

Sarcoidosis: oral and perioral manifestations

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Abstract

A 46-year-old white woman with lesions on the lower lip, perioral area and in the soft tissues of the oral cavity (gingivae and palate) was examined. The clinical signs were recorded, and incisional biopsies from the oral lesions were taken. The diagnosis of sarcoidosis was established by the histopathological evidence of typical non-caseating granulomas from tissue biopsy, supported by serum ACE- 57.9 U/L, blood calcium 16.83 mEq/L and 24-hrs urine calcium 600 mg). Oral lesions may be the first or the only sign of sarcoidosis in an otherwise healthy patient. Hippokratia 2009; 13 (2): 119-121

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Sarcoidosis is a relatively rare, acquired systemic granulomatous disease, affecting multiple organs and tissues. The respiratory system is most commonly affected, with approximately 90% of patients presenting pulmonary findings during the course of their disease. Cutaneous manifestations occur in around 25% of cases, but are more common in chronic forms¹⁻³. Head and neck lesions of sarcoidosis are manifested in 10 to 15% of patients.

In the maxillofacial region the salivary glands are frequently involved, while sometimes, xerostomia and bilateral parotid swelling are present^{4,5}. Lesions that occur in the soft tissues of the oral cavity and/or in the jaws are rare.

The purpose of this study is to present an interest case of sarcoidosis in the oral cavity and perioral region.



Figure 1: Clinical view of the skin lesions of the lower lip and chin.

Case report

A 46-year-old white woman from Russia was referred in 2007 to the Department of Oral Medicine and Maxillofacial Pathology, of our University, because of lesions on the skin of the lower lip and chin area, as well as in the soft tissues of the oral cavity. According to the patient, the lesions were present for the last 6 years and progressively enlarged. The patient's medical history was negative for previous illnesses.

Her physical examination was significant for the presence of well-circumscribed violaceous plaques of the skin of lower lip and the chin, and a single painless subcutaneous nodule, which was located under the vermilion of the lip (Figure 1). Intraoral examination revealed the presence of a well-defined asymptomatic red-violet nodular mass in the middle of the palate, measured 2x2.5cm in diameter (Figure 2). Also, diffuse erythematous and hyperplastic gingival in the upper incisor area was observed (Figure 3). The regional lymph nodes were not palpable, and salivary glands were normal.

Incisional biopsies were taken from the lesions of the hard palate, and from the skin of the lip. Histopathologic examination of the specimens showed numerous non-caseating epithelioid granulomas that consisted of histiocytes and Langhans' or foreign body multinucleated giant cells. In some of the giant cells Schaumann bodies were observed (Figure 4, 5).

Since the histopathological findings were compatible with sarcoidosis, in order to confirm the diagnosis, we proceeded with laboratory tests that strongly support the diagnosis for sarcoidosis. The results were: serum angiotensin converting enzyme (ACE) 57.9 U/L (normal value: 18-55 U/L), blood calcium 16.83 mEq/L (normal value:



Figure 2: Intraoral asymptomatic red-violet mass in the middle of palate, with well-defined borders and nodular surface.

9-11 mEq/L) and 24-hrs urine calcium 600 mg (normal value <180 mg). Erythrocyte sedimentation rate was 21 mm/h. Acid-fast bacilli were not found in smears from specimens of the palate and culture for isolation of *Mycobacteria* sp. in Löwenstein-Jensen medium was negative. The radiograph of the maxillofacial region did not show any abnormality. Based on the above laboratory findings the diagnosis of sarcoidosis was established. The patient was referred to the University Clinic of Pulmonary Diseases for further investigation, which however, was negative for systemic organ involvement.

Discussion

Oral involvement generally appears in patients with chronic multisystem sarcoidosis and seldom occurs in the acute stage. The oral lesions may be solitary, multiple or part of a generalized disease. In some cases, oral involvement is the first, or only, manifestation of the disease^{1,3,5}, and appears as a nontender well-circumscribed brownish red or violaceous swelling, as papules, or as submucosal nodules that can occasionally either show superficial ulceration or be symptomatic. Gingival involvement presents as red gingival enlargement^{6,7}.



Figure 3: Clinically, the gingival and predominantly the interdental papillae, of the patient appeared swollen and eruptive.

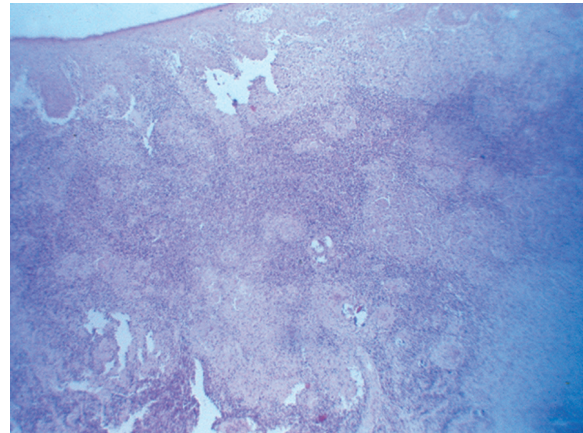


Figure 4: Histological appearance of the patient's hard palate showed typical sarcoid granuloma, and the absence of central necrosis (H-E. Original magnification X10).

In the present case the lesions were multifocal, including the lips, the gingivae and the hard palate. The clinical signs (red-violet nodular mass in the middle of the palate, and the erythematous and hyperplastic gingival in the upper incisor area) were in accordance with other researchers' results¹⁻³. Moreover, oral sarcoidosis may be asymptomatic or mildly symptomatic with minimal discomfort during eating or drinking, especially if the lesions involve the tongue⁸.

Although the etiology of sarcoidosis is unknown, many factors may be accused of the pathogenesis of this disease. In these causative factors are involved: infections (fungal, viral, bacterial), genetic predisposition, environmental factors and miscellaneous^{9,10}. The specific tests for fungal (mycology tests for *Candida* spp-cultivation in SDA and CHROMAgar *Candida*), viral (Abs to HIV, EBV, CMV) and bacterial (for mycobacterium-skin test and AFB) infections were negatives.

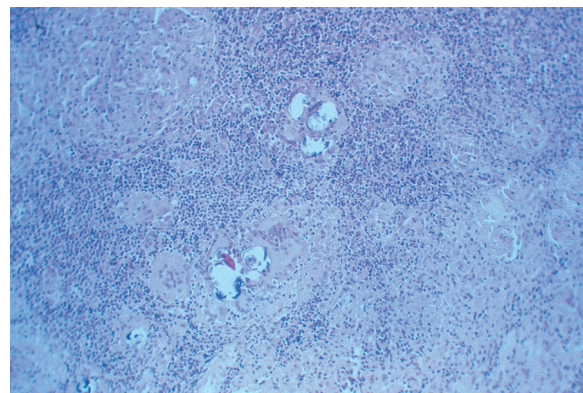


Figure 5: Scattered non-caseating epithelioid cell granulomas consisted of chronic inflammatory cells-histiocytes and Langhans' type multinucleated giant cells, some of which contained Schaumann bodies (H-E. Original magnification X100).

In most cases of oral involvement, sarcoidosis is diagnosed before the oral manifestations become apparent. Occasionally, as in the present case, oral involvement is the first or only manifestation of the disease¹⁻⁴. The diagnosis of sarcoidosis is established when clinical features are supported by histopathological evidence of typical non-caseating epithelioid granulomas and other laboratory tests. The differential diagnosis of oral soft tissue lesions must consider other granulomatous conditions, such as infections (tuberculosis, leprosy, tertiary syphilis, systemic mycoses, and cat-scratch disease), Crohn's disease, Melkersson-Rosenthal syndrome (including Miescher's cheilitis or cheilitis granulomatosa), Wegener's granulomatosis, foreign body reactions and hairy cell leukemia¹¹.

Oral glucocorticoids are the first-line treatment. Other medications include cytotoxic drugs such as methotrexate, azathioprine, chlorambucil, cyclosporine and cyclophosphamide^{1,4,12}.

Some authors suggest the surgical excision for treatment of oral soft tissue or jaws lesions^{1,5}.

Conclusively, oral lesions may be the first or the only sign of sarcoidosis in an otherwise healthy patient. Oral involvement of the disease is very rare. Prognosis of sarcoidosis correlates with mode of onset, initial clinical course, hosts' characteristics, and extent of disease. However, this multisystem disorder may never be completely

cured. Moreover, it is important to enforce a periodic follow-up of patients in order to evaluate the disease

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