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Pathology in Focus

Mid-line swelling of the palate

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Abstract
Sarcoidosis is a multi-system, non-caseating granulomatous disease of unknown aetiology that may affect any organ. The oral involvement of sarcoidosis is rare and usually an initial manifestation of the disease. In this case report the authors present a 25-year-old African-American woman with palatal sarcoidosis treated successfully with intra-lesional corticosteroid injections.

The oral manifestations of sarcoidosis are relatively uncommon and may be the only manifestation of the disease. Suspected cases of oral sarcoidosis should be biopsied and subsequently referred to a physician to rule out systemic involvement.

Key words: Sarcoidosis; Palate; Adrenal Cortex; Hormones; Pathology, Oral

Introduction

Sarcoidosis is a multi-system, non-caseating granulomatous disease of unknown aetiology that may affect any organ, including the oral cavity.[1] Common clinical presentations of sarcoidosis include hilar lymphadenopathy, pulmonary infiltration, dermal, and ocular lesions.[2] The oral involvement of sarcoidosis is rare.[3] In this case report a patient is presented with sarcoidosis restricted to the hard and soft palate.

Case report

A 25-year-old African-American female presented with chief complaints of 'painless, enlarging swellings, and redness of roof of the mouth' of three months duration. The clinical history revealed that she had consulted her dentist who had treated her palatal lesions with systemic antibiotics, saline and chlorhexidine rinses without response. Past medical, family, and dental history were non-contributory.

Physical examination revealed a well-developed, afebrile patient with normal vital signs and no regional lymphadenopathy. Intra-oral examination revealed three discrete erythematous/purplish exophytic masses on the hard palate, soft palate and uvula, the largest measuring approximately 4.0 × 3.0 cm across (Figure 1). On palpation, the masses were soft and non-tender. Posterio-anterior chest radiographs revealed no abnormal findings. Computerized tomography (CT) scans of the head and neck, chest and abdomen were performed and revealed destruction of the premaxilla, hard palate, inferior portion of the vomer and the perpendicular plate of the ethmoid. CT scans of the chest, and abdomen showed a clear mediastinum and no lymph node involvement. Incisional biopsy of the lesions was not diagnostic and showed only necrotic and inflammatory tissue. VDRL, ELISA and repeated blood cultures for acid fast bacilli and fungi were negative. All other blood laboratory studies were within normal limits. Additional biopsies of the hard and soft palate were carried out under general anaesthesia and revealed non-caseating granulomas with epithelioid cells and occasional multinucleated giant cells associated with extensive inflammation, bone destruction but no evidence of necrosis and/or vasculitis (Figures 2 and 3). Acid fast, Periodic acid Schiff (PAS) and Gomori's methenamine staining were all negative. A diagnosis of sarcoidosis was made. Subsequently, a whole body bone gallium scan was

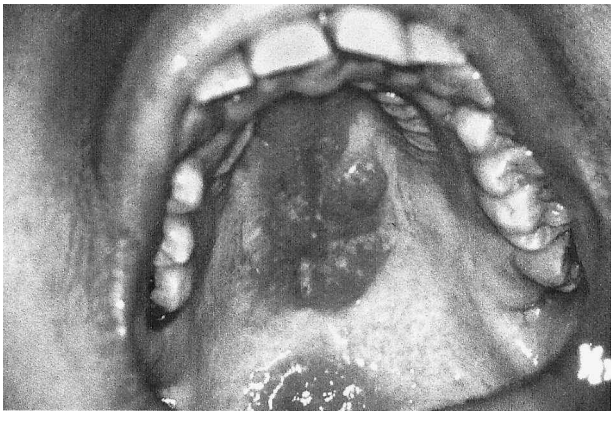

Fig. 1
Erythematous, exophytic masses on the hard palate, soft palate and uvula.

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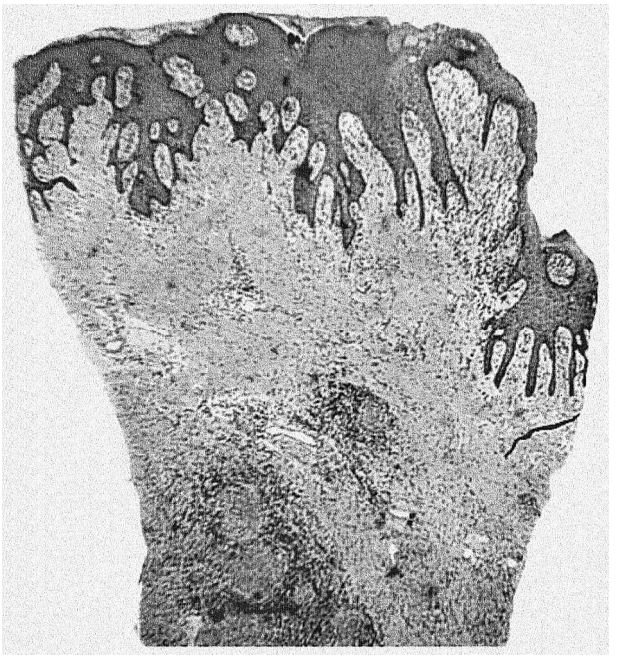

FIG. 2

Photomicrograph of incisional biopsy of the hard palate demonstrating granulomas in the connective tissue (H&E; ×100).

performed and showed increased activity in the nasal and palatal bones, both eyes, parotid gland and left hilum consistent with sarcoidosis. Pulmonary function and diffusing capacity tests, nasopharyngoscopy, and slit-microscope eye examination were normal. Additional diagnostic serum levels of angiotensin-converting enzyme, and calcium levels were within normal limits. The patient received a series of eight treatments of intra-lesional local corticosteroid injections (2 ml of hydrocortisone, 5 mg/ml) under local anaesthesia upon which there was a complete resolution of the palatal lesions. The patient failed to attend her subsequent follow-up appointments.

- **The oral manifestations of sarcoid are uncommon**
- **This case report describes a patient with midline palatal swelling due to sarcoid that was successfully treated with injections of corticosteroids**
- **The paper reminds us that an oral manifestation may be the only feature of sarcoid**

Discussion

Sarcoidosis is a multiorgan granulomatous disorder of unknown aetiology. Current epidemiological and laboratory studies suggest an immunological pathogenesis, where T-helper 1 lymphocytes (Th1) play a central role.[4] The formation of granulomas is thought to be the result of a local amplification modulated by Th1 in response to the tissue deposition of poorly soluble antigenic material.[4]

The clinical presentation of sarcoidosis depends on several factors, including; ethnicity, chronicity of illness, site and extent of involvement of the organ, as well as activity of the granulomas. One third of the patients with sarcoidosis can present with nonspecific constitutional symptoms such as fever, fatigue, malaise or weight loss.[5]

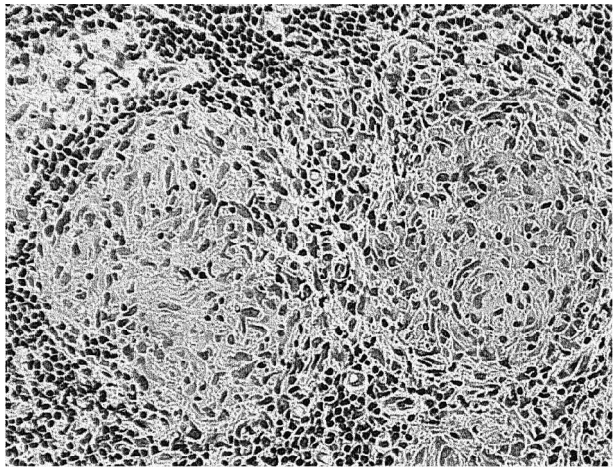

FIG. 3

Photomicrograph showing non-caseating granulomas with epitheloid cells exhibiting a rim of lymphocytes (H&E; ×400).

The most common presentation of sarcoidosis consists of pulmonary infiltration with hilar lymphadenopathy (90 per cent of the cases), followed by ocular (50 per cent) and dermal lesions (25 per cent). Sarcoidosis affecting the palate is rare.[6,7] The clinical differential diagnosis of midline palatal swellings causing bone destruction includes; salivary gland neoplasms, Kaposi sarcoma, angiocentric T-cell lymphoma (mid-line lethal granuloma), Wegener's granulomatosis, sarcoidosis and various infectious diseases of bacterial (syphilis, tuberculosis, leprosy) and/or fungal aetiology (zygomycosis, blastomycosis). Microscopic examination and culture for bacteria and fungi help to differentiate these lesions.

Sarcoidosis is a diagnosis of exclusion. No diagnostic tests or specific markers have been established yet.[2,9] The diagnosis is based upon positive history (occupational or environmental exposure to dust, pollen, clay) decreased pulmonary function tests (forced expiratory volume, vital capacity),[10] elevated serum calcium and serum angiotensin-converting enzyme levels,[11,12] radiographs,[10,13] and histological studies. The histology of sarcoidosis will show non-caseating granulomas with the presence of epithelioid macrophages, lymphocytes and occasional multinucleated giant cells.[14] The giant cells result from the fusion of the epithelioid macrophages and may occasionally contain many basophilic inclusion bodies such as Schumann bodies and/or asteroid bodies.[14–16]

Not all patients with sarcoidosis require treatment but if treatment is required, corticosteroids are the therapeutic mainstay. Immunosuppressant drugs such as methotrexate, cyclophosphamide and azathioprine also have been shown to be beneficial in the treatment of sarcoidosis. These drugs should be used with caution due to their adverse side effects. Drugs inhibiting TNF-alpha such as etanercept, infliximab, pentoxifylline, and thalidomide have also shown promise in the treatment of sarcoidosis, but more controlled clinical trials are necessary to show their effectiveness.[17]

Conclusion

The oral manifestations of sarcoidosis are relatively uncommon and may be the initial and/or the only manifestation of the disease. Suspected cases of oral sarcoidosis should be biopsied and subsequently referred to a physician to rule out systemic involvement.

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