An Unusual Case of Otitis Media

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Abstract
This case report of bilateral otitis media in a 39-year-old woman secondary to Wegener’s Granulomatosis highlights, for the audiologist, the importance of being aware of some of the less common etiologies of middle ear disease. Bilateral otitis media that resists usual forms of medical treatment may represent one of the earliest signs of Wegener’s Granulomatosis, a potentially life-threatening disease. According to recent literature, involvement of the ears, nose, and throat can in many instances be the only early manifestation of the disease. It is important that audiologists be aware of the early signs in order to make appropriate referral for treatment, particularly since they are usually among the first to see these patients. Early diagnosis and referral is critical since the mean survival of untreated WG is five months, with 82% of patients dying within one year, and more than 90% dying within two years. The disease involves a systemic vasculitis that may involve any organ system; however, pulmonary or renal disease appear to be among the later signs, while both middle ear and upper respiratory involvement are frequently among the earliest.

Key Words: Autoimmune disease, cytoxan, methotrexate, otitis media, prednisone, systemic vasculitis, Wegener’s granulomatosis

Abbreviations: WG = Wegener’s granulomatosis

Sumario
Este reporte de caso de una otitis media bilateral en una mujer de 39 años de edad, secundario a una Granulomatosis de Wegener, destaca para el audiólogo la importancia de ser consciente de algunas de las etiologías menos frecuentes en la enfermedad del oído medio. Una otitis media bilateral que se resiste a las formas habituales de tratamiento médico, puede ser uno de los signos más tempranos de la Granulomatosis de Wegener, una enfermedad potencialmente mortal. De acuerdo a la literatura reciente, el compromiso de los oídos, la nariz y la garganta, puede a menudo ser la única manifestación temprana de la enfermedad. Es importante que los audiólogos conozcan estos signos tempranos, para hacer una referencia apropiada para tratamiento, particularmente por ser quienes primero ven al paciente. Un diagnóstico y una referencia temprana son críticos pues la supervivencia media de una WG no tratada es de cinco meses, con muerte del 82% de los pacientes dentro de un año y de más de 90% a los dos años. Esta enfermedad produce una vasculitis sistémica que puede involucrar cualquier sistema de órganos; sin embargo, la enfermedad pulmonar o renal parece ser un signo tardío, mientras que el compromiso del oído medio y del tracto respiratorio superior está frecuentemente entre los signos más tempranos.

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Technical information from this paper was included in a Research Podium presentation at the American Academy of Audiology 15th Annual Convention and Exposition, San Antonio, TX, Apr. 2–5, 2003.
The sudden onset of otitis media that resists usual medical treatment may signal a serious progressive disease. The following case study illustrates such progression in a patient ultimately diagnosed with Wegener’s granulomatosis, a potentially life-threatening disease.

**CASE STUDY**

A female patient, age 39, in previous good health, presented a moderate bilateral conductive hearing loss secondary to sudden onset of bilateral otitis media, accompanied by painful sinusitis. Middle ear pain began suddenly the day after she had helped a friend move out of a very dusty apartment. The day of dust exposure apparently concluded with an allergic reaction that manifested itself with sinus drainage and acute otitis media with middle ear pain. By the following day, the middle ear pain was intense. Severe bilateral otitis media was subsequently diagnosed by an ear, nose, and throat specialist. Treatment began immediately, but both the otitis media and severe sinus pressure persisted without remission well beyond expectations in spite of typical medical treatment and did not subside to any degree that would indicate recovery.

Figure 1 shows the patient’s pure-tone audiogram at the most advanced stage of the disease, approximately four months after the onset of initial symptoms. Unmasked air-conduction thresholds were measured via insert receivers. There was a moderate conductive loss of 30–40 dB in both ears. Tympanograms were flat. Word-recognition scores were unimpaired. Over the next several months, other signs and symptoms emerged including persistent dry cough accompanied by a large granulomatous cyst of the right lung requiring surgery, painful symptoms of arthritis in the hands and feet, chronic fatigue, persistent low-grade fever, night sweats, and unexplained weight loss, when in fact this patient was described as a previously healthy person. Over the next six months, she was seen by several medical specialists but with no suggestion of a diagnosis. However, the symptoms continued to worsen.

By the conclusion of a six-month period of illness, she was jointly diagnosed by an immunologist and rheumatologist as possessing Wegener's granulomatosis (WG), and treatment began immediately. At that time, treating physicians noted that, if the disease had gone without a correct diagnosis, the patient would in all probability have died within several months. However, with aggressive treatment, the patient has now been in remission for five years.

**Figure 1.** Pure-tone audiograms four months after onset of the disease at its most advanced stage.
CLINICAL MANIFESTATIONS OF WG

Persistent adult onset bilateral otitis media that resists usual forms of medical treatment may represent the earliest symptom of WG. This has been recognized as early as 1980 by McCaffrey et al (1980) and in 1986 by Coloday et al (1986) and has been confirmed more recently by Banerjee et al (2001) and Takagi et al (2002), among many others. In fact, according to Banerjee et al (2001), involvement of the ears, nose, or throat may at times be the only early manifestation of the disease. Prompt recognition and treatment of the underlying disease process is important since the mean survival of untreated WG is five months, with 82% of patients dying within one year, and more than 90% dying within two years (National Institute of Allergy and Infectious Diseases [NIAID], 2002; Wegener’s Granulomatosis Association, 2002).

According to Macias et al (1998), WG is a systemic vasculitis that may involve any organ system. However, according to Lebovics (2000), it appears to affect primarily the head and neck in its early stages. It is frequently described as an autoimmune disease (Murray, 2000), but an etiology has not yet been identified. According to Banerjee et al (2001), WG is most frequently characterized by granulomatous lesions of the sinuses, nasal passages, the middle ear, and lungs, and it belongs to one of the seven forms of vasculitis classified by the American College of Rheumatology.

Murray (2000) summarizes several studies involving the characteristics of WG, and presents commonly regarded descriptors and symptoms. Those include a description of WG as an autoimmune multisystem disease whose cause remains unknown. However, according to his summary, it is currently felt that WG may represent an antigen triggered immunologic reaction to environmental elements, but what those are also remains unknown. Interestingly enough, it also resembles an infectious process, although no viral, bacterial, or other causative agent has been identified.

PREVALENCE OF PRIMARY SIGNS AND SYMPTOMS OF WG

The prevalence of primary signs and symptoms of WG is summarized by Murray (2000), and the Johns Hopkins University Vasculitis Center (Helman, 2002). These involve (1) disease of the ear, nose, and throat (95%); (2) symptoms of a persistent “cold” or “sinusitis” accompanied by otitis media that fails to respond to traditional treatment (about 90%); (3) granulomatous or other similar lesions of the lungs, with chronic cough (85%); (4) kidney malfunction (75%); (5) arthritic symptoms of the joints including hands, knees, ankles, and feet (70%); and (6) double vision or difficulty focusing the eyes (50%).

DEMOGRAPHICS OF WG

A review of the literature reveals the following (1) Age: WG can occur at any age but has its peak in the 40s and 50s; the age range of patients is currently found to range from ages 5–91 years; 85% percent of patients are above age 19 years; the average age
of patients with WG is 41 years; (2) Race: 97% are Caucasian, 2% are African American, and 1% are from other races; (3) Sex: WG affects males and females nearly equally, although females demonstrate a slightly higher prevalence (Carruthers et al, 1996; Murray 2000; Banerjee et al, 2001; Helman, 2002; NIAID, 2002).

**TREATMENT OF WG**

Until 20 years ago, most persons with WG died, since treatment alternatives for the symptoms of the disease did not generally exist or were not effective. Current treatment involves attempts at suppressing the patient’s immune system in order to eliminate its aggressiveness in attacking the patient’s vital systems. A prescribed regimen of three drugs is generally the course of treatment today. They include methotrexate, prednisone, and cyclophosphamide as the disease and its symptoms necessitate (Langford, 2003). These are powerful medications, and the combined affects are similar to those of chemotherapy, including hair loss, and others. However, at the present time, those drugs alone or in combination are the regimen of choice for many physicians, primarily hematologists and immunologists who diagnose and treat persons with the disease.

**SUMMARY**

In view of many of the early signs and symptoms of the disease, audiologists may be the first to see patients with WG. It is, therefore, imperative that they become familiar with the signs and symptoms that may represent early signs of this dangerous disease so that proper referral can be made. In fact, according to Takagi et al (2002), the possibility of WG should typically be included in the differential diagnosis in cases of atypical inflammatory states of the ear. The reason for the urgency of accurate referral, as stated earlier, is that untreated Wegener’s granulomatosis is generally fatal. But if the disease is discovered and treated appropriately and in a timely manner, patients now have an excellent chance for remission. However, according to the Wegener’s Granulomatosis Support Group (2000), it has been found to take up to 5–15 months to make a diagnosis of WG, primarily because many persons in positions to make the diagnosis may not be aware of the symptoms of the disease. Since a person with undiagnosed WG can die within five months of the onset of the disease, it is critical that a diagnosis be made as quickly as possible. Audiologists are frequently in positions in which the early signs and symptoms of the disease can be recognized and appropriate referral can be made.

**REFERENCES**


