflammatory disorder with unknown etiology, but a genetic predisposition may be possible . It is a rare inflammatory disorder first described by Miescher in 1945. Usually, it appears during the second decade of life, with no racial or ethnic predilection and it regresses with age. Although it may resolve spontaneously, after several recurrences it could become chronic, resulting in multiple non-infective and non-necrotic granulomas. Cheiliti granulomatosa (CG) is characterized by chronic swelling of one or both the lips due to granulomatous inflammation. It is one manifestation of orofacial granulomatosis (OFG), which is a clinical entity describing facial and oral swelling in the setting of non-caseating granulomatous inflammation and in the absence of systemic disease such as Crohn's disease and sarcoidosis. It is a monosymptomatic form or an incomplete variant of Melkersson-Rosenthal syndrome (MRS); a triad of recurrent orofacial edema, recurrent facial nerve palsy and fissuring of the tongue. Presentation of complete MRS with all three elements of the triad in a single patient is rare, being reported in only 10-20% of cases. The presence of CG without lingua plicata or facial palsies also is called Miescher syndrome or Miescher's Cheilitis. In 1985, Wiesenfield introduced the concept of Orofacial granulomatosis (OFG). Today both CG and MRS are considered subsets of OFG. Disease is often replaced by some other diseases that are accompanied by increased lips and swelling of the face. We will show seven patients who were previously diagnosed with chronic macroheilia, allergic angioedema and hereditary angioedema.

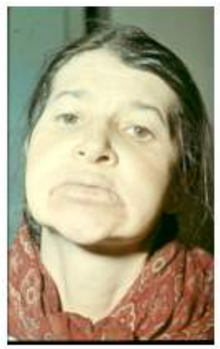
Various treatments for CG have been reported, including antibiotics like tetracycline and clofazimine tranilast, oral and intralesional steroids, and surgical resection.

Keywords: Cheilitis granulomatosa, Miescher's syndrome, Lip, Swelling of the lips and face, Cases report.



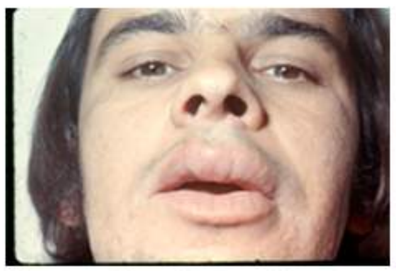
Pretreatment view of the swelling in the first patient showing diffuse, erythematous swelling of upper ,lower lip and perioral area

Pretreatment view of the swelling in the second patient showing diffuse, erythematous swelling of upper lip and face



Chronic swelling of the lips and face

Diffuse, erythematous swelling of upper lip and face



Chronic swelling of the lip and face due to granulomatous inflammation

References

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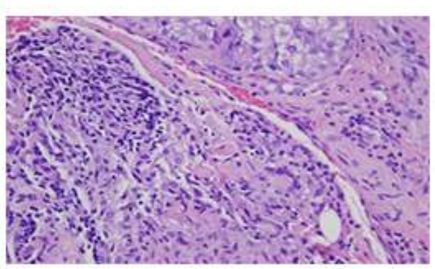

Sub-epithelial edema, increased number of dilated lymphatic vessels, inflammatory infiltrate and non necrotic and non caseating granulomas, consisting of lymphocytes and epithelioid histiocytes.

SUBJECTS AND METHODS

In this study we examined the clinical records of seven patients, one females and six male presenting, by enlargement lips and swelling of the face, with the clinical and histologic diagnosis of granulomatous inflammation of the lip, which were examined and treated in the Clinic reconstructive and plastic surgery, Sarajevo. One patient had swelling upper and lower lip (FIGURE1), three patients had swelling of the upper lip (FIGURE2,FIGURE 3, FIGURE 4), and onepatients had swelling of the lower lip and face (FIGURE 5). The youngest patient was 19 years old and the oldest 34 years old. The average age of the sick was 25.6 years. Four patients were man and one was woman . The woman was the oledest (34 years old). The average duration of the disease of all patients was 1.3 years. Three patients were treated earlier as allergic angioedema but not respond to antihistamines or steroids, one patient were treated as chronic forms of macrocheilia and one patient were treated as hereditary angioedema. Patients had no history of applied irritants, local trauma or atopy. All patients also reported burning and peeling off in perioral area. The patients referred lowered local sensation, functional limitation, but, above all, dramatic social embarrassment with a complete lack of confidence. Consistency of the lips and face was firmly elastic, thus to limit facial mimic.

We perform an appropriate evaluation, which includes a chest radiograph to exclude other etiologies of granulomatous disease, such as sarcoidosis or Mycobacterium infection. Gastrointestinal tract endoscopy and radiography used to exclude Crohn's disease. Patch tests used to exclude allergy in the pathogenesis of CG. No signs or symptoms of systemic granulomatous disease were noted. Histopathologically analysis were characterized by sub-epithelial edema, increased number of dilated lymphatic vessels, inflammatory infiltrate and non necrotic and non caseating granulomas, consisting of lymphocytes and epithelioid histiocytes (FIGURE 6). RESULTS: Patients did not agree on a surgical procedure. The treatment with the intralesional injections of of 40 mg triamcinoleone once a months, for a total of three administrations, in 3 cases . In one cases, 20 mg triamcinolone every other week,for four injections, was successful. One of the patients refused to be treated. CONCLUSION: CG seems to respond well to steroid treatment and the need for surgery is minimal and should be reserved for recalcitrant cases. Surgical intervention and radiation have been proposed in the management of CG in cases of severe disfigurement but post-surgical relapses are common. A thorough work-up to eliminate other etiologies of granulomatous disease is essential when a patient presents with granulomatous inflammation of the lip and swelling of the face.