Case Report: Sequential bilateral otitis media and bilateral facial nerve paralysis as presenting symptoms of Wegener’s Granulomatosis

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ABSTRACT

Wegener’s granulomatosis (WG), a well-known but uncommon immunologic disease, can present in a variety of forms requiring the involvement of multiple medical specialties in the care of these patients. 1,2 The characteristic features are granulomatous lesions of the respiratory tract, glomerulonephritis, and small vessel necrotizing vasculitis. 3 The most common otolaryngologic manifestations are in the nose and paranasal sinuses occurring in 85% of cases with presenting symptoms of congestion, epistaxis, septal perforation, and saddle-nose deformity. 3

While the ear is reportedly involved in approximately 35% of all cases, it is rare to have facial nerve dysfunction. 3 Facial nerve paralysis alone or in combination with hearing loss is reported in about 5% of patients. 4 A case report is presented of a 35-year-old male with sequential bilateral otitis media and progressive mixed hearing loss, tinnitus, otalgia, otorhea, and, later, bilateral facial nerve paralysis. A review of the literature demonstrates only six cases of bilateral facial palsy from WG worldwide with this case representing the first in the United States. 1,3,4

INTRODUCTION

While Wegener’s granulomatosis bears the name of German pathologist Friedrich Wegener who first described it in 1931, it was later defined in 1969 by Peter C. McIndoe, a Scottish otolaryngologist. The destructive involvement of the nose and paranasal sinuses, which is what McBride reported, is the most common manifestation, present in up to 85% of all cases. 3 The necrotizing vasculitic syndrome is thought to be caused by an autoimmune attack on the small to medium-sized blood vessels by anti-neutrophil cytoplasmic antibodies (ANCA). The inflammation that results in the destruction of granulomatous tissue. 3,4,5

The diagnosis of WG can be difficult to obtain so in 1990, the American College of Rheumatology established four criteria to aid with the identification. These are: non-specific sinusitis (red cell casts greater than 5 red blood cells per high power field), abnormal chest radiograph findings, oral ulcers or nasal discharge, and granulomatous inflammation on biopsy. According to the publication, the presence of 2 or more of the criteria was associated with a sensitivity of 88.2% and a specificity of 92.0%. The criteria’s purpose was to distinguish WG from other types of vasculitis to guide treatment. 4-10 Another important diagnostic finding, with a reported sensitivity as high as 97%, is the presence of elevated levels of ANCA, most specifically the cytoplasmic pattern (c-ANCA). 2

The ear is rarely the presenting symptom in WG. In these cases, the differential diagnosis includes chronic infections such as tuberculosis, sarcoidosis, and syphilis. 3-9,10 The otologic manifestations have been divided into five categories, listed in decreasing prevalence: serous otitis media, sensorineural hearing loss, chronic otitis media, vertigo, and facial palsy. 3-4,9 To date, 26 cases of unilateral facial paralysis and only 6 cases of bilateral facial paralysis are reported worldwide in the literature. 1,3,4

ABSTRACT (cont’d)

It is well recognized that Wegener’s granulomatosis is an intractable vasculitis that can be life threatening when end-organ damage occurs. Symptoms most commonly affect the nose, lungs, and kidneys. We present a case study of a 35-year-old male who presented with sequential bilateral otitis media, progressive mixed hearing loss, tinnitus, otalgia, otorhea, and, later, bilateral facial nerve paralysis despite aggressive antibiotic intervention. The accompanying review of the literature demonstrates only six other patients worldwide with these presenting symptoms. Treatment options and outcome will be discussed.

CASE REPORT

A 35-year-old male merchant marine with extensive overseas travel and past medical history significant for diverticulitis began complaining of right otalgia. Over the next 3 months with only minimal improvements from multiple courses of topical and oral steroids, his symptoms worsened to include brown ototrauma, ipsilateral hearing loss, and occasional nonpulsatile tinnitus.

Initially, he presented to the Emergency Department for continued pain located posterosuperiorly to his right ear. His exam was only remarkable for a thickened and intact right tympanic membrane and he was given two otic drops, one with benzocaine and the other with hydrocortisone. The following day, he developed sudden ipsilateral facial nerve paralysis (Figure 3). After an initial hospitalization, the facial paralysis was believed to be due to a medication reaction. However, he was admitted to the hospital for imaging and 7 days of intravenous antibiotics including ciprofloxacin.

The clinical picture was suggestive of otitis media and was further supported by a CT scan, which demonstrated findings compatible with myringitis (Figure 1). A wide-field myringotomy was performed but failed to relieve his symptoms. The patient also received a CT chest as a pulmonary nodule was noted on a prior outpatient chest x-ray. It demonstrated an 8-mm non-calcified right upper lobe nodule with a probable small area of central cavitation. A PPD was placed but read as negative.

Upon discharge, the patient presented to have right facial paralysis (6/6 on the House-Brackmann scale) with a normal Bell’s phenomenon. The microscopic otologic exam was remarkable for purulent drainage and eustachian tube stenosis. The right external auditory canal, a sclerotic tympanic membrane, and a whitish mass in the middle ear space. An audiogram demonstrated severe to profound mixed hearing loss in the right, mostly of sensorineural nature with a 40-decibel air-bone gap in the low frequencies. He had a type B tympanogram with 20% speech discrimination. The left ear was within normal limits.

After a few days of no improvement, a right-sided tympanomastoidectomy debridement was performed. The pale whitish mass noted in the middle ear space was removed and cultures were sent. The pathology demonstrated severe to profound mixed hearing loss with a 40-decibel air-bone gap in the low frequencies. The left ear was within normal limits.

The patient’s pulmonary function eventually responded to the antibiotics, and he was able to be successfully extubated. He was discharged from the hospital on day 53. Upon discharge, his bilateral facial nerve paralysis and hearing loss remained unchanged; however, further improvement is expected with outpatient neurophysiological therapy.

Figure 1: Axial CT temporal bones images from second hospitalization demonstrates left facial nerve paralysis and bilateral ear space and mastoid air cells (Figure 3). A repeat PPD was negative along with a quantiferon assay.

Figure 2: High power (40X) view of the necrotic amorphous middle ear mass shows ghost nuclei in a fibrotic stroma. The fibrous tissue focally extends into the right petrous apex without evidence of bony erosion. The left side shows well-intact normal ear cells.

Figure 3: Axial CT temporal bones images from second hospitalization demonstrates opacification of the mastoid air cells and middle ear space on the left (right). The right ear shows progressive changes from the OME (middle ear opacity with mucosal creases (left).