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Short Communication

Heerfordt Syndrome: About a Case and a Review of the Literature

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Introduction

Heerfordt syndrome is a rare form of sarcoidosis manifested by a picture of uveitis, fever, paratidomegaly and facial paralysis. This picture was described by Heerfordt in 1909 and Waldenstrom in 1937 linked this syndrome to sarcoidosis [1].

The latter is a chronic inflammatory disease with an incidence of 3 to 10 per 100,000 inhabitants. It is dominated by respiratory manifestations but remains multifocal [2].

The diagnosis is based on the histopathological study of salivary secretions. The outcome is often favorable with total remission.

Treatment is based on oral corticosteroid therapy after a pre-therapeutic assessment.

Patient and Observation

This is a 21-year-old patient with no particular pathological history admitted to the emergency room for poor visual acuity and facial asymmetry that had been developing for several days. The latter had a sudden onset with a general preserved, afebrile condition. Examination of the face revealed right facial paralysis and a positive Charles-Bell sign, as well as total swelling of the ipsilateral parotid gland.

Ophthalmological examination confirmed the presence of bilateral granulomatous, synechiant and non-hypertensive anterior uveitis.

The biological examination showed a biological inflammatory syndrome, preserved renal function. Ultrasound confirmed diffuse right parotid hypertrophy. Chest CT revealed mediastinal and para-hilar lymphadenopathy.

Faced with this clinical trial, Heerfordt syndrome has been suggested; a biopsy of the salivary glands was performed followed by a pathological examination confirming the presence of an inflammatory reaction with gigantocellular granuloma without caseous necrosis.

After a pre-therapeutic assessment, corticosteroid therapy of 1 mg/kg was started with local ocular means.

From the third month a notable regression of the paralysis with a spectacular improvement in ocular signs.

There are no conflicts of interest between the authors and they accept publication

Discussion

Sarcoidosis, also known as Besnier-Boeck-Shaumann disease (BBS) is a rare disease. Heerfordt syndrome represents 5 to 10% of sarcoidoses and 4.4 to 5.6% as reported by Fukuhara et al [3]. This syndrome is an active form of sarcoidosis, it has a complete form if the picture is total or incomplete if there is the presence of at least two signs.

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In our study we report the case of a 21-year-old woman. This is consistent with the literature which describes a female predominance due to hormonal factors with a second peak between 45-65 years of age [4].

Ocular manifestations are constant depending on the series, they are found in 25 to 60% of cases and are indicative of the disease in 10 to 20% of cases [5]. It is mainly anterior uveitis in 70 to 75% of cases, which is quite characteristic as we observed in our study. On the other hand, posterior uveitis is more common in white people, between 65 and 83% [6].

All structures of the eyeball may be involved. This is sometimes indicative of illness.

Involvement of the accessory salivary glands is present in 65% of cases with rare involvement of the main salivary glands sometimes leading to bilateral parotitis (observed in 1 to 4% of cases) with xerostomia [3,7]. In our study the biopsy came back positive.

Fatigue is a common symptom in patients with sarcoidosis, occurring in up to 80% of patients [8]. The level of fatigue seems to be associated with the presence of extrapulmonary sarcoidosis [9].

Sarcoidosis can cause a diffuse goiter or rarely a solitary thyroid nodule [10,11]. Almost all cases are euthyroid, although cases of clinical hypothyroidism caused by diffuse thyroid replacement have been reported [12,13].

Pathogenically, the lesions are of autoimmune origin in reaction to an unknown antigen (bacteria, virus) leading to a cytokine disorder, in particular an increase in the secretion of Tumor Necrosis Factor (TNFα), which participates in the formation granulomas in different organs. Usually sporadic, SNELL had described two observations of Heerfordt syndrome in two sisters [14].

It is typical to note an elevation of Angiotensin Converting Enzyme (ACE) in 60% of cases. This elevation is linked to abnormal production of ACE by activated macrophages and epithelioid cells in granulomas.

The tuberculin IDR is negative in 80% of cases (tuberculous anergy) and takes on its full value in the event of previous positivity. Quantiferon is negative in our patient.

On an evolutionary level, spontaneous regression is possible. The treatment allows for remission of clinical signs over a period of 12 to 36 months. The occurrence of pulmonary localization and the spread of the disease can be seen in the long term. Mortality is between 1 to 5% [1].

The differential diagnosis arises with tuberculosis, Wegener's disease, malignant lymphomas, Castleman's disease. Therapeutic treatment is essentially medical and immediately finds its indication in the face of ocular and neurological damage (facial paralysis). Corticosteroid therapy represents the first-line treatment for Heerfordt syndrome. The systemic route with a dose of 20 to 40 mg/day (some cases may benefit from 80 mg/d) for 1 to 3 months then reduce the dose by 5 to 10 mg/d every 2 to 4 weeks to reach a dose maintenance of 5 to 10 mg/day for 6 to 9 months [1-15].

Conclusion

Heerfordt syndrome is a rare manifestation of sarcoidosis. Diagnosis is easy if the form is complete, but you should always think about it in atypical cases. The treatment is medical and the prognosis is often good.

In refractory or diffuse forms, the approach must be multidisciplinary.

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