Case Report

Sequential Bilateral Otitis Media and Bilateral Facial Nerve Paralysis as Presenting Symptoms of Wegener's Granulomatosis

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Received 28 March 2013; Accepted 6 May 2013

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Abstract Wegener's granulomatosis is an incurable 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, otorrhea, 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.

Keywords Wegener's granulomatosis; facial paralysis; facial nerve dysfunction

1. Introduction

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 [3,7]. 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 [1].

While the ear is reportedly involved in approximately 35% of all cases, it is rare to have facial nerve dysfunction [1]. Facial nerve paralysis alone or in combination with hearing loss is reported in about 5% of patients [9]. A case report is presented of a 35-year-old male with sequential bilateral otitis media, progressive mixed hearing loss, tinnitus, otalgia, otorrhea, 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 [1,2,4,6,7,8].

2. 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 antibiotics, the patient symptoms worsened to include brown otorrhea, 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 but intact right tympanic membrane and he was given two otologic drops, one with benzocaine and the other with hydrocortisone. The following day, he developed sudden ipsilateral facial nerve paralysis (5/6 on the House-Brackmann scale). Initially, the paralysis was believed to be due to a medication reaction. However, he was admitted to the hospital for imaging and 7 days of treatment, including IV antibiotics, steroids, and antivirals.

The clinical picture was suggestive of otitis media and was further supported by a CT scan, which demonstrated findings consistent with right-sided otomastoiditis (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 was noted 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 erythema in 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 on 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.

Figure 1: Axial head CT demonstrates right-sided otomastoiditis that extends into the right petrous apex without evidence of bony erosion. The left side shows well-aerated mastoid air cells.

Figure 2: High power (40X) view of the necrotic amorphous middle ear mass shows ghost nuclei in a fibrotic stroma. The fibrous tissue focally lined by pseudostratified epithelium, with marked fibrosis, chronic inflammation, and areas of necrosis, is suggestive of chronic otitis media.

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. On pathology, the mass measured $2.1 \times 0.4 \times 0.4$ cm and was noted to have findings most consistent with chronic otitis media (Figure 2). The mass was negative for fungal forms and acid fast bacilli, and there was no evidence of sarcoidosis.

The recovery was uneventful until approximately 2 weeks postoperatively when the patient began complaining

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 postoperative changes from the CWU mastoidectomy with osseous erosion (left).

of left otalgia and mild ipsilateral hearing loss. Physical exam was unchanged despite the new complaints. After failing another course of antibiotics, a left myringotomy was performed when a retracted, thickened tympanic membrane was noted on exam, and he was admitted to the hospital for a comprehensive work-up. A chest X-ray on this admission showed the pulmonary nodule was now 3 cm in diameter so a CT-guided biopsy on hospital day #3 was performed and showed areas of necrosis, fibrosis, and benign alveolar parenchyma with reactive atypical pneumocytes.

Unexpectedly on hospital day #6, the patient developed complete left facial nerve paralysis. Shortly after this finding was noted, he began complaining of bilateral joint pain and had one isolated episode of hemoptysis. Repeat CT temporal bones demonstrated expected postoperative changes on the right side with ossicular chain disruption. On the left, opacification without osseous erosion was noted in the middle ear space and mastoid air cells (Figure 3). A repeat PPD was negative along with a quantiferon assay.

On this admission, serum leukocytosis was 13,500 and ESR 76. When all the hematologic, immunologic, rheumatologic, and metabolic tests returned normal except for 2+ C-ANCA positivity, the diagnosis of Wegener's granulomatosis was made and treatment was begun with corticosteroids and cyclophosphamide. Unfortunately, the patient's respiratory status declined rapidly and required prolonged intubation with a week of extracorporeal membrane oxygenation (ECMO). In addition to the medications, the patient also required plasmapheresis for the treatment of WG.

The patient's pulmonary function eventually responded, 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 neuropsychological therapy.

3. Discussion

While Wegener's granulomatosis bares the name of German pathologist Friedrich Wegener, it was first described in 1897 by Peter McBride, 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 [1]. 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 ensues causes the formation of granulomas [1,5,10].

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 abnormal urinary sediment (red cell casts or greater than five red blood cells per high power field), abnormal chest radiography findings, oral ulcers or nasal discharge, and granulomatous inflammation on biopsy. According to the publication, the presence of two 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. Another important diagnostic finding, with a reported sensitivity as high as 97% depending on the disease activity, is the presence of elevated levels of ANCA, most specifically the cytoplasmic pattern (c-ANCA) [3].

The ear is rarely the presenting symptom in WG. In these cases, the differential diagnosis includes chronic infections such as tuberculosis, sarcoidosis, and syphilis [5,9,10]. 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 [1,5,9]. To date, 26 cases of unilateral facial paralysis, and only 6 cases of bilateral facial paralysis are reported worldwide in the literature [1,2,4,6,7,8].

The cause of the otologic symptoms is unclear. It is believed that the involvement of the nose and paranasal sinuses leads to nasopharyngeal inflammation, causing Eustachian tube dysfunction [5,9,10]. As for the hearing loss, two theories exist. The vasa nervorum and the cochlear vessels could be affected by vasculitis, or there could be an immune-mediated labyrinthitis caused by immune complexes depositing on the labyrinth. Therefore, the facial nerve paralysis would be caused by the vasa nervorum vasculitis or compression of the nerve in the middle ear space [1,5,9,10].

The standard treatment of WG is the combined use of steroid and immunosuppressive drugs. In the unusual presentations like the one described above, early diagnosis is extremely difficult to obtain, and proper treatment is frequently delayed. Operative management often proves to be

futile [1,5]. The otitis media responds poorly to antibiotics and pressure-equalizing (PE) tubes are usually placed for the drainage of the effusion [8]. Commonly, mastoidectomy only reveals the obliteration of the air cells by granulation tissue. In fact, when facial palsy is noted, nerve decompression is rarely useful and may even aggravate the problem [1,10].

With proper treatment, remission rates of 70–90% have been reported, especially in the absence of renal involvement [1,7]. The major determinant in survival is the extent of the progressive renal disease such that if left untreated, 82% of patients die within one year [1,8]. In the few cases of facial nerve paralysis, most patients experienced only mild to moderate improvement of their facial function [1,2,4,6,7,9]. Due to the short follow-up and rarity of the presentation, a generalization cannot be made regarding expectant recovery.

4. Conclusion

The early diagnosis of WG and initiation of proper treatment is essential in patient survival and prevention of progression of the disease. Prior to operative management or placement of PE tubes, it is recommended to examine the nasopharynx as part of the initial assessment. We stress that in cases of refractory or new-onset otitis media with effusion in an adult, even in the absence of significant rhinologic findings, to consider the diagnosis of WG.

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