Coexisting Orofacial Granulomatosis with Discoid Lupus Erythematosus: Report of a Rare Case

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Abstract

Orofacial granulomatosis (OFG) comprises a group of diseases characterized by noncaseating granulomatous inflammation affecting the soft tissues of the oral and maxillofacial region. The classic presentation of OFG is a nontender recurrent labial swelling that eventually becomes persistent; however, the clinical presentation can be highly variable, making the diagnosis difficult to establish. Herein, we report the rare case of a 15-year-old patient, suffering from OFG and discoid lupus erythematosus; this coexistence of two such rare entities together hardly finds a mention in the literature.

Keywords: Coexistent, discoid lupus erythematosus, orofacial granulomatosis

INTRODUCTION

Orofacial granulomatosis (OFG) is a unifying term encompassing many heterogeneous conditions which on histopathological examination reveal noncaseating granulomas.[1,2] OFG has varied manifestations with most common being recurrent painless swelling of one or both lips which becomes persistent over time. The etiopathogenesis is unknown, but various hypotheses include a role of infections, autoimmunity, genetic predispositions, and allergies.[3] Discoid lupus erythematosus (DLE) is an autoimmune inflammatory disorder of the skin characterized by well-defined red, scaly patches of variable size, which heal with atrophy, scarring, and pigmentary changes. Coexistent OFG and DLE have hardly been reported in the literature. Herein, we report one such rare case.

CASE REPORT

A 15-year-old boy reported with complaints of recurrent episodes of multiple oral ulcers, swelling and fissuring of lips, black discoloration of mucosa of oral cavity, and red raised itchy lesions over face of 8-year duration. Oral ulcers were associated with pain and intolerance to spicy food. Oral ulcers and swelling of both the lips had a relapsing and remitting chronic course with periods of complete resolution of symptoms for months. Over the next 2–3 years, he gradually developed patchy bluish-black pigmentation over mucosa of oral cavity including both lips. Five months after the onset of oral cavity symptoms, he developed an itchy red raised, coin-sized lesion over nose which was associated with photosensitivity. Patient denied a history of any oral or dental procedure, swelling of gingiva, deviation of face to one side, loose motions or pain abdomen, or any history suggestive of allergies. There was no history of joint pains or seizures. He was developmentally normal for age and height.

General and systemic examination was essentially normal. Dermatological examination revealed two well-defined erythematous plaques with central hyperpigmentation and surrounding erythema over left half of nose and left cheek [Figure 1]. There was diffuse rubbery nontender swelling of both lips along with fissuring and hemorrhagic crusting seen at places [Figure 2]. Areas of hypopigmentation and hyperpigmentation were also seen over the lips. Both lower eyelids showed areas of reddish, erythematous, slightly infiltrated plaques with scaling. Oral cavity revealed patchy hyperpigmentation with superimposed reticulate plaques over hard palate, bilateral buccal mucosa, upper...
gingival, and labial mucosa [Figure 3]. There was no lymphadenopathy. Examination of scalp, palms, soles, and nails was normal.

Relevant hematological and biochemistry profile was normal. Workup for tuberculosis, sarcoidosis, and leishmaniasis was within normal limits. Antinuclear antigen profile was negative. Skin biopsy from a plaque on nose revealed prominent interface dermatitis with marked follicular plugging [Figure 4]. Incisional biopsies from lip and oral cavity revealed acanthotic epidermis with dense lymphomononuclear infiltrate with foci of noncaseating epitheloid cell granulomas in dermis [Figure 5]. Ziehl–Nielsen stain, Gram, Grocott, Gomori and periodic acid–Schiff stains were negative. Based on the clinical and histopathologic findings, the final diagnosis of DLE with OFG was made. Patient was treated with tablet hydroxychloroquine 200 mg twice daily and tablet dapsone 100 mg daily along with intralesional triamcinolone injections with good response in the form of marked resolution of swelling of both lips. Lesions on the nose and cheeks regressed with mid potent

Figure 1: Two well-defined erythematous plaques with central hyperpigmentation and surrounding erythema over left half of nose and left cheek

Figure 2: Diffuse swelling involving both lips with areas of hemorrhagic crusting, areas of hypopigmentation as well as depigmentation

Figure 3: Oral cavity revealed patchy hyperpigmentation with superimposed reticulate plaques over hard palate, buccal mucosa, and gingiva

Figure 4: Biopsy from nose revealed interface dermatitis with marked follicular plugging (H and E, ×100)
granulomatosus in which OFG can present as an oral manifestation of systemic disease.[10] OFG includes Melkersson–Rosenthal syndrome (MRS) and cheilitis granulomatosa (CG) of Miescher. MRS generally consists of a triad of persistent lip or facial swelling, recurrent facial paralysis, and fissured tongue. CG of Miescher is characterized by recurrent or persistent swelling restricted to one or both lips.[3]

OFG has a variable clinical appearance with most common manifestation being recurrent painless swelling of one or both lips which becomes persistent over time. Other features include involvement of tongue, gingiva, buccal mucosa, oral ulcers, fissuring of the tongue, facial nerve palsy, erythema of the face, and cervical lymphadenopathy.[4] Granulomas can cause lymphedema due to lymphatic obstruction, leading to swelling of the lips. It is a rare condition with onset usually in young children and adults but can appear at any age.[6] It has a slight female predilection. The etiopathogenesis is unknown, but various hypotheses include a role of infections, genetic predispositions, and allergies. Delayed hypersensitivity to various dental materials and amalgams has been implicated. Chronic antigenic stimulus leading to monoclonal lymphocytic expansion in OFG lesions which produce cytokines responsible for granuloma formation is another hypothesis.[5,6]

The diagnosis is confirmed by histopathological findings of noncaseating granulomas. Known causes presenting with OFG including Crohn’s disease, sarcoidosis, and Wegener’s granulomatosis need to be excluded.[7] Other conditions which may show granulomatous inflammation include tuberculosis, leprosy, systemic fungal infections, and foreign body reactions which should be ruled out by appropriate investigations. Treatment of OFG is difficult, though, rarely spontaneous remission is possible. First-line therapy included local (intralesional) and systemic corticosteroids. Relapses are common, and long-term treatment is required.[8] Other therapeutic measures with variable success rates reported in the literature including hydroxychloroquine, methotrexate, clofazimine, metronidazole, minocycline alone or in combination with oral prednisone, thalidomide, dapsone, and danazol.[3,4] Cheiloplasty may be an option, especially in cases complicated by major lip deformation or inadequate response to local corticosteroid therapy.

DLE, the most common subtype of cutaneous lupus erythematosus, is a chronic dermatological disease that can lead to scarring, hair loss, and hyperpigmentation changes in skin if it is not treated early and promptly. Lupus erythematosus is a polygenic autoimmune disease linked to various HLA subtypes, immune signaling, and environmental factors, which ultimately leads to autoantibody production and T-cell dysfunction.[9,10] Till date, the exact etiology of DLE is not well understood. It has been suggested that a heat-shock protein is induced in the keratinocyte following ultraviolet light exposure or stress, and this protein may act as a target for T-cell-mediated epidermal cell cytotoxicity.[10]

Our case is really exceptional because of the association of OFG with DLE. Nazzaro et al. have reported a similar case of coexistent DLE with CG.[11] This association could help to clarify the pathogenesis of OFG as an immunological autoimmune disease, but further studies are required.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given his consent for his images and other clinical information to be reported in the journal. The patient understands that name and initial will not be published, and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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