Sarcoidosis and its oral manifestations: A case report study

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Abstract

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Abstract

A patient was referred to the oral medicine department with symptoms of redness and swelling of the lips and cheek, and intra-oral lesion. Biopsy was taken and laboratory factors were higher than normal, suggesting diagnosis of sarcoidosis. In this study we analyze the oral findings associated with sarcoidosis.

Written informed consent was obtained from the patient to publish this report in accordance with the journal's patient consent policy.

Introduction

Sarcoidosis is a multisystem granulomatous disorder of unknown etiology in which T lymphocytes, mononuclear phagocytes, and granulomas destruct the tissues affected(1). Boeck termed it sarcoidosis (Greek meaning "flesh-like condition")(2).

The diagnosis of sarcoidosis is made by the presence of supporting clinical factors with the presence of noncaseating granulomas in the biopsy sample. And elevated serum angiotensin-converting enzyme (ACE) levels could support the diagnosis(1-3). It shows an increased prevalence in females(2). The disease may present acutely or demonstrate a chronic course with periods of remission.

In two-third of the patients, oral signs were the first manifestation of the disease. And the most involved intra-oral soft tissue site is the buccal mucosa, gingiva, lips, tongue, and palate(2). Treatment of this disease may range from only observation of the patient, to taking systemic corticosteroids and steroid-sparing agents(1).

In this study, we emphasize on the oral manifestations of sarcoidosis, a multi-factorial disease that involves vital organs. Because the patient's first symptoms appeared in the mouth, it is important for dentists to be aware of these oral signs and pay close attention not to ignore it. Also, in this stage, the disease has a better prognosis.

Case Presentation

A 47-year-old woman was referred to the Oral and Maxillofacial department at Mashhad University of Medical Sciences, for redness on the skin of her left cheek, and non-tender diffuse swelling of lips (especially the upper lip) that had appeared five months previously. She declared that no improvement was obtained with prescription and use of Tetracycline ointment and Amoxicillin. Her medical records revealed a history of hypertension and hypothyroidism and usage of Captopril 250 mg (twice a day) and Levothyroxin 50 mg.

Extraoral examination showed diffuse. A swelling with slight redness in the lower lip and edematous inflammation of the upper lip with redness, particularly in the upper left area with mild swelling and erythematous on the skin of the left cheek. Asymmetry is clearly observed with lymphadenopathy in the left submandibular area with firm consistency but without tenderness in touch.

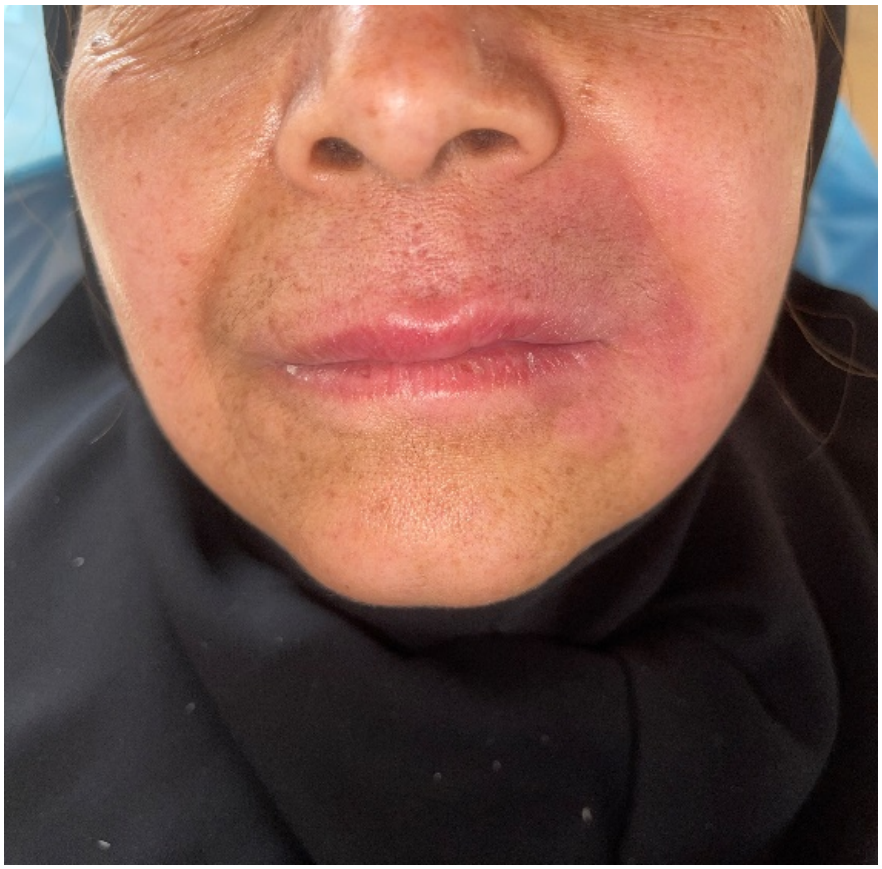

Figure 1- The extra-oral view of the patient. Diffuse swelling of the lips and redness of skin around the lips is visible.

The intraoral examination consisted of a multi-lobular exophytic lesion in the left buccal mucosa with a smooth surface, rubbery consistency, and normal color.

After intra-oral and extra-oral examination, due to the redness and swelling of the lips, OFG was the initial diagnosis. To verify this diagnosis, other local and systemic factors must be ruled out, and the first step, is a biopsy test.

The macroscopic results of the biopsy examination were as followed: soft tissue with elastic consistency and grayish color of the mucous membrane. Microscopic results showed granulomatous inflammation with bundles of histocytes that were surrounded by a lymphocytic rim, with foreign body giant cells, and asteroid body with stellate inclusions. Hence, the diagnosis turned out to be Sarcoidosis.

Later on, a serum Angiotensin-converting enzyme level test (ACE) and chest x-ray were done. The laboratory results showed a reasonably high ACE level (78 IU/L) and a slightly higher than normal ESR level (25 mm/h), which supported the diagnosis of sarcoidosis.

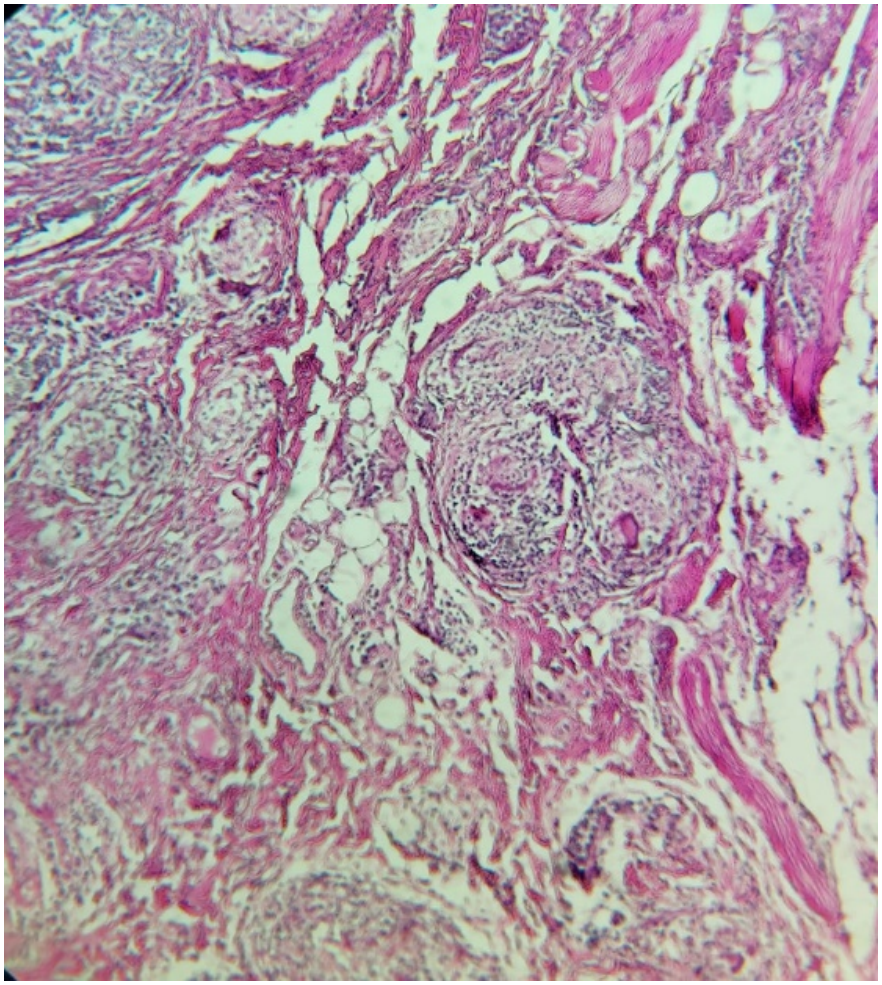

Figure 2- Microscopic view of giant cell bodies and Asteroid bodies and bundles of histiocytes.

With the consultation of a rheumatologist, Nisopred 50 mg was prescribed and after a 2-month follow-up, the patient showed a relative response to the drug, and was referred to a pulmonologist. and Due to the relative response to the previous drug, and single coughs that had started prior to taking it, Salbutamol

spray was also prescribed. A chest x-ray was performed and not showed anything in particular.

Within another follow-up, and after quitting Nisopred, the intra-oral signs and lesions recurred, and therefore with the consultation of another rheumatologist, this time Hydroxychloroquine therapy was considered. Although currently the patient hasn't checked in for a visit and did not start taking Hydroxychloroquine(4), but with follow-up on the phone, the patient reported betterment of the lip redness.

Discussion

Sarcoidosis is described as a disease in which the clinical signs and symptoms are not severe enough to cause alarm. Although the prognosis of sarcoidosis is good and in 60% of cases it regresses on its own, it is still a disease associated with heart, kidney, CNS, and lung involvement. And therefore must be taken seriously, due to the fact that almost 4-10% mortality rate is based on these involvements(5).

Some researchers claim that lesions in the soft tissue of oral cavity due to sarcoidosis are not very common(6), but our patient had a lesion on her buccal mucosa.

Swelling of the cheek and lips suggested it to be Orofacial Granulomatosis, although systemic diseases such as Tuberculosis, Sarcoidosis, Crohn's disease, and Melkersson-Rosenthal syndrome had to be ruled out first(7), therefore further microscopic testing and examination were required.

Also, in the intra-oral aspect of OFG, cobble-stone swellings, grooved tongue, recurrent labial swelling, and gingival inflammation can be seen(8). On the other hand, intra-oral signs of sarcoidosis includes the occurrence of multiple nodules, xerostomia, and involvement of salivary glands(9). The microscopic view of OFG shows aggregates of non-caseating granulomatosis inflammations(10), while in Sarcoidosis we can see aggregates of epithelial histiocytes and a surrounding rim of lymphocytes and Asteroid bodies and Schaumann bodies.

Some studies suggest a biopsy of affected tissue for demonstration of non-caseating granulomas that strongly support the diagnosis of sarcoidosis(11). And in our case, as mentioned above, the biopsy result revealed a granulomatous inflammation along with non-caseating granulomas.

Measurement of serum angiotensin-converting enzyme (ACE) can be helpful in diagnosis and monitoring the response to treatment, as the level of this enzyme is raised in about 60-80% of patients with sarcoidosis(11, 12). Although due to its poor sensitivity, an increase in ACE levels does not necessarily indicate the diagnosis of sarcoidosis(13). Laboratory tests were performed on our patient and the results were much like expected. ACE levels were higher than the normal range (78 IU/L) which could support the diagnosis.

Another factor that could support the diagnosis of this condition is the Erythrocyte Sedimentation Rate (ESR) which has a normal range of 0-22 in women, and a recent case report study claims that some patients with sarcoidosis can have an increase of ESR level(14). Laboratory tests supported the slight increase of ESR levels to 25 mm/h in our patient as well, although not significantly enough to verify the diagnosis.

Management of this particular disease can range from no interventions to systemic corticosteroids to surgical excision(15). Because it is known that corticosteroids are generally considered beneficial in the acute phase of sarcoidosis, some studies have found that oral glucocorticoids are the first option(16), much like our case in which the first-line treatment was the prescription of Nisopred 50mg.

In cases of sarcoidosis, usually, an absence of treatment response is rare and urges for verifying the absence of a diagnosis error(17). But in this case, we can see resistance to treatment response from the patient.

Nevertheless, studies have shown that oral sarcoidosis can be a manifestation of a systematic disorder(18), therefore follow-up of the patient in this stage is necessary. We hope to bring awareness to other dentists and fellow researchers to notice the oral symptoms of sarcoidosis which is usually assumed to be irrelevant to the disease.

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