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CASE REPORT

Bilateral Facial Nerve Paralysis in Wegener's Granulomatosis: A Case Report

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Wegener's granulomatosis (WG) is a systemic inflammatory disease that typically presents with renal, pulmonary, and head and neck manifestations. Seventy-five percent of WG patients present with head and neck signs or symptoms, and approximately 90% of patients with WG will have ENT involvement during the disease course. Unilateral facial nerve paralysis occurs in approximately 5% of WG cases. We present a rare instance of bilateral facial nerve paralysis.

The clinical notes and operative reports of a patient with WG were reviewed. Relevant literature was reviewed. This case reinforces the importance of medical intervention of WG that includes both high dose steroids and an immunosuppressive agent. It also serves as a reminder for the otolaryngologist that facial nerve paralysis should heighten awareness for WG, as earlier intervention can result in better overall outcomes for patients with WG.

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Introduction

Wegener's granulomatosis (WG) is a systemic inflammatory disease that usually presents with renal, pulmonary, and head and neck manifestations, though any part of the body can be affected.[1] We present an unusual case of bilateral facial paralysis in a man who developed systemic WG. Case reports do not require Institutional Review Board (IRB) approval at our institution.

Case Report

A 60-year-old man was evaluated by an outside physician for ear pain and subsequently diagnosed with left otitis media. Oral antibiotics and steroid treatments were initiated by the outside physician; unfortunately, detailed records of dosages, routes, and duration of medications were not available for our review. The patient was unresponsive to treatment, developed paralysis of the left facial nerve two weeks later, and did not improve clinically with additional oral steroids and antiviral medications. Further clinical examination and imaging revealed fluid in the mastoid air space and the middle ear; as a result, myringotomy, aspiration, and ear tube placement were performed. The patient subsequently developed right facial paralysis two weeks later and was started on unspecified oral antibiotics and steroids due to similar findings in the right ear. He developed respiratory symptoms of cough and dyspnea, and was started on intravenous antibiotics. Imaging studies revealed multiple cavitary pulmonary lesions, laboratory testing revealed C-ANCA positivity, but no definite diagnosis was made at that time. Because of clinical decompensation worrisome for meningitis, including worsening fevers, deteriorating mental status, new-onset deafness, and respiratory distress, the patient was then transferred to our institution. Clinical examination revealed an obtunded patient with significant nasal crusting and granulation tissue, left-sided ear drainage, right ear tube blockage, and bilateral House-Brackman grade VI facial nerve function. A computerized tomography (CT) scan of the sinuses revealed bilateral pan sinusitis and opacification of the bilateral middle ears and mastoid cavities (Figure 1). Electroneuronography (ENOG) testing of the facial nerve showed no response bilaterally at 30 milliamps (Figure 2), and his audiogram revealed profound mixed hearing loss

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bilaterally. The patient had been medically treated for nearly one month as both an outpatient and inpatient at the outside institution and continued to deteriorate clinically; we therefore decided to intervene surgically for purposes of obtaining biopsy tissue to confirm diagnosis, to debride the nasal cavity, and to explore a middle ear and mastoid due to the facial nerve paralysis. We performed bilateral endoscopic nasal debridement with biopsies, limited left mastoidectomy, and placement of an additional right ear tube. Extensive amounts of granulation tissue were found in the nasal cavities as well as the left mastoid and middle ear spaces (Figure 3). Portions of the nasal septum, left turbinates, and left ossicles had been destroyed, as well as portions of the left ear ossicles and tympanic membrane. Pathology results from nasal biopsies proved consistent with WG.

Postoperatively, the patient was placed on 1 gram Solu-Medrol pulse dose daily for three days, then 80mg prednisone daily. Additionally, he has been maintained on cyclophosphamide at 2mg/kg/day. He has experienced substantial overall clinical improvement as well as an improvement in hearing. Facial nerve function is now grade I on the right and grade IV on the left. Follow-up CT imaging of his middle ears, mastoids, and paranasal sinuses have been stable; specifically, his facial nerve canals appears to be intact on the CT imaging. His rheumatologist and pulmonologist are closely monitoring him for scheduling of steroid and cyclophosphamide tapering.

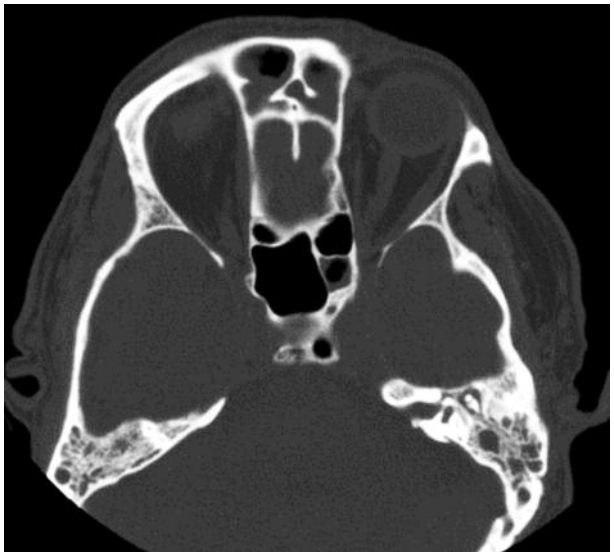

Figure 1. Preoperative CT view showing partial opacification of the inferior frontal sinuses, opacification of the left middle ear, and opacification of the bilateral mastoid cavities..

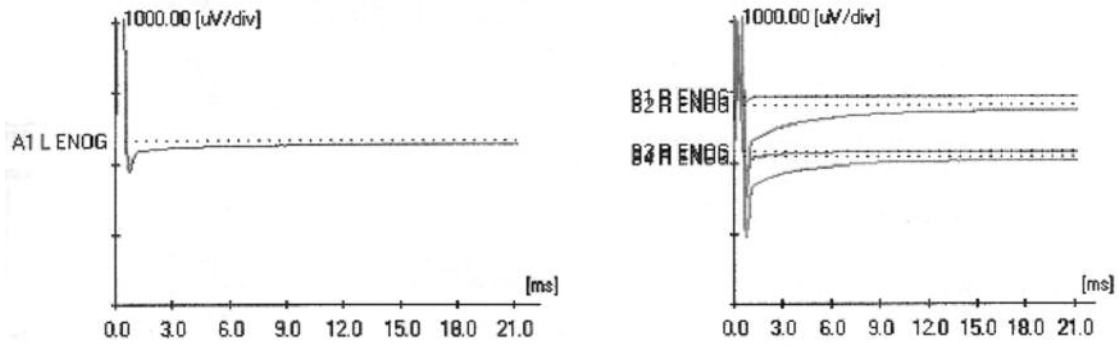

Figure 2. Electroneuronography (ENOG) of the patient, showing no response for the left and right facial nerves, respectively.

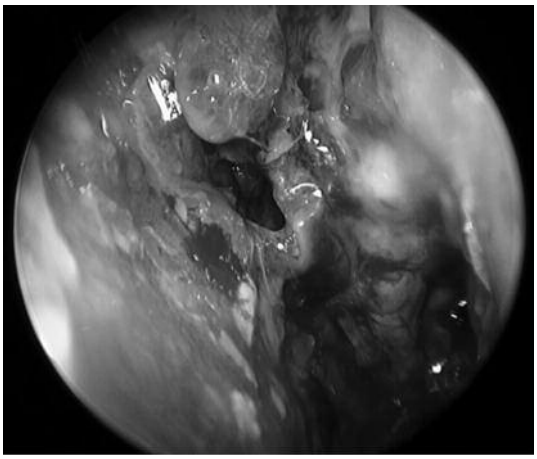

Figure 3. Predebridement view of granulation tissue and secretions in the left nasal cavity in Wegener's granulomatosis.

Discussion

Wegener's granulomatosis is a rare disease, occurring in approximately 3 per 100,000 people, most commonly in Caucasian (>90%) adults, and without gender bias.[1] Signs and symptoms involving structures of the nasal passages, sinuses, trachea, or ears are seen as a presenting symptom in approximately 75% of patients with WG, and approximately 90% of patients with WG will have ENT involvement at some point during the disease course,[2] including nasal obstruction (54% of WG cases), epistaxis (50%), nasal crusting (56%), sinus involvement (33%), new-onset deafness (24%), and hoarseness or stridor due to subglottic stenosis (15%).[3] In fact, ENT manifestations are the most common systems involved by WG,[2] thus placing the otolaryngologist as a central figure in diagnosis and, at times, management of WG. In contrast, unilateral facial paralysis is estimated to occur in approximately 5% of WG cases.[4] A literature review demonstrated only four previous reported cases of bilateral facial nerve paralysis in WG.[4,5,6,7]

This case reinforces the importance of primary medical intervention with both high dose steroids and an immunosuppressive agent, as steroids alone are generally not sufficient in treatment of systemic WG.[8] Importantly, others have demonstrated the ability to achieve at least partial facial nerve recovery with early medical intervention.[4] Recommended regimens include cyclophosphamide treatment at 1-2mg/kg/day and glucocorticoids at 1mg/kg/day with tapering doses.[1] Surgical intervention is typically reserved for either biopsy if WG diagnosis is not clear, or for refractory WG head and neck manifestations after the disease is under control medically. Surgical intervention during active disease can lead to worsened inflammation and impaired healing. However, deteriorating clinical situations may warrant surgery acutely.[1] Examples of surgical interventions include endoscopic sinus surgery or myringotomy with ear tube placement for intracranial infection, extension from pansinusitis, and mastoiditis, respectively. Interestingly, the patient has had a persistent grade IV facial nerve paralysis on the left side; this is the side that initially became paralyzed and was also the ipsilateral side to surgical exploration. Intraoperatively, we did not identify any dehiscent areas of the facial nerve. While postoperative CT imaging has shown the remainder of the facial canal to be intact, we cannot completely rule out iatrogenic injury to the nerve. This case also serves as a reminder for the otolaryngologist that facial nerve paralysis should heighten awareness for WG, as earlier intervention can result in better overall outcomes for patients with WG.

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