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Sarcoidosis (Heerfordt syndrome): A case report

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SUMMARY

We report the case of a 22-year-old woman who is suspected of having primary Sjögren´s syndrome. She complaining of bilateral swelling of eyelids and the parotid glands of three weeks duration. Physical examination revealed a bilateral enlargement of both parotid glands, which were solid and painful. Sjögren´s syndrome was suspected at that stage, and the serologic and specific analysis were done. All these tests didn´t find any autoimmune or visceral features typical of Sjögren´s syndrome and autoantibodies were negative. During follow-up time the right facial nerve palsy developed. Pulmonary radiography revealed bihilar lymphadenopathy and labial salivary gland biopsy revealed non-caseating granuloma. The patient was classified as having stage I sarcoidosis. This case demonstrates the importance of being aware of the leading clinical signs and symptoms in case of Heerfordt syndrome.

Key words: Sjögren syndrome, sarcoidosis, Heerfordt syndrome.

INTRODUCTION

Sarcoidosis is a condition characterized by multiple nodular lesions in the skin, internal organs, eyes, salivary glands, and with enlargement of lymphnodes [1]. Danish ophthalmologist C. F. Heerfordt first described the symptom triad – uveitis, parotid gland enlargement and cranial nerve paresis in 1909 [2]. Sarcoidosis represents a granulomatous disease of obscure etiology. Although cutaneous manifestations of this disease were recognized as early as 1877, it was later demostrated that sarcoidosis was a systemic disorder involving visceral tissues as well [3,4].

Sarcoidosis occurs throughout the world, affecting all genders, races and ages. The disease consistently shows a predilection for adults under age 40, peaking in those aged 20 to 29 [5]. In Scandinavian countries and Japan, there is a second peak incidence in women over age 50 [6]. Most studies suggest a slightly higher disease rate for women. Sweden, Denmark, and USA appear to have the highest prevalence rates in the world [7].

Geographic and environmental factors may explain differences between countries and age groups [8].

An abnormal immune response to a yet unidentified antigen is suspected as the cause.

Recently, using molecular biologic techniques, mycobacteria have been identified in sarcoidal tissues, raising the possibility of *M. tuberculosis* or a related organism as a causative agent [9].

The several pathogenic models have been proposed in recent years that suggest that the disease might result from exposure of genetically predisposed individuals to a kind of specific environmental infectious agent [10, 11]. Among the infectious agents suspected to be involved in the etiology of sarcoidosis are several DNA viruses (Epstein-Barr-virus, human herpes virus, retrovirus, HIV).

Sarcoidosis is usually about 50% asymptomatic upon presentation. Patients may complain of lethargy, chronic fatigue, and anorexia, with specific signs and symptoms related to the organ involved.

The organs most affected are: lungs – 90%, liver – 60%, skin – 25%, eyes – 25%, upper respiratory tract – 20%, parotid glands – 6% of cases [12,13,14].

Heerfordt syndrome – a combination of fever, parotid enlargement, anterior uveitis, and cranial nerve seven palsies.

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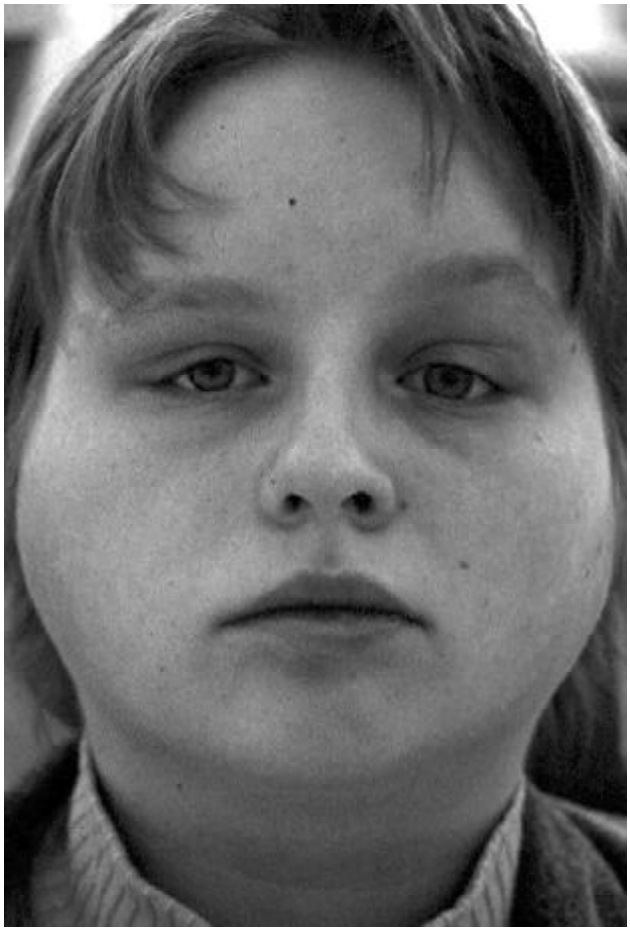

Fig. 1. Bilateral upper lid oedema, enlargement of parotid glands

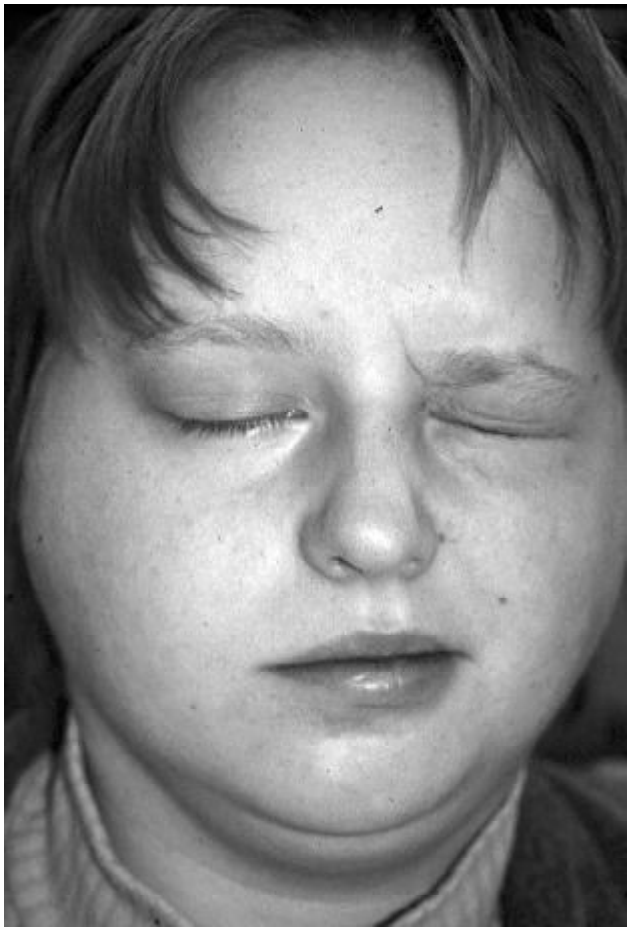

Fig. 2. The 3rd day after hospitalization: facial nerve palsy occurred

This report describes a case of saroidosis having the symptoms triad characteristic for Heerfordt syndrome.

CASE REPORT

A 22-year-old woman suspected of having primary Sjögren's syndrome was referred to the Department of Maxillofacial Surgery, Tartu University Hospital, complaining of bilateral swelling of the eyelids and the parotid glands, also complained *xerostomia* and *xerophthalmia* of three weeks duration and visual disturbance that developed in the evenings. She denied fatigue, fever, night sweats or any other signs of general malaise, otherwise, the patient was in good general health.

Physical examination revealed a bilateral enlargement of both parotid glands, which were solid, plain and painful to pressure, especially right side (Fig.1). On the third day after her arrival in our department right facial nerve palsy and taste disturbance developed (Fig.2).

The following investigations were performed:

1) Laboratory investigations: S-CRP 1 mg/L; S-Prot-Fr-74 g/L; IgM 0,91 g/L, IgG 14,30 g/L; IgA 5,00 g/L; B-ESR 17 mm/H; WBC 6,93x10^9 /L.

2) Serologic investigations S-HZV, S-CMV, Epstein Barr virus (EBV), mumpsvirus IgM negative.

3) Specific analysis: SS-A, SS-B antibodies; Human Leukocyte Antigen (HLA alleels-B7, B8, B27, B35 by PCR method ; ANA–panel analysis. These data of investigations were normal, only IgA level was higher – 5.00 g/L (normal 0,85-4,5 g/L).

Chest radiography revealed bihilar lymphadenopathy. Polycystic circumscribed enlargement of pulmonal hiluses was observed, indicating a stage I sarcoidosis (Fig.3). CT – axial tomogram demonstrated bilateral parotid gland enlargement and destruction foci were observed on the right gland tissue (Fig. 4).

Excision biopsy from the small salivary glands of lower lip was performed. Nodules consisting of epithelioid cells, surrounded by dense fibrous tissue and centrally Langerhans´ type giant cell is observed. There is no evidence of caseation (Fig. 5). Epitheliod cell granuloma without caseous necrosis is the hallmark of sarcoidosis.

The patient was classified as having stage I sarcoidosis.

She recovered completely within 6 weeks under immunosuppressive therapy with steroid, but she con-

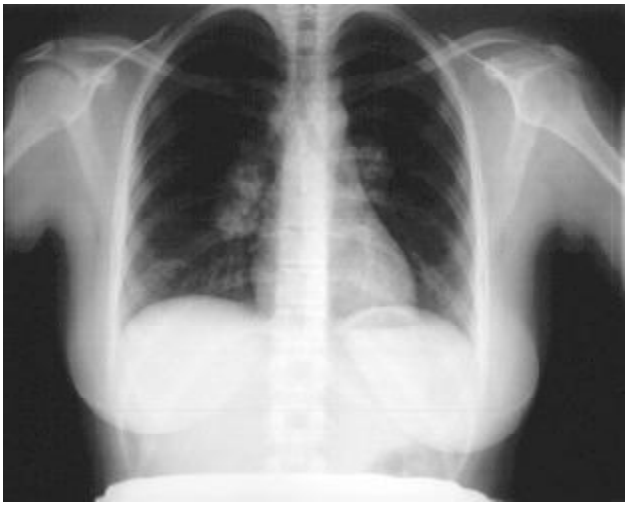

Fig. 3. Chest X-ray showing bilateral lympadenopathy, polycystic circumscribed enlargement of pulmonal hiluses

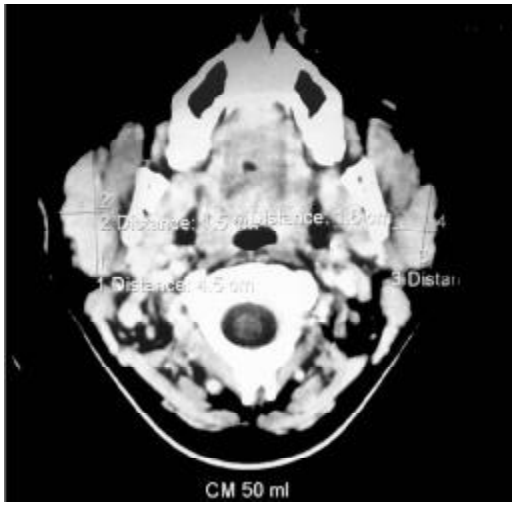

Fig. 4. CT axial tomogram. Bilateral enlargement of parotid glands and destruction foci on the right gland tissue

tinued periodically monitored with chest radiographs and laboratory tests.

DISCUSSION

Sarcoidosis may manifest in different ways. Hilar lymphadenopathy is present in 90% of cases [15], head and neck manifestations in 15% and enlargement of parotid glands in 6% of cases [16].The salivary glands are less commonly involved, with unilateral or bilateral swelling of the parotid gland reported in only 6% to 8% of sarcoidosis patients [17].

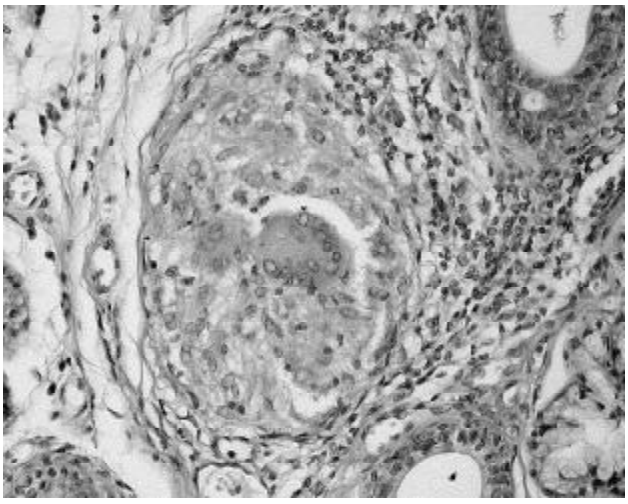

Fig. 5. Section of labial minor salivary gland. Nodule of epithelioid cells, surrounded by dense fibrous tissue. Centrally Langerhans type giant cell, no evidence of caseation (HE x 400)

Although parotid gland enlargement is a common clinical problem, the diagnosis of sarcoidosis can be difficult. Differential diagnosis must include several other systemic diseases, mainly Sjögren´s syndrome.

Xerostomia, xerophthalmia, both symptoms are clinical criteria defining Sjögren´s syndrome. The swelling of the parotid glands and lacrimal glands and also *xerostomia* and *keratoconjunctivitis sicca* in the current case persisted as the leading clinical symptoms for 3 weeks.

In this case specific immunologic markers (HLA) didn´t confirm both diagnosis neither Sjögren´s syndrome nor sarcoidosis.

Only IgA level was elevated in serum, which is often found in case of sarcoidosis [1].

The bihilar lymphadenopathy seen in the chest radiograph, uveitis with blurry vision in the evenings and facial nerve palsy could be interpreted as a signs of sarcoidosis.

In the present case the clinical picture was most compatible with a diagnosis of sarcoidosis. It is important to point out the biopsy obtained from the minor salivary gland showing pathognomnic noncaseating granulomas [18].

This case demonstrates the importance of being aware of the leading clinical signs and symptoms in case of Heerfordt syndrome. These systemic signs and symptoms observed may be quite modest. Therefore chest x-ray, serologic testings and tissue evaluation have a great diagnostic value.

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