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(CASE REPORT)

Systemic sarcoidosis revealed by Heerfordt's syndrome: A case report

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Abstract

Heerfordt's syndrome (or evaporated fever) is a rare clinical form of sarcoidosis (occurring in less than 5% of cases) combining uveitis, parotitis, fever, and facial nerve palsy. We report the case of a 46-year-old female patient in whom isolated peripheral facial palsy preceded the onset of pulmonary, cutaneous, and ophthalmological involvement by several months. The diagnosis of systemic sarcoidosis (radiological stage II) was confirmed by skin biopsy after the formal exclusion of tuberculosis. The patient's condition improved favorably under corticosteroid therapy.

Keywords: Sarcoidosis; Heerfordt's Syndrome; Facial Palsy; Granuloma; Tuberculosis

1. Introduction

Sarcoidosis is a systemic granulomatosis of unknown etiology. Heerfordt's syndrome, first described in 1909, represents a historical and rare variant, classically defined by the association of fever, parotid gland enlargement, anterior uveitis, and facial nerve palsy. Its clinical presentation is frequently incomplete or temporally dissociated, making the initial diagnosis complex. We report here a case illustrating this diagnostic challenge, where an inaugural facial palsy preceded the full systemic picture.

2. Case Report

We report the case of a 46-year-old female farmer, exposed to wood smoke and poultry, with a history of hysterectomy and thyroid nodules, admitted for the investigation of symptoms evolving over four months. The initial clinical picture was marked by a transient right peripheral facial palsy, followed by the appearance of a generalized macular rash sparing the palms and soles, exertional dyspnea associated with chest pain, dysphonia, dysphagia, and a bilateral decrease in visual acuity with xerostomia, all occurring in a context of general health deterioration.

Physical examination upon admission revealed cervical and supraclavicular lymphadenopathy, scarring skin lesions, alopecia, as well as active bilateral anterior uveitis with a positive Schirmer's test. Paraclinical investigations demonstrated a micronodular interstitial syndrome with peri lymphatic distribution associated with mediastinal lymphadenopathy (radiological stage II) and splenic involvement on the thoraco-abdomin-pelvic CT scan, while cervical ultrasound revealed chronic bilateral parotitis.

Laboratory tests showed hypercalciuria and anicteric cholestasis with a normal angiotensin-converting enzyme (ACE) level. The infectious workup (GeneXpert PCR, sputum smear microscopy for acid-fast bacilli) allowed for the exclusion of tuberculosis. The diagnosis of systemic sarcoidosis revealed by Heerfordt's syndrome was confirmed by the demonstration of epithelioid and giant cell granulomas without caseous necrosis on skin biopsy. The outcome was favorable under oral corticosteroid therapy (40 mg/day), with regression of cutaneous and respiratory symptoms.

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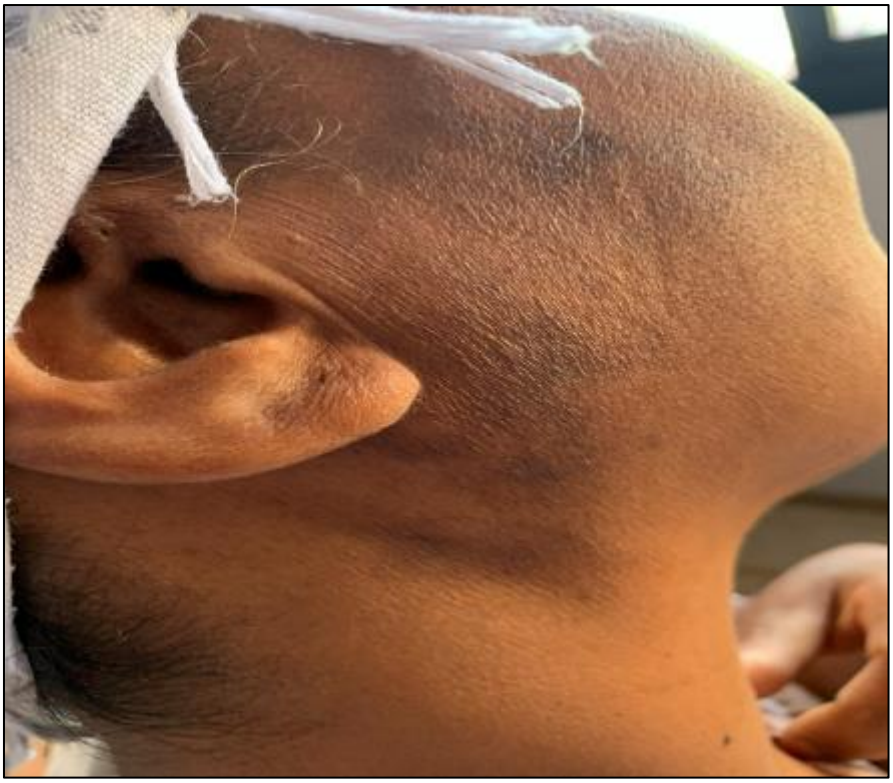

Figure 1 Right parotid swelling

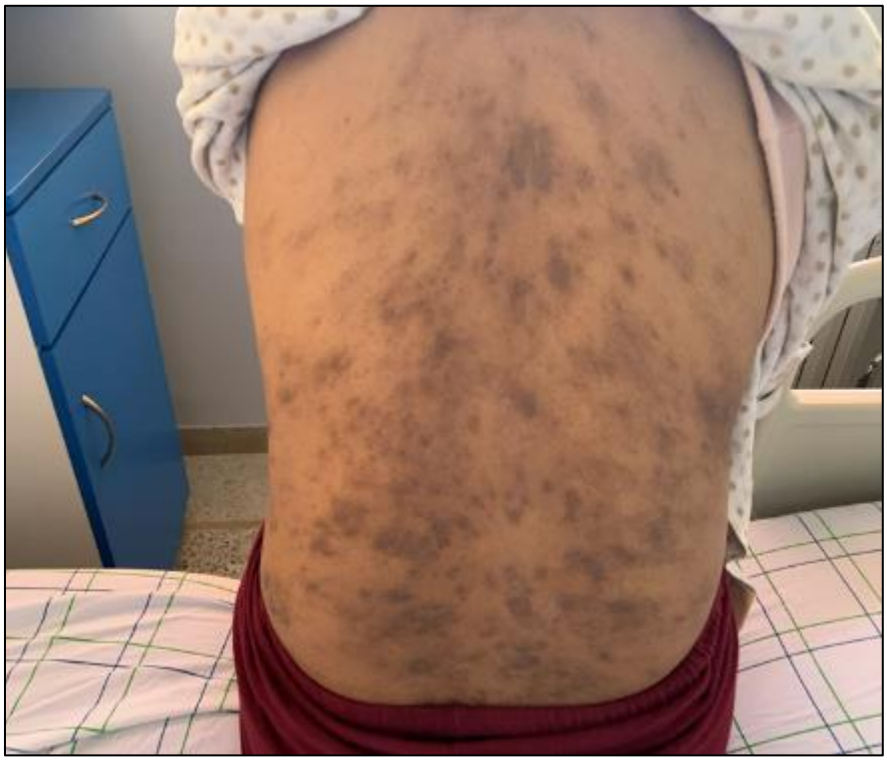

Figure 2 Diffuse macular lesions on the back in the process of healing

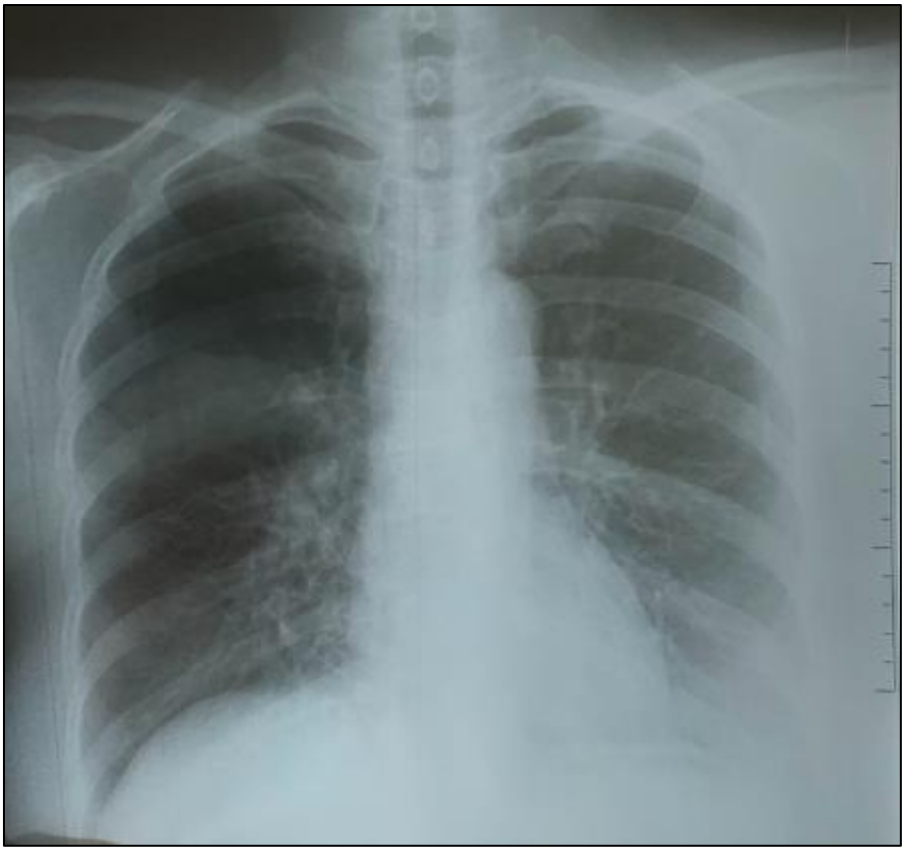

Figure 3 Chest X-ray showing a diffuse bilateral interstitial syndrome with right hilar lymphadenopathy

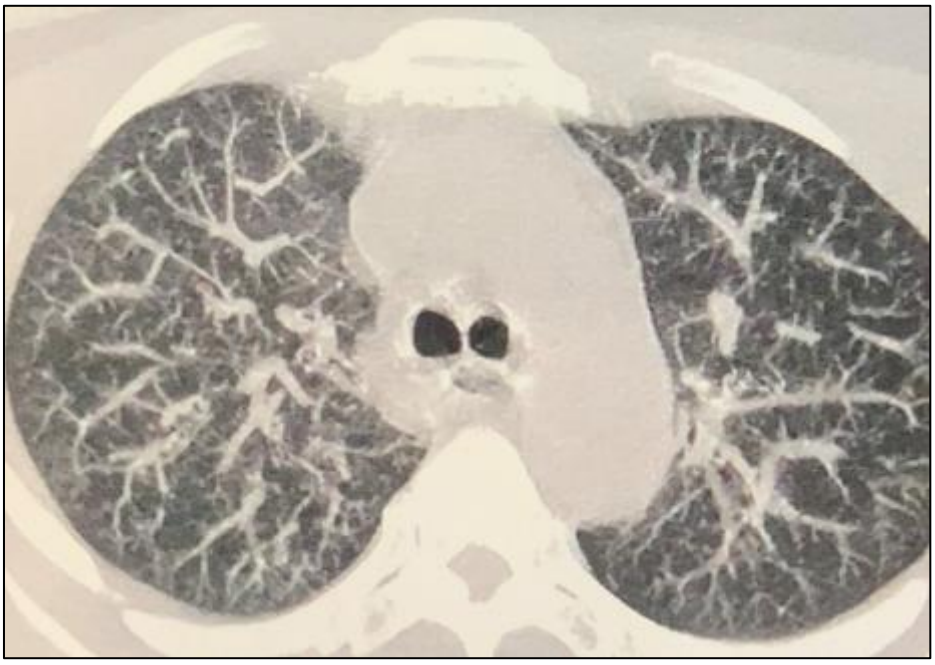

Figure 4 CT scan section showing a peri lymphatic distribution of micronodules, typical of sarcoidosis

3. Discussion

Heerfordt's syndrome, or evaporated fever, constitutes a rare and historical phenotypic form of systemic sarcoidosis, affecting less than 5% of patients and defined by the pathognomonic tetrad associating uveitis, parotitis, fever, and facial nerve palsy [1-3]. While a complete presentation at onset is exceptional, our observation illustrates the insidious and temporally dissociated nature of this syndrome, marked here by an inaugural and isolated peripheral facial palsy preceding mediastina-pulmonary involvement by several months.

This involvement of the 7th cranial nerve, the most common manifestation of neurosarcoidosis (25 to 50% of nerve involvements) [4, 5], typically results from direct compression by granulomatous infiltration or basilar meningitis. Its early occurrence requires increased diagnostic vigilance to avoid overlooking subclinical parotitis, which in our patient was only detected via ultrasound [6].

In a context of high tuberculosis endemicity such as Morocco, the diagnostic challenge is significant. The formal exclusion of tuberculosis, whose radio clinical mimicry is perfect (fever, weight loss, interstitial involvement), constitutes an absolute prerequisite, validated here by the negative results of molecular microbiological investigations [7, 8]. Furthermore, the angiotensin-converting enzyme (ACE) level, which is normal in nearly 40% of active forms, cannot be considered a reliable exclusion marker due to its low negative predictive value [9].

Definitive diagnosis relies ultimately on histological evidence of an epithelioid and giant cell granuloma without caseous necrosis. This is easily obtained via skin or minor salivary gland biopsy (yield of 20 to 60%) [10], allowing for the early initiation of systemic corticosteroid therapy (0.5 to 1 mg/kg/day). This treatment is imperative given the functional threats to vision (uveitis) and the nervous system, and its rapid efficacy confirms the inflammatory and granulomatous nature of the pathology [11, 12].

4. Conclusion

Heerfordt's syndrome is a rare entity that should be considered in any case of unexplained peripheral facial palsy, by systematically searching for parotid involvement (clinical or sonographic) and uveitis. Histological confirmation, easily accessible via skin or salivary biopsy, and the rigorous elimination of tuberculosis in endemic zones, allow for early and effective management.

Compliance with ethical standards

Disclosure of conflict of interest

The authors declare no conflicts of interest.

Statement of informed consent

All participants included in the study provided informed consent.

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