Parotid Sarcoidosis

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Citation

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Abstract

Sarcoidosis is a multisystem granulomatous disorder of unknown etiology, commonly affecting lungs. The salivary glands are rarely involved. A 40-year female was admitted with bilateral symmetrical non-tender, firm and smooth parotid swelling. Histopathological study showed an extensive non-caseating epitheloid cell granulomas with multinucleated giant cell, langerhan's foreign body cell type destroying the normal parenchyma of the parotid gland, suggestive of sarcoidosis. The patient responded remarkably well to a regime of corticosteroids.

INTRODUCTION

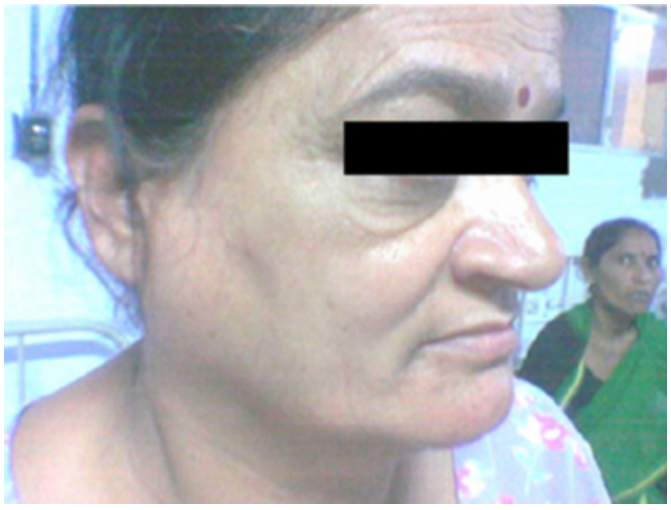
Jonathan Hutchinson first reported sarcoidosis, a multisystem granulomatous disorder of unknown etiology in 1869. [1] Several causative agents have been implicated in the pathogenesis of this disease, which includes infectious agents such as mycobacterium as well as non-infectious agents such as exposure to beryllium dust.[2,3]

CASE REPORT

A 40-year-old female was admitted with history of bilateral symmetrical, non-tender, firm and smooth parotid enlargement along with xerostomia since three weeks. (FIG-1)

Figure 1

Figure 1: photo showing bilateral parotid gland enlargement in the patient with parotid sarcoidosis.



She also noticed maculo-papular eruptions over the right shoulder since one week. The patient reported no history of fever and there was no evidence of peripheral lymphadenopathy or hepatosplenomegaly. Cardiovascular, respiratory and central nervous system examination revealed no abnormality.

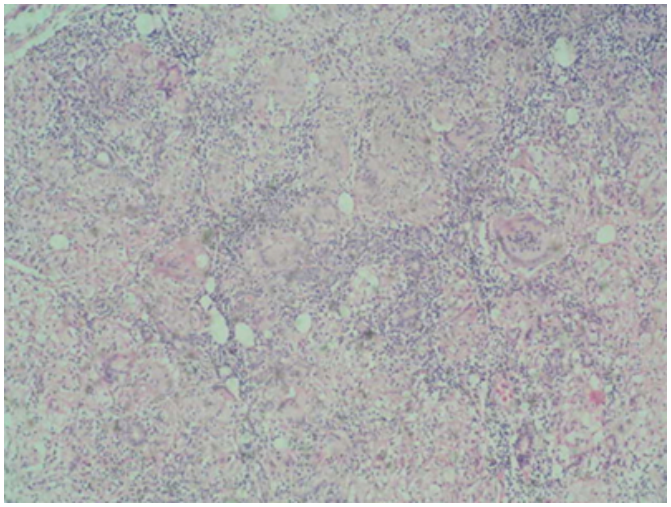
On diagnostic work-up her hemoglobin was 10.2 gm/dl. Total leukocyte count was 7660 per cu. mm. and differential count showed neutrophils 67%, lymphocytes 32% and eosinophils 1%. Platelet count was 1.5 lakhs per cu. mm. chest radiograph was normal. Erythrocyte sedimentation rate and Mantoux test were within normal limits.

Fine needle aspiration cytology from both the enlarged parotid gland showed few epitheloid cell granulomas amidst tiny clusters of epithelial cells, which at few places were forming acinar structures and a few occasional multinucleated giant cells without any evidence of necrosis, suggesting a chronic granulomatous inflammation.

Incisional biopsy was planned which revealed an extensive non-caseating epitheloid cell granulomas with multinucleated giant cell, langerhan's foreign body cell type destroying the normal parenchyma of the parotid gland. Intervening tissue showed dense chronic lymphocytic infiltration. (FIG-2)

Figure 2

Figure 2: histopathology of the parotid gland showing non-caseating granuloma.

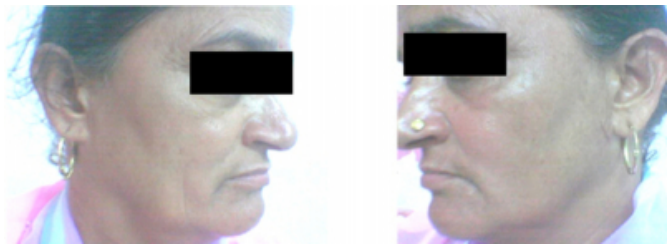


By integrating the clinical data with presence of non-caseating granulomas, a diagnosis of Sarcoidosis of parotid was made.

The patient was initiated a regimen of corticosteroids in the form of oral prednisolone 1mg/kg body wt./day for 4 weeks, followed by slow tapering over 6 weeks. She responded remarkably well to the above regime. (FIG-3)

Figure 3

Figure 3: photo showing absence of parotid swellings following treatment.



DISCUSSION

Sarcoidosis most commonly affects lungs, followed by lymph nodes and spleen. The salivary glands are rarely involved and usually manifests in a variety of clinical patterns. [4]

They commonly presents as major salivary gland enlargement with only histopathological involvement of the minor salivary glands. The second clinical pattern is characterized by the absence of clinical salivary gland swelling with only histopathological involvement of minor salivary glands. Heerfordt's syndrome or uveoparotid fever is the third pattern, which consists of a triad of symptoms including parotid swelling, uveitis and facial palsy.

Parotid gland involvement occurs in 0.5-15 % of patients of sarcoidosis and may manifest as unilateral or bilateral painless swelling. [5] Xerostomia may be associated depending upon the extent of granulomatous infiltration of the gland.[4]

There is no specific laboratory test for Sarcoidosis. Markers of activity include raised levels of serum angiotensin converting enzyme (ACE), abnormal calcium metabolism, a positive Kveim-Siltazbach skin test and radioactive gallium scanning (Gallium-67 citrate) of parotid gland. [6,11]

Diagnosis is confirmed by the presence of non-caseating epitheloid cell granulomas on histopathology of the affected parotid gland. [7] Since non-caseating granulomas are occasionally seen in tuberculosis, special stains such as Ziehl-Neelsen may be used to rule out the above condition. [4]

The main goal of the treatment is to minimize or prevent inflammation and granulomas formation that ultimately causes end stage organ destruction by the development of hyaline fibrosis.

Corticosteroids remain the mainstay of therapy. Methotrexate, Azathioprine, Hydroxychloroquine, Cyclophosphamide and recently Infliximab has been tried with varying success. [9,10] However spontaneous regression of this disease have also been reported. [8]

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