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M. Carter Denny & A. Domnica Fotino

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The Heerfordt-Waldenström syndrome as an initial presentation of sarcoidosis

M. Carter Denny, MD, MPH, and A. Domnica Fotino, MD, MPH

Sarcoidosis is a granulomatous disease of unclear etiology, which commonly presents with cough, dyspnea, chest pain, fever, weight loss, arthralgias, and erythema nodosum. Heerfordt-Waldenström syndrome, a rare presentation of sarcoidosis, is characterized by the presence of parotid gland enlargement, facial palsy, anterior uveitis, and fever. Here we present a case of a 59-year-old nonsmoking African American woman who presented with 3 days of progressively worsening left facial droop, difficulty swallowing, and blurred vision. Over the prior 4 months, she had had a productive cough, fevers, night sweats, and an unintentional 30-pound weight loss. Physical examination revealed a left facial droop involving the forehead, cheek, and chin with an inability to close the left eyelid. Her serum angiotensin-converting enzyme level was twice the upper limit of normal. Prominent hilar markings were identified on chest x-ray, but no focal opacity was seen. Fine-needle aspiration of a preauricular lymph node revealed noncaseating granulomas consistent with granulomatous lymphangitis. The patient was given a diagnosis of Heerfordt-Waldenström syndrome, or uveoparotid fever. Treatment with a high-dose steroid improved her parotid gland enlargement, facial palsy, and anterior uveitis.

CASE REPORT

A 59-year-old nonsmoking African American woman presented with a 3-day history of progressively worsening left facial droop, difficulty swallowing, and blurred vision. Over the prior 4 months, she had had a productive cough, fevers, night sweats, and an unintentional 30-pound weight loss. Physical examination revealed a swollen left cheek and left facial droop involving the forehead, cheek, and chin. She was unable to close her left eyelid completely. Her posterior pharynx was erythematous, but no areas of oropharyngeal fluctuance were noted. Tender, mobile submandibular and cervical lymph nodes were palpable bilaterally, but more prominently on the left. Chest, lung, abdominal, and extremity exams were unremarkable. No rashes were noted. The remainder of the neurologic exam was normal.

Blood cultures, tuberculosis testing, and HIV antibody testing were negative. Antinuclear antibody and extractable nuclear antigen profiles were within normal limits. Serum angiotensin-converting enzyme (ACE) levels were twice the upper limit of normal. Rheumatoid factor was not checked on initial presentation. Computed tomography (CT) of the chest showed hilar lymphadenopathy with bibasilar interstitial lung disease. CT of the neck soft tissues demonstrated enlarged lymph nodes, the largest of which measured 3.2 × 1.2 cm *(Figure 1)*. Esophagogastroduodenoscopy showed pharyngeal edema but no evidence of obstruction. The patient's ophthalmologic exam was notable for impaired corneal sensation, punctate epithelial erosions in both eyes, and no overt flare or leukocytes seen on slit lamp exam. Fundi were normal in appearance. Fine-needle aspiration of a left preauricular lymph node revealed multinucleated giant cells and noncaseating granulomas consistent with granulomatous lymphangitis *(Figure 2)*. No bacteria or viruses were grown on culture of the biopsied lymph node. The patient was given a diagnosis of Heerfordt-Waldenström syndrome, or uveoparotid fever, which is a rare initial presentation of sarcoidosis.

DISCUSSION

Given the patient's acute onset of unilateral facial droop, the differential diagnosis includes vascular, infectious, granulomatous, neoplastic, and autoimmune causes of cranial nerve VII injury *(Table 1)*. The principal vascular causes of unilateral facial weakness are ischemic stroke and intraparenchymal hemorrhage involving the cranial nerve VII nucleus. Infectious causes of unilateral facial palsy include herpes simplex virus–associated Bell's palsy, Lyme disease, HIV, and varicella zoster virus–associated Ramsay-Hunt syndrome. Tuberculosis, orofacial granulomatosis, and sarcoidosis are granulomatous diseases that cause unilateral facial palsy. In tuberculosis, the mastoid, middle ear, or petrous bone is often involved (1, 2). In sarcoidosis, neoplasms such as lymphoma and adenocarcinoma, and autoimmune etiologies such as systemic lupus erythematosus and Sjögren's disease, the paralysis is thought to be related to inflammation or compression of cranial nerve VII.

Sarcoidosis is a systemic disease characterized by granuloma formation. Typical clinical findings include cough, dyspnea, chest pain, fever, weight loss, arthralgias, and

From the Department of Medicine, Tulane University School of Medicine, New Orleans, Louisiana.

Corresponding author: A. Domnica Fotino, MD, MPH, Assistant Professor, Department of General Internal Medicine and Geriatrics, Tulane University Health Sciences Center, 1430 Tulane Avenue, SL-16, New Orleans, LA 70112 (e-mail: dfotino@tulane.edu).

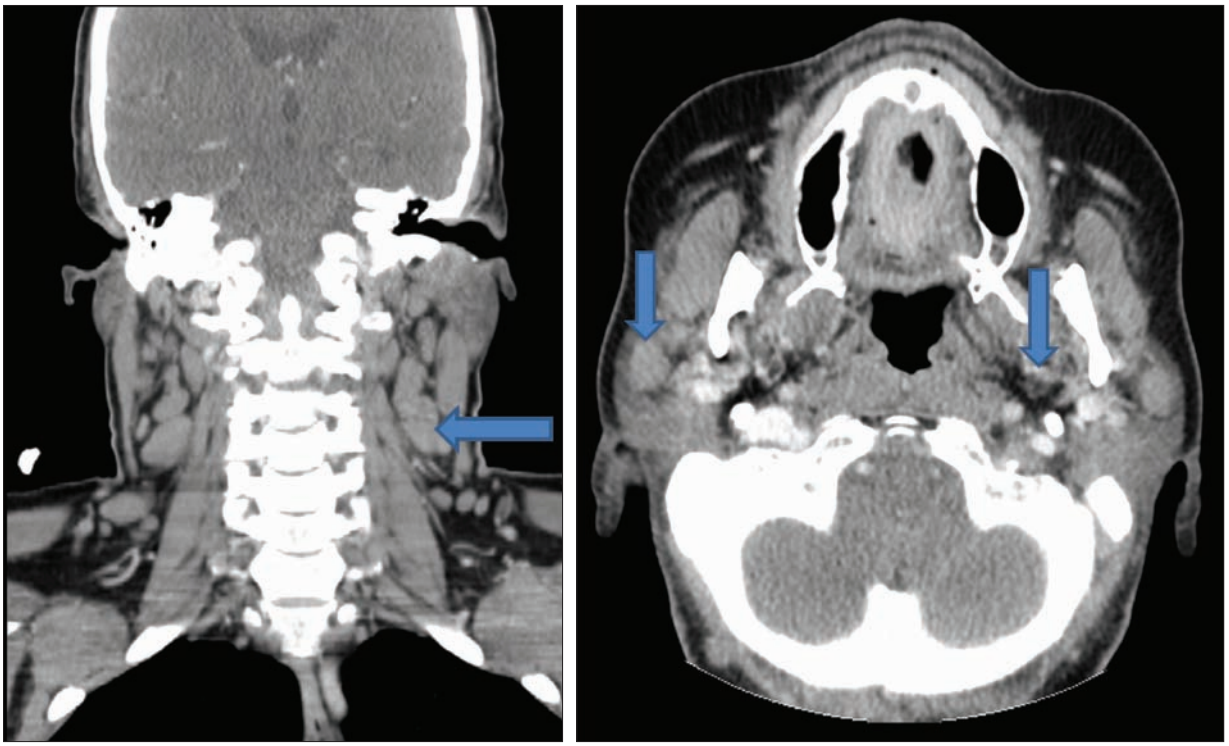

Figure 1. CT of the neck soft tissues demonstrating prominent cervical lymphadenopathy.

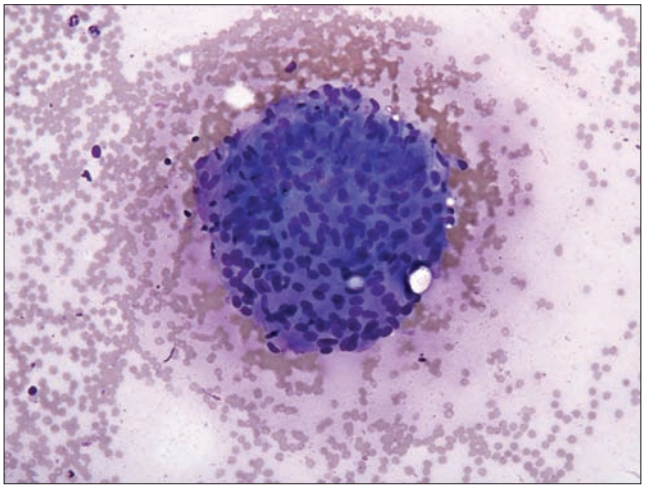

Figure 2. Preauricular lymph node biopsy showing a noncaseating granuloma.

Table 1. Etiologies of facial nerve palsy

- Ischemic stroke of the pons
- Intraparenchymal hemorrhage of the pons
- Herpes simplex virus–associated Bell's palsy
- Human immunodeficiency virus
- Lyme disease
- Varicella zoster virus–associated Ramsay-Hunt syndrome
- Sarcoidosis
- Tuberculosis
- Adenocarcinoma
- Lymphoma
- Sjögren's syndrome
- Systemic lupus erythematosus

erythema nodosum. Only 5% of sarcoidosis cases have neurologic involvement, with the most common manifestation being facial nerve palsy (3–6). Heerfordt-Waldenström syndrome, also called uveoparotid fever, is characterized by the presence of parotid gland enlargement, facial nerve palsy, anterior uveitis, and fever (7–9). Dr. Christian Heerfordt first described this constellation of symptoms in 1909 (10). Dr. Jan Waldenström made the observation that this syndrome was associated with sarcoidosis in 1937 (11). Isolated case reports have described Heerfordt-Waldenström syndrome, but the exact prevalence is not known.

The parotid gland enlargement and cervical lymphadenopathy observed in this patient are typical of Heerfordt-Waldenström syndrome and are caused by a granulomatous inflammatory reaction. The facial nerve palsy is associated with epineural granulomas and perineural inflammatory infiltrates of cranial nerve VII (12). Anterior uveitis classically presents with infected conjunctiva, blurred vision, or eye pain (13). This patient's blurred vision and eye pain were thought to be related to uveitis. The patient had already received high-dose steroids for 3 days at the time of the full ophthalmologic exam, which may explain why the slit lamp and funduscopic exams were fairly unremarkable. Salivary gland involvement likely led to the pharyngitis seen on esophagogastroduodenoscopy and the patient's difficulty swallowing.

The diagnosis of Heerfordt-Waldenström syndrome is made by the constellation of symptoms along with an elevated serum ACE level and lymph node biopsy demonstrating noncaseating granulomas. Granulomas in sarcoidosis are known to produce ACE; however, the serum ACE level does not always correlate precisely with disease activity (14, 15). As with other manifestations of sarcoidosis, corticosteroids are the mainstay of treatment. Immunosuppressants such as mycophenolate mofetil, cyclosporine, and infliximab can be utilized in patients whose symptoms are refractory to steroids (16, 17). When complete eye closure is not possible, the patient should use artificial tears while awake, use lubricant eye ointment at night, and cover the affected eye during sleep to prevent exposure keratopathy. If the facial nerve palsy is not treated with steroids, the likelihood of permanent facial paralysis is increased. When the facial paralysis is misdiagnosed as being a simple Bell's palsy, the opportunity to treat the systemic sarcoidosis is missed. This patient had marked improvement of symptoms after 4 days of prednisone therapy. Two months after discharge, her facial nerve palsy, parotitis, fevers, and uveitis had resolved.

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