

Case Report

Volume 2 Issue 3 – December 2016
DOI: 10.19080/GJO.2016.02.555590

Glob J Otolaryngol

Copyright © All rights are reserved by Ait-el-kerdoudi M

Heerfordt's Syndrome: A New Atypical Clinical Presentation

Ait-el-kerdoudi M*, Chafiki Z, Lakhdim M, Merzouki B, Rouadi S, Abada R, Roubal M and Mahtar M

Department of ENT, Ibn Rochd University Hospital, Morocco

Submission: December 01, 2016; **Published:** December 08, 2016

***Corresponding author:** Ait-el-kerdoudi Mehdi, Department of ENT, Hopital 20 Aout, ENT Service, CHU Ibn Rushd, Casablanca, Morocco.

Abstract

Heerfordt's syndrome is an unusual clinical manifestation (less than 5%) of sarcoidosis, characterized by parotitis, uveitis, and peripheral facial palsy, febrile syndrome is usually associated. Oral corticosteroid is the treatment of choice. We report a case of a patient in whom the syndrome of Heerfordt was retained in an incomplete form thereof.

Keywords: Heerfordt's syndrome; Parotitis; Uveitis; Sarcoidosis; Facial palsy

Introduction

The sarcoidosis is a systemic granulomatosis of unknown cause, characterized by its clinical polymorphism and a wide variety of its modes of presentation. The combination of fever, uveitis, parotitis and peripheral facial paralysis defines Heerfordt's syndrome which presents an unusual manifestation of this disease. We report a case of a patient in whom the syndrome of Heerfordt was found.

Case Report

A 49 year-old female with no medical history, followed in ophthalmology for bilateral uveitis, was referred to the ENT consultation for nasal obstruction with crusting rhinitis. General examination was unremarkable except a low-grade febrile state. Extraoral examination revealed no visible facial swelling neither facial palsy nor cervical lymphadenopathy. Intraoral examination revealed the mucosa to be normally moist. The patient has undergone a rhinocavoscopy which revealed the presence of nodular lesions in the lower horn. A biopsy of the lesions was performed which results normal. A month later, the symptomatology was marked by the appearance of erythema nodosum on the face and the upper limbs (Figure 1). The diagnosis of sarcoidosis was confirmed by the biopsy of the cutaneous lesions which showed non-caseating epithelioid granulomas. Facial CT scan highlighted a multinodular bilateral chronic parotitis (Figure 2), therefore, the diagnosis of Heerfordt's syndrome was retained in view of these highly evocative criteria even if the symptomatology was incomplete given the absence of facial palsy. The patient was initiated on long-term decreasing steroid therapy (Prednisone) that was started at the dose of 1

mg / kg / day. After a month of treatment with oral steroids, clinical symptoms have shown a clear improvement.



Figure 1: Erythema nodosum of the face.



Figure 2: Facial CT scan - frontal section- bilateral parotid enlargement.

Discussion

Sarcoidosis is a chronic inflammatory disorder of unknown etiology, characterized by noncaseating granulomas involving the lungs in more than 90% of patients. Ocular, lymph-node, and cutaneous manifestations are next in frequency, but any organ system can be affected by the disease [1]. Otolaryngologic manifestations of Sarcoidosis are identified in 10-15 % of

patients, the most common being cervical adenopathy [2]. Salivary glands are less frequently involved, unilateral or bilateral swelling of the parotid gland was reported in only 6-8% of patients with sarcoidosis [3]. It can be included in Heerfordt's syndrome (uveoparotid fever). Heerfordt's syndrome is a sarcoidosis syndrome characterized by mild fever, painless parotid gland enlargement, cranial nerve involvement, and anterior uveitis [2].

It is considered as one of the first central nervous system involvements to be described as a neurological presentation of sarcoidosis. The complete form occurs in approximately 0.3% of all cases [4]. The etiology of this condition is still unclear and, as a result, so is the pathogenesis [5]. The incidence of cranial nerve palsy in sarcoidosis is about 5 % [6], with the facial nerve followed by the optic and the trigeminal nerves being the most common nerves involved [7]. Facial palsy forms an important defining component of Heerfordt's syndrome. Its approximate incidence in this syndrome is 25-50%. However, its lack doesn't eliminate the diagnosis as we have noticed in our case, in which the diagnosis of Heerfordt's syndrome was made based on the association of other highly suggestive criteria such as uveitis and parotitis.

The only combination of both uveitis with blurred vision and facial nerve palsy could be performed as sarcoidosis [8]. As it is obviously noted in the literature that eye involvement was the most consistent finding in patients presenting with Heerfordt's syndrome, along with unilateral or bilateral facial palsy or parotid gland swelling [5]. Glucocorticosteroids is the first-line therapy of Heerfordt's syndrome [5]. The diagnosis of Sarcoidotic parotitis requires exclusion of other etiologies, specifically Sialoliths causing an obstruction, and resulting in salivary stasis which may lead to repeated infectious exacerbations [9]. Sjögren's syndrome, which is an autoimmune disease of salivary and tear glands, characterized by xerophthalmia and or xerostomia with or without extra glandular manifestations. It may be primary or secondary when associated with a connective tissue disease, such as rheumatoid arthritis, systemic lupus erythematosus, or scleroderma.

The diagnosis is based on clinical tests such as Schirmer's test and others, the detection of SSA/SSB antibody and abnormal findings of a biopsy of salivary-gland tissue strongly support the diagnosis. Tuberculosis must be also suspected in case of a long lasting parotid swelling, especially, in countries where the disease is rampant as Morocco. Tuberculous parotitis is extremely rare, it accounts for 2.5% to 10% of parotid pathologies [10]. It is likely to simulate a neoplasm [9]. It may occur either as parotitis secondary to primary location in the lung as a result of hematogenous/lymphatic spread or as primary disease due to autoinfection from the oral cavity [11]. Two clinical forms of tuberculous parotitis are, acute tuberculous sialadenitis, and chronic sialadenitis, this latter manifests itself as an asymptomatic localized lesion within the parotid gland, slowly growing in size for many years [11]. The

diagnosis should be suspected in case of personal or a family history of tuberculosis. Imaging finding may be non-specific, but fine-needle aspiration cytology (FNAC) can provide a cytological diagnosis and confirmed later by culture [9].

Other infectious causes of unilateral or bilateral chronic parotitis are also seen due to fungal and parasitic infections. Parotid gland swelling may be encountered in other conditions (neoplasms, sialadenitis, human immunodeficiency virus, anticholinergic medications), it is slow in onset, persistent and usually painless and usually do not disappear spontaneously [9]. In spite of the several differential diagnoses that may be evoked before the symptoms already described, Heerfordt's syndrome is to be considered when at least 2 or more of the symptoms are present especially if the sarcoidotic origin is highly probable. This case emphasizes the importance of recognizing the main signs and clinical symptoms which may be quite modest to indicate the diagnosis of this syndrome.

Conclusion

Head and neck manifestations of Heerfordt's syndrome are non-specific; therefore, a high suspicion is required to diagnose the condition early. In our case, Heerfordt's syndrome was retained given the association of parotitis and uveitis, confirmed subsequently by the histological examination. The presence of peripheral facial paralysis is not necessary to make the diagnosis and must not delay its management.

References

1. Rao DA, Dellaripa PF (2013) Extrapulmonary Manifestations of Sarcoidosis. *Rheum Dis Clin North Am* 39(2): 277-297.
2. Badhey AK, Kadakia S, Carrau RL, Jacob C, Khorsandi A (2015) Sarcoidosis of the Head and Neck. *Head Neck Pathol* 9(2): 260-268.
3. Levent Tabak, E Ağırbaş, Dilek Yılmazbayhan, U Güç (2001) The value of labial biopsy in the differentiation of sarcoidosis from tuberculosis. *Sarcoidosis, vasculitis, and diffuse lung diseases* 18(2): 191-195.
4. Sugawara Y, Sakayama K, Sada E, Kajihara M, Semba T, et al. (2005) Heerfordt syndrome initially presenting with subcutaneous mass lesions: Usefulness of gallium-67 scans before and after treatment. *Clin Nucl Med* 30(11): 732-733.
5. Chappity P, Kumar R, Sahoo AK (2015) Sahoo Heerfordt's Syndrome Presenting with Recurrent Facial Nerve Palsy Case report and 10-year literature review. *Sultan Qaboos University Med J* 15(1): e124-e128.
6. Burns TM (2003) Neurosarcoidosis. *Arch Neurol* 60(8): 1166-1168.
7. Christoforidis GA, Spickler EM, Recio MV, Mehta BM (1999) MR of CNS sarcoidosis: Correlation of imaging features to clinical symptoms and response to treatment. *AJNR Am J Neuroradiol* 20(4): 655-669.
8. Boucher RM, Grace J, Java DJ (1994) Sarcoidosis presenting as multiple cranial neuropathies and a parotid mass. *Otolaryngol Head Neck Surg* 111(5): 652-655.
9. BN Gayathri, R Kalyani, K Manjula (2011) Primary tuberculous parotitis. *Journal of Cytology* 28(3): 144-145.
10. Iseri M, Aydinler O, Celik L, Peker O (2005) Tuberculosis of the parotid gland. *J Laryngol Otol* 119: 311-313.
11. Hamdan AL, Hadi U, Shabb N (2002) Tuberculous parotitis: a forgotten entity. *Otolaryngol Head Neck Surg* 126(5): 581-582.

**Your next submission with JuniperPublishers
will reach you the below assets**

- Quality Editorial service
- Swift Peer Review
- Reprints availability
- E-prints Service
- Manuscript Podcast for convenient understanding
- Global attainment for your research
- Manuscript accessibility in different formats
(Pdf, E-pub, Full Text, Audio)
- Unceasing customer service

**Track the below URL for one-step submission
<http://juniperpublishers.com/online-submission.php>**