

Heerfordt's Syndrome

An interesting and under-recognised manifestation of sarcoidosis

Jeanie McGee and *Alireza Minagar

متلازمة هيرفورد

ظاهرة من مظاهر الساركويد غير المألوفة و المثيرة للاهتمام

جيني ماغي وعلي رضا ميناچار

SARCOIDOSIS IS A SYSTEMIC GRANULOMATOUS disease which principally involves the pulmonary and lymphatic systems. However, such a formal definition of sarcoidosis is simplistic since it can resemble many other diagnoses and may therefore have 'a thousand faces'. In this issue of SQUJ, Chappity *et al.* describe an atypical case of Heerfordt's syndrome and discuss findings from a systematic review of the literature over the last 10 years.¹ Heerfordt's syndrome is an atypical manifestation of acute sarcoidosis presenting with fever, *uveitis* and swelling of the parotid glands and, most often, unilateral facial nerve palsy. Because most patients with Heerfordt's syndrome present with atypical symptoms at onset, many are misdiagnosed or remain undiagnosed. In Chappity *et al.*'s case report, a 52-year-old woman had recurrent facial nerve palsy on two different occasions four months apart; at first unilateral and, later, bilateral. The patient reported a recent history of swelling of the parotid region and low-grade fever. She reported no significant visual disturbances or other ophthalmological symptoms. However, upon examination, the patient was found to have anterior intermediate *uveitis*. She was subsequently diagnosed with Heerfordt's syndrome and treated with steroids.¹

The authors present an interesting clinical example of an atypical presentation of Heerfordt's syndrome, emphasising the need for a careful comprehensive examination of patients presenting with facial nerve palsy.¹ Historically, a clear presentation of Heerfordt's syndrome involves the onset of unilateral facial nerve palsy, parotid gland swelling, fever, night sweats, weight loss and optical disturbances.^{2,3} However, as the authors observed in their review of published literature, most cases of Heerfordt's syndrome present with ambiguous manifestations and do not fall within

the standard norms of diagnostic criteria.¹ Due to the fact that most cases of unilateral facial nerve palsy with an unknown aetiology are diagnosed as Bell's palsy, there is a considerable need for a systematic approach in the recognition and diagnosis of Heerfordt's syndrome. Sudden onset of facial nerve palsy is common in 25–50% of patients with Heerfordt's syndrome and is normally preceded by enlargement of the parotid gland.^{1,4} Other potential indications of the syndrome include sensorineural hearing loss, labyrinthine involvement with vestibular dysfunction, epistaxis, nasal pain or obstruction, low-grade non-specific fever, night sweats and visual disturbances.^{1–3}

In their systematic review, Chappity *et al.* found that the most commonly documented features associated with Heerfordt's syndrome included symptoms or signs demonstrating the involvement of the seventh or fifth cranial nerves; the presence of parotid swelling; fever, and an array of ophthalmic anomalies.¹ These anomalies included *panuveitis*, posterior and anterior *uveitis*, bilateral granulomatous *uveitis*, bilateral swelling of the eyelids and *myodesopsia* (floaters), with cytology demonstrating either parotid or pre-auricular lymph node nonceasing granulomas. Other clinical features often associated with this syndrome, including night sweats and weight loss, were not commonly reported in the recent literature.¹

The findings of Chappity *et al.*'s case report and systematic literature review underline the importance of obtaining a comprehensive and current medical history from patients presenting with facial nerve palsy.¹ For neurologists and other clinicians, recognising the significance of a patient's self-reported symptomology within the past week is critical to the differential diagnosis and consideration of Heerfordt's syndrome.⁴ Patients presenting with either unilateral or bilateral facial nerve palsy should be questioned

regarding and assessed for visual disturbances and an ophthalmology consultation should be secured immediately. The presence of fever, night sweats, a cough, throat swelling, difficulty swallowing or even headaches could indicate a potential case of Heerfordt's syndrome.^{5,6}

In conclusion, the authors have done an excellent job in educating the clinician regarding the rare diagnosis of Heerfordt's syndrome and recognising the need for an updated awareness of its clinical features.¹ Early diagnosis and treatment with steroid therapy can prove effective in reducing morbidity and promoting recovery. The alert clinician may therefore need to pursue additional diagnostic work-up beyond the initial clinical assessment as well as seek to obtain a comprehensive medical history when evaluating patients with facial weakness.^{1,6-8}

References

1. Chappity P, Kumar R, Sahoo AK. Heerfordt's syndrome presenting with recurrent facial nerve palsy: Case report and 10-year literature review. *Sultan Qaboos Univ Med J* 2015; 15:129.
2. Denny MC, Fotino AD. The Heerfordt-Waldenström syndrome as an initial presentation of sarcoidosis. *Proc (Bayl Univ Med Cent)* 2013; 26:390–2.
3. Tamme T, Leibur E, Kulla A. Sarcoidosis (Heerfordt syndrome): A case report. *Stomatologija* 2007; 9:61–4.
4. Joseph FG, Scolding NJ. Sarcoidosis of the nervous system. *Pract Neurol* 2007; 7:234–44. doi: 10.1136/jnnp.2007.124263.
5. Dua A, Manadan A. Images in clinical medicine: Heerfordt's syndrome, or uveoparotid fever. *N Engl J Med* 2013; 369:458. doi: 10.1056/NEJMicm1303454.
6. Sharma SK, Soneja M, Sharma A, Sharma MC, Hari S. Rare manifestations of sarcoidosis in modern era of new diagnostic tools. *Indian J Med Res* 2012; 135:621–9.
7. Kimura J. Electrodiagnosis of the cranial nerves. *Acta Neurol Taiwan* 2006; 15:2–12.
8. Sugawara Y, Sakayama K, Sada E, Kajihara M, Semba T, Higashino H, et al. Heerfordt syndrome initially presenting with subcutaneous mass lesions: Usefulness of gallium-67 scans before and after treatment. *Clin Nucl Med* 2005; 30:732–3. doi: 10.1097/01.rlu.0000182264.76461.74.