Multiple Cranial Neuropathies Related to Wegener’s Granulomatosis: An Unusual Presentation

Rajanya S. Petersson, MD, Jan L. Kasperbauer, MD, Colin L. W. Driscoll, MD
Department of Otorhinolaryngology, Mayo Clinic, Rochester, MN

BACKGROUND

Wegener’s granulomatosis is an uncommon, systemic vasculitis characterized by necroizing granulomas of the respiratory tract, focal necroizing vasculitis, and focal necroizing glomerulonephritis.¹,² The prevalence in the United States is estimated to be three cases per 100,000.² In 90% of patients, the first symptoms arise in the head and neck, with the nasal cavity and paranasal sinuses being the most common head and neck sites.³ Other head and neck sites include the ear (usually middle ear), larynx, and oral cavity. The disease usually begins as a localized process, but can progress to a systemic disease that can be fatal due to renal and pulmonary involvement if left untreated.³

Neurologic manifestations occur in 25-30% of patients, and may involve cranial neuropathies.¹ Diagnosis is made on the basis of clinical suspicion, tissue biopsy, laboratory testing (positive cytoplasmic ANCA and anti-PR3 antibodies), and radiography.²,⁴ Treatment is medical, and involves immunosuppressive agents.² We present an unusual case involving the skull base and multiple cranial neuropathies that became apparent after undergoing mastoidectomy for presumed chronic ear disease.

CASE REPORT

A 22 year old female presented with an 8 month history of worsening left ear pain and conductive hearing loss after multiple courses of antibiotics and various otic drops. Microscopic otoscopy revealed opacification of the tympanic membrane. Computed tomography imaging of her temporal bones revealed left middle ear and mastoid opacification (Figure 1). She underwent mastoidectomy with facial recess approach. Intraoperative findings included dense, fibrotic inflammatory tissue in the middle ear, surrounding the ossicles, and throughout the mastoid. Pathological evaluation of the middle ear tissue was consistent only with chronic inflammation; no cholesteatoma was identified. A tympanostomy tube was placed at the time of surgery, and the patient was placed on ciprofloxacin/dexamethasone otic drops.

On postoperative day 2, she was admitted for nausea and vomiting, and it was noted that she had neuropathies of cranial nerves III, V, VII, X, and XII on physical exam. Magnetic resonance imaging revealed enhancement of the left parapharyngeal space extending through foramen ovale to involve the dura (Figure 2). She also had a persistent conductive hearing loss on the left.

Workup for meningitis and other infectious processes were negative. Further workup included positive c-ANCA and anti-PR3, suggesting Wegener’s granulomatosis. Re-review with special staining by the pathologist of the previously submitted ear tissue again did not reveal necroizing granulomas. Tissue biopsy was requested by the rheumatology consulting service from the left parapharynx or dura, but was not performed due to the risks of these procedures.

However, six days after readmission, mucosal ulcerative changes consistent with Wegener’s granulomatosis were noted by the left eustachian tube orifice on nasopharyngoscopy. Biopsy showed necroizing granulomas, consistent with Wegener’s granulomatosis. Initiation of treatment with high dose steroids resolved all cranial neuropathies within days. The patient was started on rituximab as an outpatient.

At last follow-up, seven months after diagnosis, the patient had had her last rituximab infusion five months prior, and had been tapered down to a daily prednisone dose of 8 mg daily. She continues to have left conductive hearing loss and middle ear opacification (Figure 3). As the Wegener’s granulomatosis appears to be in remission, a middle ear exploration will be planned.

METHODS

Case report and review of the literature.

REFERENCES