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Idiopathic Orofacial Granulomatosis-Diagnostic Dillemas-Case Report.

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ABSTRACT

We report a case of 67 years old women who presented to our Department with indurated lower lip swelling. First histopathological diagnosis was highly suggestive of sarcoidosis and she was sent to do chest X ray and angiotenzyne serum converting enzyme (both finding being negative as was M. Tuberculosis testing). The second biopsy was consistent with diagnosis of granulomatous cheilitis. Patient was treated with intralesional injections (triamcinolone acetonide, 8 mg per visit divided into equal parts and injected in four parts of the lower lip) once a week during period of four weeks and afterwards lesion subsided.

Keywords: orofacial, granulomatosis,

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INTRODUCTION

Wiesenfeld et al. in 1985 [1] proposed term Orofacial granulomatosis (OFG) as a descriptive term for non- infectious granulomatous disorders of the lips, face and oral cavity that are histologically associated with non caseating granulomas and multinucleated giant Langhan's cells. Term OFG integrates spectrum of various disorders including Melkersson-Rosenthal syndrome and granulomatous cheilitis and has been shown to be most frequently correlated with sarcoidosis, Crohn's disease and tuberculosis. Tilakaratne et al. [2] proposed term idiopathic orofacial granulomatosis when lesions are restricted to oral region without identifiable granulomatous disease. The exact underlying mechanism of the disease is poorly understood. Previously genetic background has been postulated however, recent studies do not support this correlation [3, 4]. Regarding allergies it seems that various substances could evoke delayed type hypersensitivity reactions and it has been shown that 60% of patients affected with OFG are atopic [5]. Some authors advocated the role of dental materials in patients with OFG [6] but the results of these studies are inconclusive. In the case of only gingival enlargement, gingivectomy is a therapeutic method. Swellings in the orofacial region are treated with intralesional steroids. The swelling is initially soft but becomes firmer due to the fibrosis. Clinical oral presentation of OFG might be swelling, hypertrophy, erythema or ulcerations. However, the clinical presentation can be highly variable making the establishment of proper diagnosis difficult [2].

CASE REPORT

One year ago, a 66 year old female patient was referred to maxillofacial surgeon due to the nodular enlargment of the lip and chin (Picture 1 and 2). At the time, oral biopsy was performed and the pathologist finding was highly suggestive of sarcoidosis. Therefore, she was sent to immunologist in order to confirm systemic sarcoidosis, however, the findings excluded systemic sarcoidosis (negative serum angioconverting enzyme and chest radiograph). The patient was routinely seen by dermatologist as she suffered from pruritus which was diagnosed as neurodermatitis. She is hypertensive and has chronic gatritis, however, she didnt have any other GIT disturbances. Serum blood test were normal except elevated MCV 101.1 fL (normal range 83-97.2), decresed RDW-KV 12,4 % (normal range 14.6-16.5 %), reduced zinc levels being 8.5 L µmol/L (normal range 10.4-16.4 µmol/L), increased eosinophil granulocytes 12,8 (normal range 0-7), increased AST 32 U/L (normal range 8-30 U/L). At the time, ENA, ANF and anti ds DNA were negative but SS-A (anti Ro) was increased being 58 AU/ml (positive more than 40 AU/ml). IgG and IgG were within normal range, however IgM was decresed, i.e. 0,26 g/L (normal range 0,4-2,3 g/l). PPD test was negative. IgG and IgM antibodies to Toxoplasma gondii were negative. C1-inhibitor was within normal range, i.e. 1,29 (normal range 0,70-1,30), C3-complement component was decreased 0,68 (normal range 0,9-1,8 g/L) while C4 complement component was within normal range. However, IgE was 15 800 Iu/ml (normal range is below 100 IU/ml). Patch testing for inhalational and nutritive allergens showed positive finding for ambrosia, grass, cats, *Dermatophagoides pterony*, home dust and peanuts. Ultrasound of the abdominal organs showed kidney cyst.

One year later she was referred to the Department of Oral medicine for evaluation of nodular tissue seen as yellowish indurated infiltration on the labial aspect of the lower lip.

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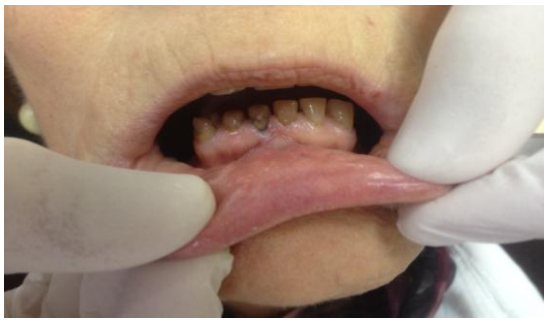
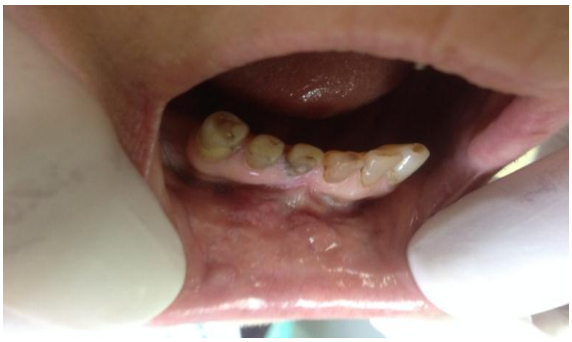
Biopsy from mucosal part of the lip was taken and diagnosis of chronic granulomatous inflammation was established by second pathologist. Histopathological finding showed partially ulcerated mucosa with several nodules consisting of epitheloid cells with some multinucleated giant cells without central necrosis. Acid resistent bacteria were not noticed. The finding is consistent with granulomatous cheilitis and differential diagnosis include sarcoidosis, Crohn's disease or biological cause. The patient was treated with intralesional corticosteroid injections (triamcinolone acetonide, Kenalog®, 8 mg per visit divided into four equal parts and applied on four locations in her lower lip once a week during four weeks and after chin lesion subsided.



Picture 1. Firm infiltration of the lower lip.



Picture 2. Granulomatous tissue involving lower lip.

DISCUSSION

The most common complaint seen in OFG patients regarding oral cavity is lip swelling or swelling in the other orofacial parts, however, it can only be manifested as gingival swelling. In our case there wasn't any gingival hyperplasia, a finding which has been reported by several authors [7, 8]. Recently, it has been reported that various hypersensitivity reactions to food, preservatives, cosmetics, oral hygiene products might predispose development of orofacial granulomatosis [5]. Our case strongly suggest that atopy might have influence on the development of OFG as our patient had increased serum IgE levels being 11000 (normal range 20-100) and was allergic to various antigens such as ambrosia, grass, cats, *Dermatophagoides pterony*, home dust and peanuts. Furthermore, in seven of these patients there was an improvement of OFG after elimination diet. However, it seems that these diets are exhausting and time consuming and results are usually minimal. We did not suggested elimination diet in our patient.

The role of dental materials in OFG has been postulated in few case reports [6, 9]. Patient with amalgam filling had swelling of the mucosa in the nearby region and was positive for mercury on patch test, following amalgam replacement, soft tissue swelling completely resolved. The other patient had soft tissue swelling adjacent to amalgam and was positive for mercury patch testing but refused amalgam replacement. However, in our patient there were no amalgam fillings near lip swelling. The role of infectious agents had been also proposed, primarily M.tuberculosis, leprae, Borrelia burgdorferi, Saccharomyces cerevisiae, systemic fungal infection. However, there is still insufficient evidence to support a definitive role for infection as causative factor in the OFG [10].

List of differential diagnosis regarding lip enlargement might include sarcoidosis, Crohn's disease, foreign body reactions, Melkersson-Rosenthal syndrome, Wegener's granulomatosis, cheilitis granulomatosa or hairy cell leukemia, hypersensitivity reactions (food substances and additives, cosmetic antigens, angioneurotic oedema, infective agents such as tuberculosis, actinomycosis, syphilis, systemic mycoses, leprosy and cat-scratch disease [2,10].

Therapy of oral lesions consists of intralesional steroid injections for tretament of OFG, clofazimine, ofazimine, low dose thalidomide, infliximab, adalimumab in relacitrant cases. Regarding intralesional steroid injections, various treatment modalities have been proposed such as once a week, twice a week and every day, however, our experience shows that intralesional injections once a week during period of three to four weeks are sufficient to eliminate symptoms in most of the patients [11,12].

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