Isolated Sarcoidosis of the Tonsil: Case Report and Literature Review

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Objectives

1. To present only the eighth published case of sarcoidosis presenting as unilateral tonsil hypertrophy; and even more rare, to present sarcoidosis limited only to the tonsil in the absence of a history of acute or chronic tonsillitis.

2. To review the existing literature as it relates to evaluation, management, and follow-up of this finding including the association with malignancy.

Abstract

Study Design: Case report and literature review.

Methods: A case is presented with appropriate history, laboratory values, imaging results, and pathology slides. Then the literature on the topic of sarcoidosis of the tonsil is systematically reviewed.

Results: A 25 year old black woman with no history of acute or chronic tonsillitis presented with unilateral tonsil hypertrophy and had biopsy showing non-necrotizing granulomas consistent with sarcoidosis. She had elevated angiotensin converting enzyme (ACE), but no other systemic signs or symptoms at diagnosis or upon follow-up evaluation.

Conclusions: This case and literature review support the finding of sarcoidosis limited to and presenting as asymmetric tonsillar hypertrophy. However, it is still important that such patients receive appropriate work-up, referral, and follow-up for development of systemic manifestations of sarcoid and to be vigilant for any signs or symptoms of malignancy.

Introduction

Sarcoidosis is a multi-systemic disease of unknown etiology characterized by the presence of noncaseating granulomas composed mainly of epithelioid cells with the presence of Langhan's giant cells and histiocytes.1,2 Manifestations of sarcoidosis in the head and neck are present in 10 to 15% of patients with sarcoidosis, and may include cervical adenopathy, enlarged lacrimal glands, uvula, orbit, mass, nasopharyngeal lesions, or bilateral parotid enlargement.3,4 Tonsillar manifestations are rare, comprising less than 0.03% of cases.5

Case Report

A 25 year old African American woman presented to her internist with one month of noticing her uvula was being pushed to one side beginning after a self-limited common cold. This progressed to pain and mild odynophagia without fevers or chills. She also noted that her right tonsil was enlarged as compared to the left. She was sent for an MRI and referred to an otolaryngologist by her primary medical doctor for further evaluation.

She had no significant medical history, including no history of tonsillar infections. She occasionally smoked and reported drinking a small amount of alcohol socially. She took no medications and has no family history of malignancy.

Preoperative laboratory values showed normal CBC, basic metabolic panel, and LFT except for borderline low platelets (148) and elevated AST (50 U/L, normal 15 to 41 U/L) and ALT (83 U/L, normal 14 to 54 U/L). Imaging is shown below.

The patient was taken to operating room for tonsil biopsy which showed non-necrotizing granulomas within a lymphoid infiltrate just deep to squamous epithelial surface lining of tonsil which was consistent with sarcoidosis.

Follow-up evaluation showed an elevated angiotensin converting enzyme (ACE) level of 80 Units/Liter (normal 9 to 67 U/L). Chest X-ray was within normal limits without hilar lymphadenopathy. The remainder of malignancy workup and review of systems was negative. The patient was placed on steroids. One year follow-up revealed no further evidence of sarcoidosis manifestations.

Discussion

We report a new case of unilateral tonsillar enlargement and elevated ACE level as the only manifestations of sarcoidosis. Only seven authors have previously published cases of sarcoidosis presenting as tonsillar asymmetry.6-11

Granulomatous tonsillar enlargement can be the result of numerous etiologies, such as Crohn's disease, fungal infection, tuberculosis, lymphoma, squamous cell carcinoma, venereal disease, toxoplasmosis, berylliosis, foreign body reactions, and secondary responses to lymphoid tissue draining carcinoma.2,12,13 Sarcoid-like granulomas can also be a manifestation of neoplastic disease such as Hodgkin's and non Hodgkin's lymphoma.14,15,16 These causes must be excluded prior to making a diagnosis of sarcoidosis.

Some believe that development of sarcoidosis is a cellular immune response to excessive antigen stimulation resulting from an exaggerated immune response to the stimuli of chronic tonsillitis.11,13 However, the patient in this case report, had an elevated ACE level and no history of chronic tonsillitis, thereby supporting the finding of sarcoidosis as a mild self-limited finding isolated to the tonsil. It will, however, be important to follow this patient for any subsequent systemic manifestations of systemic sarcoidosis.

A number of studies have shown a relationship between sarcoidosis and malignancy. Some authors have suggested the existence of a sarcoidosis-lymphoma syndrome in which the chronic active type of sarcoidosis appears to be responsible for an increased risk of malignant transformation of lymphoid cells.14,17,18 As described by Brickner, there are three main features of sarcoidosis-lymphoma syndrome: 1) lymphoid malignancy occurs after a preceding history of sarcoid; 2) the median age of onset of sarcoidosis is 10 years above that of unselected patients with sarcoid; 3) Hodgkin's disease occurs more frequently than other types of lymphoma. Following patients for any signs or symptoms of malignancy will be important.

Conclusion

This case and literature review supports the finding of sarcoidosis limited to and presenting as asymmetric tonsillar hypertrophy. However, it is still important that the patient receive appropriate work-up, referral, and follow-up for development of systemic manifestations of sarcoidosis and to be vigilant for any signs or symptoms of malignancy.

References