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Clinical Case of Heerfordt-Waldenstrom Syndrome in Its Complete Form

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Abstract

Heerfordt-Waldenström syndrome (SHW) is a rare manifestation of sarcoidosis whose diagnosis is essentially clinical. This syndrome presents clinically in two forms: the complete form, which constitutes 0.3% of all cases of sarcoidosis, associating facial paralysis, hypertrophy of the parotid glands, anterior uveitis and low-grade fever. Treatment is based on high-dose corticosteroid therapy with a very favorable prognosis. There are very few cases in the literature, hence the interest of reporting this case of SHW in its complete form.

I. Introduction

Heerfordt-Waldenström syndrome (SHW) is a rare manifestation of sarcoidosis whose diagnosis is essentially clinical. This syndrome presents clinically in two forms: the complete form, which constitutes 0.3% of all cases of sarcoidosis, associating facial paralysis, hypertrophy of the parotid glands, anterior uveitis and low-grade fever. Treatment is based on high-dose corticosteroid therapy with a very favorable prognosis. There are very few cases in the literature, hence the interest of reporting this case of SHW in its complete form.

II. Clinical Case

A 32-year-old male patient, with no notable pathological history, no alcohol or tobacco intoxication, who presented, intermittently and for six months before his admission, bilateral ocular redness with a slight decrease in visual acuity, evolution was marked by the appearance of facial asymmetry and swelling of the left parotid region, the clinical examination revealed left peripheral facial paralysis **(figure 1)**, bilateral ocular redness **(figure 2)**, painless unilateral left parotidomegaly **(figure 3)** confirmed by Cervical MRI **(figure 4)**. The biological assessments, especially the blood count, did not show lymphopenia or hyperleukocytosis, also the blood ionogram, the phosphocalcic balance and the CRP were normal, the angiotensin converting enzyme was slightly elevated.

The ophthalmological examination revealed bilateral anterior uveitis, the chest CT scan revealed mediastinal lymphadenopathy, the tuberculin skin test was negative, a biopsy of the accessory salivary glands was performed showed a granuloma without caseous necrosis, the diagnosis of Heerfordt syndrome was based on the clinical tetrad and on the histological confirmation. A high-dose of Oral corticosteroid therapy was started which allowed a clear improvement of the symptoms.

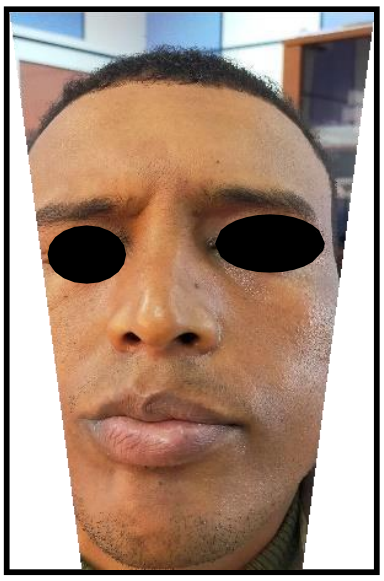

Figure 1 : Left peripheral facial paralysis.

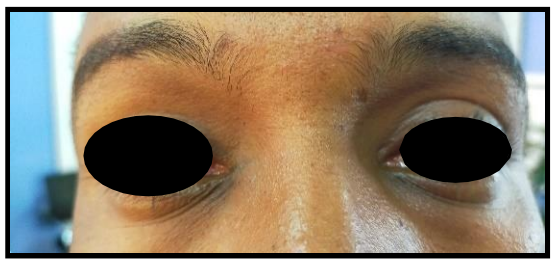

Figure 2 : Bilateral ocular redness.

Figure 3: Unilateral left parotidomegaly.

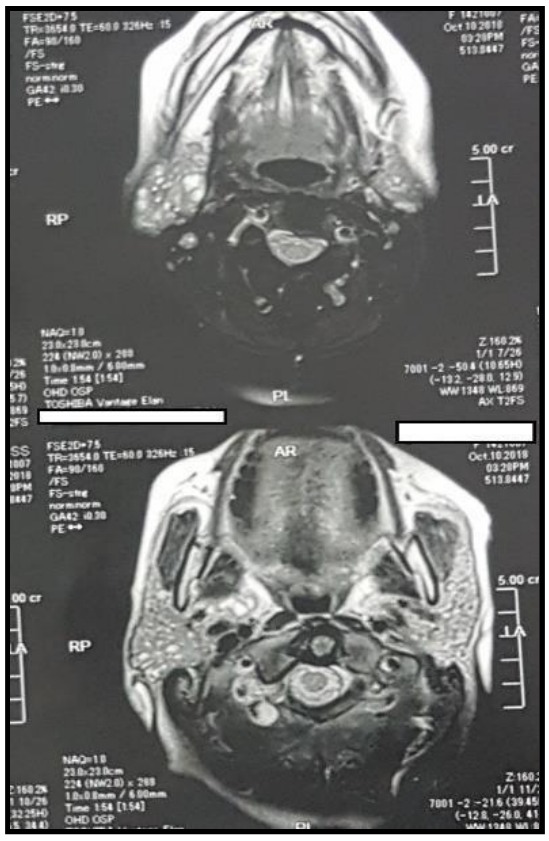

Figure 4: MRI showing bilatéral parotidomegaly

III. Discussion

Heerfordt-Waldenström syndrome (HWS) is a rare and unusual manifestation of sarcoidosis, it represents 5 % to 10 % of the forms of this disease and which it may be a mode of revelation **[1]**.

In 1909 the disease was first described by Heerfordt and in 1937 Waldenström linked this syndrome to sarcoidosis **[2.3]**.

This syndrome is more common in women, and it's characterized by the presence of a clinical tetrad including facial nerve palsy, hypertrophy of the parotid glands, anterior uveitis, and low-grade fever **[1]**.

Clinically, Heerfordt syndrome comes in two forms, the complete form combining the four main symptoms and constituting 0.3 % of all cases of sarcoidosis, while the presence of two or three symptoms constitutes the incomplete form **[4-5]**.

Darlington P et al conducted a study of 1000 patients with sarcoidosis, 16 patients (1.6 %) among them had Heerfordt syndrome, 03 (0.03 %) patients had the complete form, 13 patients (0.13 %) had the incomplete form and all had uveitis **[4-5]**. which confirms the rarity of the complete form of this syndrome. Our patient presented the complete form.

In about 6 % of cases of sarcoidosis, there is an enlargement of the parotid gland, which is due to a granulomatous inflammatory reaction. This hypertrophy is more common in women, and bilateral involvement was found in 73 % of cases **[6]**.

Neurosarcoidosis is present in approximately 5 % of patients with systemic sarcoidosis, it affects the central or peripheral nervous system, the cranial nerves can be affected: the facial nerve and the optic nerve are the most frequently affected. Facial paralysis is an important determinant of Heerfordt syndrome, with an incidence of 25-50 %. These lesions are due to spiny and perineural granulomas **[6.2]**.

Sarcoidosis affects the eyes in 30 to 60 % of cases. the different structures of the eye can be affected, but anterior or posterior uveitis remains the most common condition, often bilateral and with a favorable prognosis, rarely involving the visual prognosis **[5.7]**.

The etiology of this syndrome is still ambiguous, moreover, the pathology of neurosarcoidosis is due to a non-caseating epithelioid granuloma **[5]**, this granuloma does not exist only in sarcoidosis, diseases such as tuberculosis, fungal and parasitic infections, wegener's granulomatosis or leptomeningeal lymphoma can also have such granulomas and must be excluded by appropriate clinical and para-clinical investigations **[7]**.

The diagnosis of HWS syndrome is clinical **[2]**. The histo-pathological examination shows a giganto-cellular epithelioid granuloma without caseous necrosis, these granulomas produce angiotensin converting enzyme, but the levels do not always correlate with disease activity (10 % false positives and 40 % false negatives) **[2]**.

A therapeutic protocol has not yet been established due to the rarity of this syndrome, and therefore multidisciplinary care is required, involving otorhinolaryngologists, pulmonologists and ophthalmologists.

However, corticosteroids remain the first-line treatment for HWS. Our patient was put on high-dose corticosteroid therapy for two weeks followed by a progressive decrease **[8]**. The evolution was favorable with regression of parotid swelling and recovery of peripheral facial paralysis after one month of treatment.

The prognosis of sarcoidosis depends on the duration of the disease, its severity and the organs affected. It is usually a self-limiting disease and recovery is achieved in 12 to 36 months. Some prolonged cases have been reported. The mortality rate varies between 1 % and 5 % **[9]**.

IV. Conclusion

HWS is a rare variant of sarcoidosis, whose diagnosis is essentially clinical; it must always be kept in mind, and we know how to evoke it each time we are faced with its complete or incomplete form.

V. Conflict of Interests: The authors declare that there is no conflict of interest regarding the publication of this paper.

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