



Case Report

Otitis media triggered by Wegener's granulomatosis: A case report and review of the literature

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Article info

Article History:

Received: 08 June 2015

Accepted: 10 Oct 2015

ePublished: 30 Nov 2015

Keywords:

Otitis Media,
Systemic Vasculitis,
Wegener's
Granulomatosis

Abstract

Introduction: This case report of unilateral otitis media in a 19-year-old female secondary to Wegener's granulomatosis (WG) highlights, for the audiologist, the importance of being aware of some of the less common etiologies of middle ear disease. Unilateral otitis media that resists usual forms of medical treatment may represent one of the earliest signs of WG, a potentially life-threatening disease.

Case Report: A female patient, age 19, in previous good health, presented a moderate unilateral conductive hearing loss secondary to the sudden onset of unilateral otitis media, accompanied by painful sinusitis. However, the symptoms continued to worsen. By the conclusion of a 6-month period of illness, she was jointly diagnosed by an immunologist and rheumatologist as possessing WG, and treatment began immediately. With aggressive treatment, the patient has now been in remission for 5 years.

Conclusion: It is important that audiologists be aware of the early signs to make an appropriate referral for treatment, particularly since they are usually among first to see these patients. Early diagnosis and referral are critical.

Citation: Ghavidel A. Otitis media triggered by Wegener's granulomatosis: A case report and review of the literature. *J Anal Res Clin Med* 2015; 3(4): 261-3. Doi: 10.15171/jarcm.2015.040

Introduction

Unilateral otitis media that resists usual forms of medical treatment may represent one of the earliest signs of Wegener's granulomatosis (WG), a potentially life-threatening disease. According to recent literature, the 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 an appropriate referral for treatment, particularly since they are usually among first to see these patients. Early diagnosis and referral are critical since the mean survival of untreated WG is 5 months, with 82% of patients dying within 1 year, and more than 90% dying within 2 years. The disease involves a systemic vasculitis that may involve any organ system: however,

pulmonary or renal disease appears to be among the later signs, while both middle ear and upper respiratory involvement are frequently among earliest involved tracts.

Case Report

A female patient, age 19, in previous good health, presented a moderate unilateral conductive hearing loss secondary to sudden onset of unilateral otitis media, accompanied by painful sinusitis. An earache 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 the earache. By the following day, the ear pain was intense. Severe unilateral otitis media was

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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.

Over the next several months, other signs and symptoms emerged including persistent dry cough, 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 6 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 6-month period of illness, she was jointly diagnosed by an immunologist and rheumatologist as possessing WG, and treatment began immediately. With aggressive treatment, the patient has now been in remission for 5 years.

Discussion

Persistent adult onset unilateral otitis media that resists usual forms of medical treatment may represent the earliest symptom of WG. This has been recognized as early as by McCaffrey et al.¹ and has been confirmed more recently by Banerjee et al.² among many others. In fact, according to Banerjee et al., involvement of the ears, nose, or throat may at times be the only early manifestation of the disease.² Prompt recognition and treatment of the underlying disease process is important since the mean survival of untreated WG is 5 months, with 82% of patients dying within 1 year, and more than 90% dying within 2 years.

According to Macias et al.,³ WG is a systemic vasculitis that may involve any organ system. However, according to Lebovics⁴ it appears to affect primarily the head and neck in its early stages. It is frequently described as an autoimmune disease, but an etiology has not yet been identified. According to Banerjee et al.,² WG is most frequent 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 summarizes several studies involving the characteristics of WG, and presents commonly regarded descriptors and symptoms.⁵ 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.^{6,7}

The prevalence of primary signs and symptoms of WG is summarized by the Johns Hopkins University Vasculitis Center. 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%).^{5,8}

A review of the literature reveals the following important elements signs and symptoms of WG including: (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 to 91 years; 85% 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. 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.^{9,10}

A prescribed regimen of three drugs is generally the course of treatment today. They include methotrexate, prednisone, and cytoxan (cyclophosphamide) as the disease and its symptoms necessitate. These are powerful medications, and the combined effects 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.^{11,12}

Conclusion

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, 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 WG is generally fatal. However, 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 WG Support Group,⁵ and 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 5 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.

Conflict of Interests

Authors have no conflict of interest.

Acknowledgments

The author would like to thank Liver and Gastrointestinal Diseases Research Centre and Imam Reza Hospital, Tabriz University of Medical Sciences for preparing research materials.

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