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UPPER RESPIRATORY TRACT INVOLVEMENT IN SARCOIDOSIS: TWO CASES REPORT

Medicine

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ABSTRACT

Sarcoidosis is a multisystemic granulomatosis of unclear etiology and pathogenesis. It can affect any organ but rarely it involves the upper respiratory tract. We report two cases of isolated extrapulmonary sarcoidosis observed in our Department.

KEYWORDS

Sarcoidosis, Granulomatous Disease, Tonsillar Sarcoidosis, Nasopharyngeal Sarcoidosis.

INTRODUCTION

Sarcoidosis is a multisystemic granulomatosis of unclear etiology and pathogenesis.[1] It can affect any organ most commonly lungs and limph nodes followed by skin, eyes, liver, heart, kidney, bone, salivary glands and nervous structures.[1-2] The involvement of upper respiratory tract is rare, approximately 6% of patients with sarcoidosis.[2] Isolated Waldeyer's ring involvement in sarcoidosis is uncommon and few cases are reported in literature.[3-4] We report two case of a rare isolated nasopharyngeal and tonsillar involvement in sarcoidosis presented with nonspecific symptoms and cervica node involvement.

CASE 1

A 71-year-old woman presented with nasal obstruction, mainly on the left side for 6 months.

Nasal endoscopy revealed a sub-obstructing nasopharyngeal mass.

Physical neck examination revealed an indolent swelling of the cervical node of the right level II.

A full body CT-scan showed a mass of the left lateral wall of the cavum (Figure 1) and the presence of a single lymphadenopathy of the right level II A, with a maximum diameter of 2,5 cm (Figure 2).

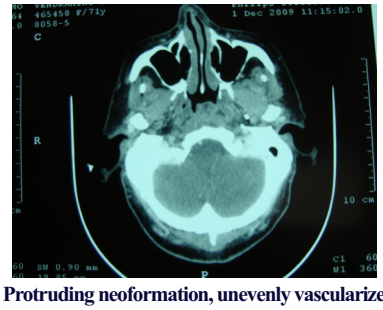

FIGURE 1. Protruding neoformation, unevenly vascularized, occupying the left lateral wall of the cavum. (CT imaging).

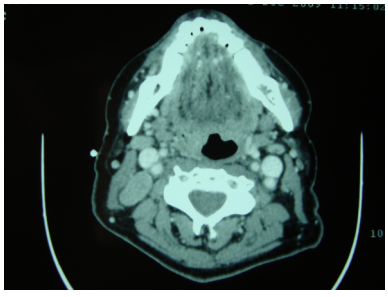

FIGURE 2. Lymphadenopathy of the level II A to the right side with the maximum diameter of 2,5 cm (CT imaging).

An endoscopic biopsy was performed and revealed the following pathological report: "chronic granulomatous phlogistic process with epithelioid cells with rare giant cells and without clear necrosis. This process is reported inside mature lymphatic tissue with mixed phenotype" (Figure 3).

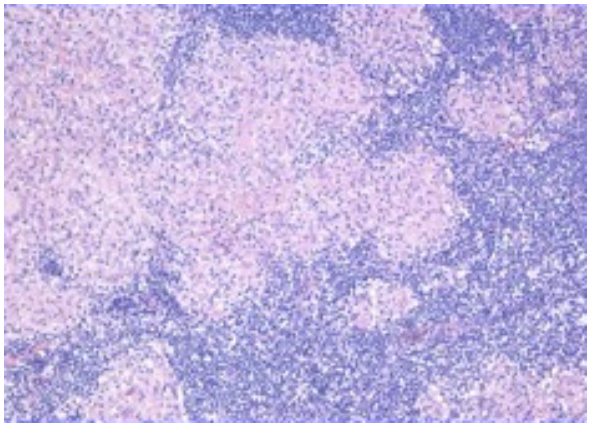

FIGURE 3. Multiple confluent granulomas formed by epithelioid cells, without central caseous necrosis. Nasopharyngeal biopsy. Hematoxylin-eosin, 100x.

Subsequently a Fine Needle Aspiration Biopsy (FNAB) of above mentioned node was performed and the cytological test showed a granulomatous phlogistic process.

Complete laboratory tests, immunological assessment and tests for Mycobacterium Tuberculosis Complex resulted negative.

The treatments performed wer the surgical removal of the nasopharyngeal mass by endoscopic approach and a selective IIa neck dissection.

Histopathologic examination highlighted a chronic granulomatous phlogistic process with epithelioid cells with rare Langhans giant cells, surrounded by a well defined area of lymphocytes, plasma cells, and hystiocytes. The granulomas was barely delimited and confluent without central caseous necrosis. Ziehl-Neelsen staining was executed to rule out infection caused by Mycobacterium tuberculosis was negative. Moreover, the negativity related to PAS (Periodic Acid Shiff) reaction confirmed the absence of other kind of bacilli.

Immunohistochemical examinations of cells expressing CD20 (Figure 4), CD3 (Figure 5) e bcl2 have been implemented with avidin-biotin-peroxidase complex method. CD20-B and CD3-T lymphocytes have been detected mainly around the granuloma.

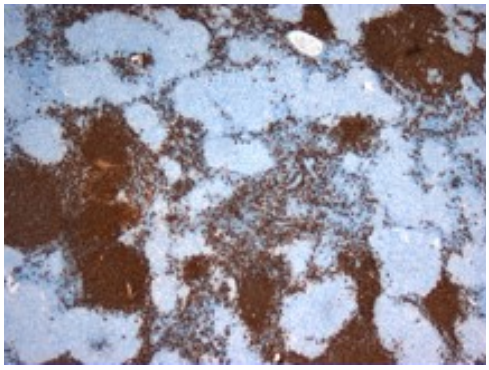

FIGURE 4. CD20-B lymphocytes surrounding granulomas; immun ohist zochemical reaction CD20, 100x.

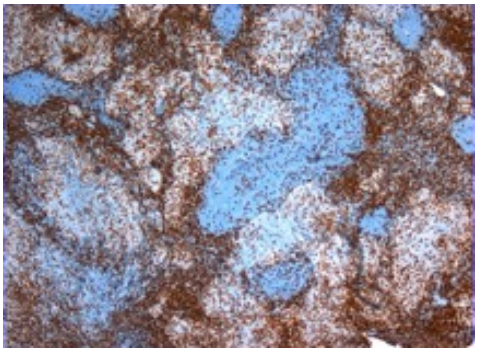

FIGURE 5. CD3-T lymphocytes in the central and peripheral area of granulomas; CD3 immunohistochemical reaction, 400x.

Histological examination of the observed specimen, jointly with the clinical-radiological data, supported a diagnostic hypothesis of sarcoidosis.

The patient was referred to the Rheumatology Department for therapy. After two years no recurrence and no signs of disease progression have been found.

CASE 2

A 32-year-old woman presented a bilateral hypertrophy palatine tonsils (grade IV according to Brodsky classification) and left cervical adenopathy (level IIA) for 5 months without other symptoms. The Magnetic Resonance Imaging (MRI) revealed a thickening of tonsillar tissue (Figure 6) and cervical lymphadenopathy of level IIA on the left side, with a maximum diameter of 1,7 cm (Figure 7).

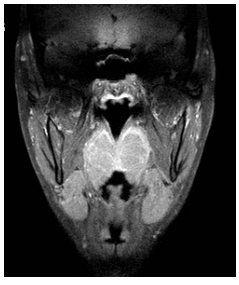

FIGURE 6. Thickening of bilateral tonsillar tissue (MRI T1-weighed with contrast).

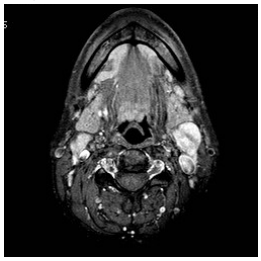

FIGURE 7. Lymphadenopathy of the level II A to the left side with the maximum diameter of 1.7 cm (MRI T1-weighed with contrast).

Chest X-ray was normal. The patient underwent tonsillar biopsy and FNAB of above mentioned node. Histopatological examination showed non-necrotizing granulomas with epithelioid and multinucleated giant cells in both samples. Subsequently a radiofrequency tonsils reduction and a neck dissection of the level IIA on the left was performed. The diagnosis of sarcoidosis was confirmed by histopatological examination (Figure 8). Also in this case the patient was referred to the Rheumatology Department for further follow-up and after one year no recurrence have been found.

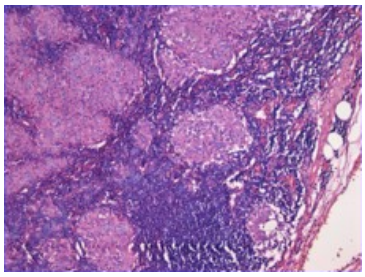

FIGURE 8. Lymph nodal biopsy. Hematoxylin-eosin, 100x.

DISCUSSION

Sarcoidosis affects mainly adult population, the onset is between III and IV decade of life, with higher frequency in women and colored races.[5]

The etiology of sarcoidosis has not been yet defined. The most widely accepted hypothesis is that an environmental or microbiological antigen may trigger an abnormal immune response into genetically susceptible subjects.[1]

The typical histopathological aspect is represented by the sarcoid granuloma constituted mainly by radially positioned differentiated cells of the macrophage line without central caseous necrosis and with little tendency toward convergence. The macrophages, the epithelioid cells, and the Langhans multinucleated giant cells, deriving from the fusion of the previous ones, occupy the central portion and are surrounded peripherally by a lymphoid (Ly Cd4+) and plasmacellular infiltrate.[1]

The association between systemic sarcoidosis and the localized form in the nasopharynx or in the palatine tonsils is variable and not quantifiable; there are no certain epidemiological data because of the rarity of presentation.

The head and neck sites most commonly involved by sarcoidosis are the salivary glands and also the cervical nodes. The parotid gland is affected in about 6% of patient.[6]

Nasopharyngeal involvement in sarcoidosis is difficult to evaluate in absence of specific symptoms, expecially in isolated forms. In the first case the nasal obstruction was the only symptom referred and the cervical lymphadenopathy was an occasional finding. In case of detection of a nasopharyngeal or tonsillar mass, with or without the enlargement of cervical lymph nodes, the diagnosis of malignancy must be firstly excluded. Secondly, the finding of a granuloma on histological examination must exclude other possible disorder as infectious disease (tuberculosis and other mycobacterial infections, fungal infections), vasculitis (Wegener's disease), or other granulomatous disorders such as Crohn's disease.[6]

The disease is suspected in presence of a suggestive clinical presentation and confirmed by endoscopical, radiological and serological data, and by the histological finding of typical non-caseating granuloma on the bioptic specimen.

Radiological investigations as X-ray chest, CT, MRI, FDG-PET (Fluorodeoxyglucose - Positron Emission Tomography) are useful to evaluate the local extension of disease and the possible involvement of other organs.[2,7] In our case there was no systemic involvement.

Laboratory tests do not show clear alterations but they can reveal elevated Angiotensin-converting enzyme levels (40-60% of patients affected) [8], hypercalcemia, hypercalciuria, elevated erythrocyte sedimentation rate (ESR), reversed albumin-globulin ratio, hypergammaglobulinemia and abnormal liver function tests.[1,8]

There are no studies in large series of effectiveness of the treatment of sarcoidosis localized in the upper airways. A codified therapeutic protocol does not exist. The Gold Standard is represented by the corticosteroid treatment and the possible association with immunosuppressive drugs. Corticosteroids are used in sarcoidosis as drug of first choice (20-40 mg/day of Prednisone, reducing this initial dose to a maintenance dose of less than 10 mg/day).[9] Local corticosteroid treatment or with intralesional injections can be used in localised form.[3]

In the localised forms or in those resistant to medical treatment, surgical approach may be useful. Surgical treatment is reserved for cases in which removing obstruction is necessary.[3]

Sarcoidosis has an unpredictable natural story, variable from one patient to another.

Despite the prolonged medical treatment, disease stabilisation is often temporary, and occasionally progresses towards chronicity. However, the systemic nature of the pathology requires a strict follow-up and the management of patients by a specialist multidisciplinary team.

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