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The Clinical Picture

Bilateral parotitis and facial nerve palsy

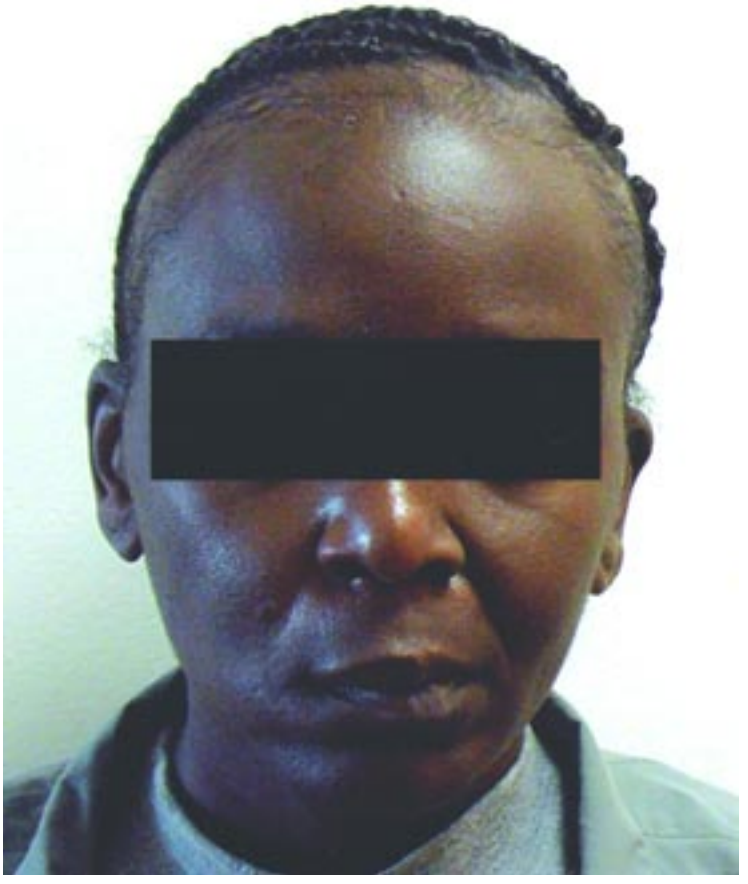

FIGURE 1

A 42-YEAR-OLD BLACK WOMAN presents with tender bilateral parotid enlargement. About 4 weeks after onset of the parotitis, she developed unilateral facial nerve palsy.

A review of systems elicits subjective fevers, arthralgias, xerostomia, keratoconjunctivitis sicca, and dyspnea on exertion. She has a history of eczema. She was not taking any medications at the time her symptoms developed.

Physical examination shows bilateral parotid enlargement, right facial nerve palsy, and a small flesh-colored papule near the right nasolabial fold (FIGURE 1). An ophthalmologic examination reveals anterior uveitis.

Q: Which underlying condition is most likely in this patient?

- ❑ Sjögren syndrome
- ❑ Tuberculosis
- ❑ Sarcoidosis
- ❑ Mumps
- ❑ Human immunodeficiency virus (HIV) infection

A: The patient's presentation is consistent with uveoparotid fever secondary to sarcoidosis, a condition also known as Heerfordt syndrome, which is an occasional variant of sarcoidosis consisting of fever, facial nerve palsy, anterior uveitis, and parotid gland enlargement.

HEERFORDT SYNDROME

The syndrome was first described in 1909 by Danish ophthalmologist Christian Frederick Heerfordt, who believed mumps virus to be the cause.[1] (Indeed, our patient was initially referred to us for consideration of mumps infection despite prior vaccination.) It was not until 1937 that Waldenström recognized the condition as a manifestation of sarcoidosis, a multiorgan systemic disease characterized by noncaseating granulomas.[2]

Parotitis

Parotitis occurs in 6% of patients with sarcoidosis. It is bilateral in about 75% of cases

and is more common in women. Parotid sarcoidosis generally occurs in the setting of multi-organ systemic disease, and patients may also present with peripheral and intrathoracic lymphadenopathy, pulmonary disease, uveitis, and skin lesions.[3]

The differential diagnosis for parotid gland enlargement is extensive and includes Sjögren syndrome, sialadenitis, tuberculosis, cat-scratch disease, toxoplasmosis, actinomycosis, mumps, human immunodeficiency virus infection, neoplasm, hepatitis C, and bulimia. However, none of these diseases is associated with both anterior uveitis and facial nerve palsy.

Nerve involvement

Neurologic manifestations occur in 5% of patients with sarcoidosis. Facial nerve palsy is the most common presentation and is bilateral in half of cases. When unilateral, it affects the right and left facial nerves with equal frequency. With therapy, facial palsy resolves completely in 80% of patients.[4]

The differential diagnosis of facial nerve palsy includes Lyme disease, orofacial granuloma, HIV or human T-lymphotrophic virus 1 infection, Ramsey-Hunt syndrome, the Miller-Fisher variant of Guillain-Barré syndrome, and idiopathic Bell palsy.

Diagnostic steps

Sarcoidosis is diagnosed by histologic evidence of noncaseating granulomas in the absence of other causes (eg, malignancy or infection) in patients with an appropriate clinical presentation. Our patient had bilateral hilar adenopathy, and noncaseating granulomas were seen in biopsy specimens of a skin tag and a mediastinal lymph node. Her parotitis, uveitis, Bell palsy, and fever made the more specific diagnosis of Heerfordt syndrome.

In the United States, sarcoidosis is more prevalent in blacks than in whites (lifetime prevalence 2.4% vs 0.85%).[1]

THERAPY AND OUTCOME

Systemic corticosteroids are the mainstay of treatment for sarcoidosis that is life-threatening or is causing organ dysfunction.[5] Cytotoxic drugs and antimetabolites have also been used anecdotally for severe sarcoidosis, but benefit has not been proven.

Our patient received prednisone, and her condition improved gradually. ■

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