Case Report

An unusual case of oral sarcoidosis: A diagnostic dilemma

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Abstract

Sarcoidosis is a rare type of multi-organ granulomatous disease. It is characterised by non-caseating necrotising granulomatous inflammation. The exact mechanism causing the disease is unknown due to its variation of manifestation in patients. The clinical presentation of this disease is protean, and often, patients remain asymptomatic throughout life. The most commonly affected organ by sarcoidosis is the lung accounting for up to 90% of all cases. Oral manifestation is a relatively rare presentation of sarcoidosis, and there are only a few cases reported till date. In this case report, the authors present a case of oral sarcoidosis of the retromolar trigone region.

Keywords: Granulomatous disease, non-caseating granulomatous disease, oral manifestation, sarcoidosis

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INTRODUCTION

Sarcoidosis is a multi-organ disease, and its occurrence in the oral cavity is relatively rare. It is a non-caseating granulomatous disease with the kaleidoscopic presentation of a cluster of macrophages, epithelioid cells, mononuclear cells and $CD4^+$ T cells with a few $CD8^+$ T cells in the surrounding zone. The disease is also denoted as the disease of exclusion, to rule out other forms of granulomatous disease, which include fungal, viral and bacterial origin.

It often remains quiescent in an individual during the life span or can be fulminating causing variable and severe clinical presentation. It was first reported by an English surgeon–dermatologist, John Hutchinson, in 1875. The nomenclature was postulated by Boeck in 1899, which in Greek was translated to 'flesh-like condition'.[1] The incidence of sarcoidosis worldwide is variable. According to the epidemiological study carried out in India in 2002, it was found that in Kolkata, 10–12 patients of 1000 new registration cases were diagnosed with pulmonary sarcoidosis; however, in Delhi the figures were 61.2 per 100000 cases.[2]

The association of sarcoidosis with environmental factors have been put forward by Ungprasert *et al.* in which they described the effect of seasonal variation, which can trigger granuloma formation in sarcoidosis. Different regions of the world show seasonal variation in the epidemiology: in Turkey, the prevalence was higher in the spring season, and in India, it was during the summer season. This depicts the effect of variability of environment on different ethnicities of people.[3]

Earlier sarcoidosis was a disease in developed countries; however, recent trends have changed and it is been observed in developing countries as well.

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Pulmonary involvement is the most commonly affected, in almost 90% of all the cases.[4] However, in a case–control etiologic study of sarcoidosis cohort, there was a higher frequency of extrapulmonary lymph node (34.2% vs. 15.2%), ocular (20.9% vs. 11.8%), hepatic (17.6% vs. 11.5%), splenic (20.9% vs. 6.7%), musculoskeletal (9.6% vs. 0.5%) and cardiac (10.7% vs. 2.3%) involvement.[5]

CASE REPORT

A 25-year-old woman reported to the Department of Dentistry, with a chief complaint of swelling on the right side of the face for 2 months. The patient was apparently well 2 months before when she started noticing swelling on the right side of the cheek with multiple ulcerations inside the mouth at different locations, for which she visited a medical physician. The intraoral ulceration subsided with time, but the swelling persisted on the right side, and she presented with the same complaint. She noted a reddish area in the inner aspect mouth right back of the tooth region. There was no associated burning sensation with the intake of spicy food, bleeding or pus discharge from the affected area. Pain on deglutition was noted, which did not subside with the intake of analgesics.

On examination, the overall build of the patient was ectomorphic and of short stature and the weight was 37 kilograms. Extraorally, there was mild facial asymmetry noted on the right side of the face on profile view. On intraoral examination, a hyperplastic lesion was noted on the right retromolar trigone region measuring approximately 4 cm × 3 cm of its greatest dimension [Figure 1]. There was a mild induration noted with respect to the ulceration. The surrounding mucosa of the ulceration was firm in consistency. The overlying mucosa of the lesion showed a pinkish-to-reddish hue with areas of brownish discoloration in the margins. Calculus and plaque were noted on the affected area denoting poor oral hygiene. There is no significant bleeding or pus discharge from the affected region.

On the right side, the level II lymph node was palpable, mobile and non-tender measuring 2 cm × 2 cm in size without fixity to the overlying and underlying structures and the temperature overlying the lymph node was not raised. She gave no positive history of tobacco intake of any form. The patient is married, her sexual behaviour history was recorded and no significant findings were found. There was a history of loss of appetite, and no history fever, cough and breathlessness was found.

A contrast-based computed tomography (CT) was performed to evaluate the extent of the lesion. The scan showed an ill-defined enhancing soft tissue thickening in the buccal mucosa and retromolar trigone extending upward to the tuberosity on the right side [Figure 2].

An incisional biopsy was performed under local anaesthesia of the lesion on the right retromolar trigone region, and the histological picture revealed stratified squamous epithelium-lined tissue with superficial squamous epithelium. The subepithelium shows a dense chronic inflammation infiltrate comprising lymphocytes, histiocytes and numerous plasma cells. The stroma showed many epithelioid granuloma cells with peripheral lymphocyte rimming. At places, these granulomas were coalescing with each other. Many Langhans giant cells and foreign body giant cells were also noted within the stroma [Figure 3]. The staining of acid-fast bacilli and fungal organisms was negative, suggestive of oral sarcoidosis.

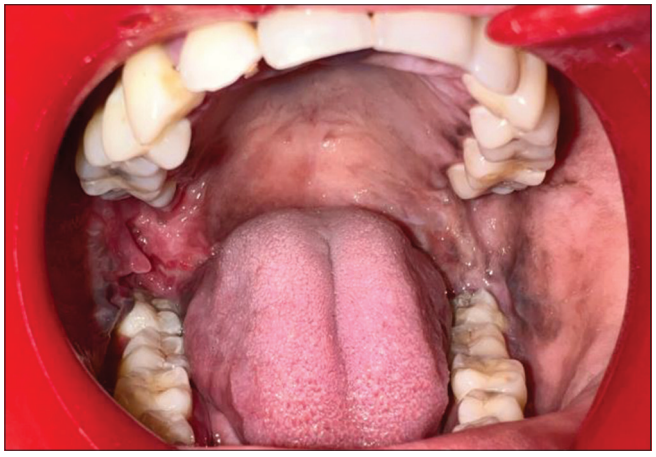

Figure 1: Oral sarcoidosis flesh-like hyperplastic lesion on the right retromolar trigone

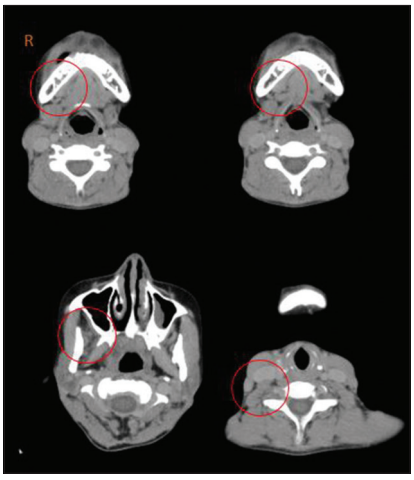

Figure 2: Contrast-based CT image showing the enhancement of the soft tissue

On further evaluation of the case, a fine needle aspiration was performed from the level II lymph node on the right side of the neck. The smear showed occasional epithelial granuloma cells in a reactive lymphoid tissue background with negative Ziehl–Neelsen staining.

This was correlated with serum angiotensin-converting enzyme (ACE) level, which was 51 U/L (normal value: 18–55 U/L), and serum calcium level was 9.2 mEq/L (normal value: 9–11 mEq/L). Haemoglobin was 8.5 mg/dl, and the erythrocyte sedimentation rate was 80 mm/hr.

A pulmonary medicine referral was carried out to rule out the possible lung involvement. A posterior–anterior view of the chest radiograph showed no significant findings, and a high-resolution computed tomography (HRCT) of the chest and abdomen was performed to rule out inconspicuous involvement. Images showed multiple chains of black pearl signs in the lung field suggestive of lung parenchymal sarcoidosis [Figure 4]. A confirmatory diagnosis of sarcoidosis was established based on all diagnostic findings. A baseline pulmonary function test was performed, which was within normal limits. Since there were no significant systemic clinical symptoms, she was kept under close follow-up.

For the oral lesion, she was given an intralesional steroid (triamcinolone) for 6 weeks and kept under close follow-up. Tooth extraction of 48 was performed due to its close approximation of the lesion, which was causing difficulty in maintaining hygiene.

The patient was followed up for 3 months, and the overall health of the patient improved. She was more energetic than before; she regained her appetite and weighed 45 kg [Figure 5]. After three months, a pulmonary function test was repeated and there was a marked improvement in forced expiratory volume 1 (FEV1) from the baseline [Figure 6].

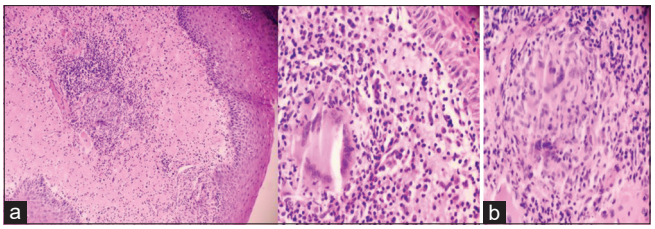

Figure 3: (a) Section examined showing epithelium lined by superficial squamous epithelium and the subepithelium showing chronic inflammatory infiltrate (H&E, 100X). (b) Stroma showing many epithelioid cell granulomas with peripheral lymphocytic rimming (H&E, 400X)

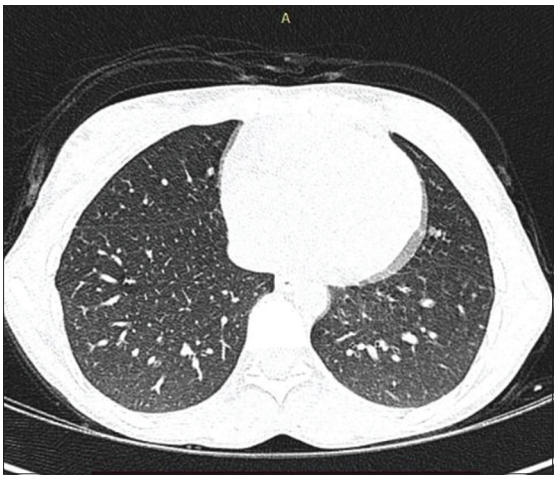

Figure 4: HRCT showing typical peribronchovascular and perilymphatic nodules

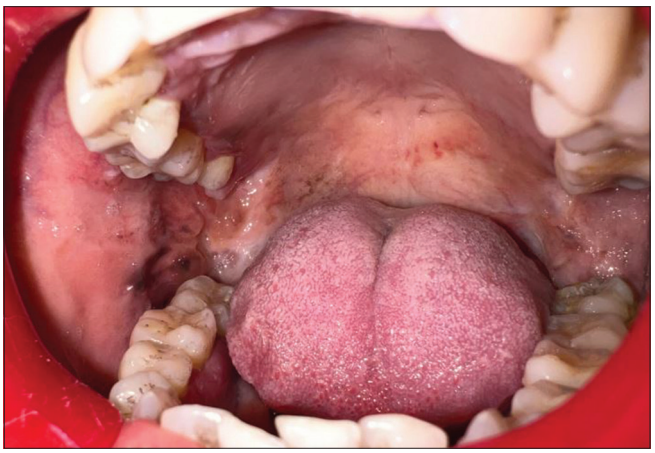

Figure 5: Follow-up after 3 months

DISCUSSION

The presence of non-caseating granulation alone is not the pathognomonic feature of oral sarcoidosis. Along with the histological findings, there should be an appropriate clinical manifestation of the diseases and exclusion of all other granulomatous inflammations. However, the only exception is pulmonary involvement. The site of biopsy is determined by the affected organ and accessibility, and the presence of non-caseating granulomatous tissue in at least one organ is considered sufficient for the diagnosis. In case the biopsy is inconclusive from any other organ, a lung parenchyma or lymph node is taken for histopathological evaluation.

The histological picture is that of granulomatous inflammation without caseating necrosis. The lesion might have many inclusion bodies such as Schaumann bodies, asteroid bodies, Hamazaki–Wesenberg bodies and calcium oxalate crystals. It is important to exclude various

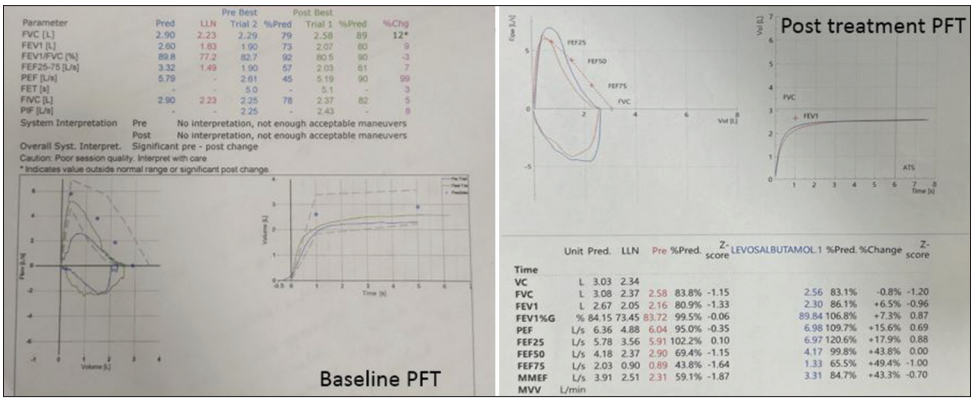

Figure 6: Illustration of pulmonary function test

other possibilities of granulomatous lesions other than sarcoidosis. There are no said markers for sarcoidosis, and serum ACE is a good indicator, which is elevated at the beginning of the treatment and monitored during therapy. The ACE levels have 10% false-positive rate and 40% false-negative rate, as the levels are raised in diabetic mellitus, cirrhosis and leprosy.[6]

The prevalence of sarcoidosis amongst the female population is found to be higher than in the male population.[7]

Due to the variation of the clinical presentation of sarcoidosis, the exact mechanism of the disease is not yet discovered. However, there are various risk factors such as genetics and environmental and infectious organisms, which can be the cause. The genetic factor is postulated due to the strong association of human leukocyte antigen (HLA) HLA-A1, HLA-B8 and HLA-DR3, and there is also a strong association with the allelic variation of HLA-DRB1 as a major contributor to disease progression.[8]

Multiple possible infectious organisms can be postulated that can cause sarcoidosis. Due to the immunological features of the disease, it is often seen that Mycobacterium plays a major role in its pathogenesis.[9]

During the summer season, exposure to sunlight helps in the conversion of 7-dehydrocholesterol to vitamin D3, which explains the hyperglycaemic state, and elevated levels of vitamin D3, which helps in the detection of such cases.[10] However, during the winter season, the pulmonary symptoms of the patients rise, resulting in the detection of pulmonary sarcoidosis.

The treatment of oral sarcoidosis is varied based on the location of the lesion and its symptoms associated with the disease. According to a review article by Suresh *et al*., oral sarcoidosis should be initially diagnosed by biopsy followed by a physician's referral to rule out any systematic involvement. Pulmonary sarcoidosis, if asymptomatic, does not require treatment, only if the pulmonary function test worsens over time treatment is indicated.[1] The oral lesion is cured by local treatments such as surgical excision, curettage and intralesional or topical application of steroids. Some lesions show spontaneous remission without any intervention.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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