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Case Report

From Muffled Hearing to Facial Nerve Palsy: A Case Report and Literature Review of Middle and Inner Ear Granulomatosis with Polyangiitis

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Abstract

This case report discusses a patient who initially presented with severe otitis media and hearing loss, which later led to a diagnosis of granulomatosis with polyangiitis (GPA). Despite only affecting the patient's ear and facial nerve, GPA is an autoimmune disease that can impact several organs in the body. Laboratory tests revealed elevated levels of PR3 and positive C-ANCA. The patient received treatment with a combination of corticosteroids and immunosuppressants, resulting in the resolution of otitis media and facial nerve palsy, while preserving their hearing. This case emphasizes the significance of considering GPA as a possible diagnosis in patients with sudden hearing loss and otitis media who may also experience facial nerve palsy, particularly in those with high C-ANCA levels.

Keywords: Granulomatosis with Polyangiitis; Cytoplasmic Anti-neutrophil Cytoplasmic Antibodies; Otitis Media; Hearing Loss

Introduction

Granulomatosis with polyangiitis (GPA) is an uncommon autoimmune disorder that affects multiple organ systems. It was initially described by Friedrich Wegener in 1936 and was referred to as Wegener's granulomatosis for many years. However, in 2012, the 2012 International Chapel Hill Consensus Conference renamed the disease as granulomatosis with polyangiitis, replacing the term Wegener's granulomatosis [2,10].

Granulomatosis with polyangiitis is characterized by vasculitis of the small- and medium-sized blood vessels, leading to the development of necrotizing granulomas in different body tissues. This autoimmune disorder primarily affects the respiratory system; however, it can also involve other organs such as the kidneys, skin, eyes, nose, paranasal sinuses, middle and inner ear, and the subglottic region of the larynx [2-10].

Granulomatosis with polyangiitis can present with several systemic symptoms such as fatigue, low-grade fever lasting for weeks or months, weight loss, joint and muscle pain, as well as stiffness. Furthermore, this autoimmune disorder can also cause diverse skin manifestations, including rashes, ulcers, and nodules. GPA may also affect the kidneys, leading to proteinuria, hematuria, necrotizing glomerulonephritis, and ultimately, renal failure [9].

The upper respiratory tract is the most affected area in the otolaryngological manifestations of granulomatosis with polyangiitis (GPA). The nose, sinuses, and ears are frequently involved. Symptoms related to the nose may include chronic or recurrent sinusitis (85%), nasal congestion, crusting and bleeding, nasal deformities, and saddle nose deformity. Involvement of the ears (approximately 35% of the cases) can cause conductive or sensorineural hearing loss, tinnitus, and vertigo [2].

Throat symptoms, such as sore throat, hoarseness, and difficulty swallowing, are less common than upper respiratory tract symptoms in granulomatosis with polyangiitis and occur in approximately 5% of cases. Facial nerve palsy is also reported in a minority of cases. However, in severe cases, GPA can cause destruction of the nasal septum, perforation of the palate, and subglottic stenosis. Involvement of the lower respiratory tract may lead to cough, hemoptysis, and shortness of breath [5,6].

The American College of Rheumatology (ACR) has established diagnostic criteria for granulomatosis with polyangiitis (GPA). According to the ACR, a patient must meet at least two of the following criteria to confirm the diagnosis of GPA: [3].

- **Nasal or oral inflammation:** Development of ulcers or crusts in the nose or mouth, with or without pain.
- **Abnormal chest X-ray:** Presence of nodules, infiltrates, or cavities in the lungs.
- **Urinary abnormalities:** Presence of red blood cells or casts in the urine or other evidence of kidney disease, such as proteinuria.
- **Biopsy showing granulomatous inflammation:** Biopsy of tissue showing inflammation characterized by clusters of immune cells known as granulomas.
- **Positive ANCA (anti-neutrophil cytoplasmic antibody) test:** Presence of ANCA, a type of antibody that attacks neutrophils, in the blood.

It is important to emphasize that a definitive diagnosis of GPA requires careful evaluation by a rheumatologist or other specialist [3].

Although granulomatosis with polyangiitis typically affects the upper and lower respiratory tracts and the kidneys, it can rarely involve the middle ear. Biopsies of the middle ear are not routinely performed for GPA diagnosis due to the rarity of middle ear involvement. However, in cases where a biopsy of the middle ear is obtained but is insufficient for diagnosis, other diagnostic tools may be utilized to confirm or exclude GPA. These include blood tests for ANCA and imaging studies such as CT scans of the chest and sinuses, which can identify other sites of disease involvement [2].

In situations where granulomatosis with polyangiitis (GPA) is suspected but not definitively diagnosed, repeated biopsies may be warranted. To ensure accurate diagnosis and proper management, a multidisciplinary approach involving specialists from otolaryngology, rheumatology, and pathology may be necessary [9].

This paper details the case of a 26-year-old female who initially presented with otitis media and subsequently developed facial palsy and mixed conductive and sensory neural hearing loss. The patient's clinical presentation, diagnosis, and management of granulomatosis with polyangiitis are described, underscoring the significance of timely diagnosis and treatment to avoid grave complications.

Case Presentation

Introduction

This case report describes a 26-year-old female who presented with bilateral ear pain, reduced hearing, and ear discharge two months after experiencing a muffled sensation in her right ear following a common cold. The patient was initially treated for acute otitis media with possible labyrinthitis, but her symptoms worsened after reducing her prednisolone dose. Further investigations revealed a positive C-ANCA and high PR3, leading to a diagnosis of GPA involving only the middle and inner ears.

On examination, the patient had bilateral swollen and vascular tympanic membranes with mucoid discharge and moderate to severe mixed hearing loss. Flexible nasopharyngoscopy showed healthy nasal mucosa and a clear nasopharynx, and there were no signs of respiratory or kidney disease. The patient was treated with co-amoxiclav, ciprofloxacin ear drops, and oral prednisolone, and an MRI scan and microbiology swabs were taken from the ear discharge.

Ten days later, the patient reported immediate improvement in her symptoms with up to 80% hearing restoration, but her pain gradually increased, and her hearing loss and discharge worsened after reducing the prednisolone dose. On examination, bilateral tympanic membrane perforations with a swollen tympanic

Photo 1: CT axial view- bone window shows opacification of the mastoids and middle ear with no bone destruction.

membrane were seen. The patient received a higher dose of prednisolone for another ten days, and a CT temporal bone with contrast and vasculitis screening investigations were requested.

Photo 2: Post gadolinium MRI scan shows dural enhancement with no evidence of leptomenengitis or abscess formation.

The vasculitic screening revealed a positive C-ANCA and high PR3 with a level of 70, leading to a diagnosis of GPA involving only the middle and inner ears. Further investigations were arranged to rule out any other systematic disease. The patient had an examination of her ears under general anesthesia with biopsy of the middle ear mucosa and grommet insertion. She was also reviewed and treated by a rheumatology team who gave her three doses of intravenous methylprednisolone 750mg daily and started Rituximab.

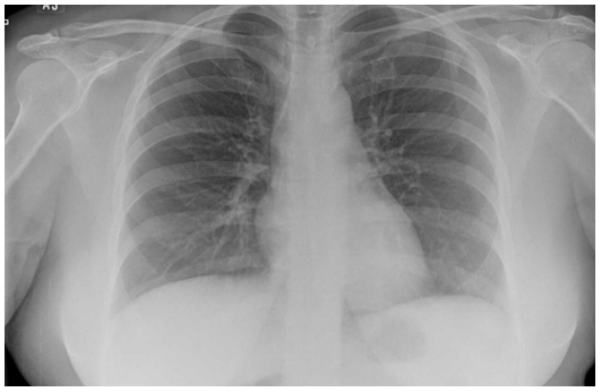

Photo 3: Chest X-ray shows no lung involvement.

Results

The patient's CT and MRI results showed opacification in the middle ear cavities and mastoids with no bone erosion. The MRI raised some concerns about meningeal involvement, which was ruled out after further review and multidisciplinary team discussion. The patient's urine PCR was standard, and her eGFR was >90. Her CRP peaked at 49 then was 6.3 after treatment.

Treatment and outcome

The patient's condition was discussed with a rheumatologist who advised her to increase her prednisolone dose to 35mg orally once daily for two weeks. She also received three doses of intravenous methylprednisolone 750mg daily, and Rituximab was started. While on treatment, the patient developed facial nerve palsy, grade 4 of the House-Brackmann Classification.

After four weeks of treatment, the patient's hearing and facial palsy significantly improved, and she had a facial nerve palsy grade 2. She is currently on prednisolone and Rituximab doses.

Discussion

This case highlights the importance of considering vasculitis in patients presenting with bilateral ear pain, reduced hearing,

and ear discharge. GPA involving only the middle and inner ears is rare, and early diagnosis and treatment can prevent significant morbidity. The use of a multidisciplinary team approach is essential for managing such cases. The development of facial nerve palsy in this patient highlights the need for careful monitoring of patients undergoing treatment for GPA.

Conclusion

In conclusion, this case report describes a rare presentation of GPA involving only the middle and inner ears in a 26-year-old female. The patient initially presented with bilateral ear pain, reduced hearing, and ear discharge, which worsened after reducing the prednisolone dose. Further investigations revealed a positive C-ANCA and high PR3, leading to a diagnosis of GPA. The patient received treatment with co-amoxiclav, ciprofloxacin ear drops, and prednisolone, followed by three doses of intravenous methylprednisolone 750mg daily and Rituximab. Despite the development of facial nerve palsy during treatment, the patient's condition significantly improved after four weeks of treatment. This case highlights the importance of considering vasculitis in patients presenting with otologic symptoms and the need for a multidisciplinary team approach in managing such cases. Early diagnosis and prompt initiation of appropriate therapy can improve outcomes and prevent potential complications.

Bibliography

1. Kühn D., *et al*. "Manifestation of Granulomatosis with Polyangiitis in Head and Neck". *Clinical and Experimental Rheumatology* 2 (2018): 78-84.

2. Djerić D., *et al*. "Otitis media with effusion as an initial manifestation of granulomatosis with polyangiitis". *SAGE Open Medical Case Reports* 9 (2021): 2050313.

3. Wawrzecka A., *et al*. "Granulomatosis with polyangiitis with bilateral facial palsy and severe mixed hearing loss". *Case Reports in Otolaryngology* 2016 (2016): 5206170.

4. Kukushev G., *et al*. "Two clinical cases of granulomatosis with polyangiitis with isolated otitis media and mastoiditis". *Reumatologia* 55 (2017): 256-260.

5. Wojciechowska J and KręCicki T. "Clinical characteristics of patients with granulomatosis with polyangiitis and microscopic polyangiitis in ent practice: a comparative analysis". *Acta Otorhinolaryngologica Italica* 38 (2018): 517-527.

6. Goh KY., *et al*. "Intractable chronic otitis media, rhinosinusitis and facial palsy: a case of active granulomatosis with polyangiitis". *BMJ Case Reports* 13 (2020): e233685.

7. Carnevale C., *et al*. "Head and Neck Manifestations of Granulomatosis with Polyangiitis: A retrospective analysis of 19 patients and review of the literature". *International Archives of Otorhinolaryngology* 23 (2019): 165-171.

8. Andrada-Elena M., *et al*. "Wegener's granulomatosis with orbital involvement: case report and literature review". *Romanian Journal of Ophthalmology* 65 (2021): 93-97.

9. Trandafir CM., *et al*. "Granulomatosis with Polyangiitis (GPA)-A Multidisciplinary Approach of a Case Report". *Medicina* 58 (2022): 1837.

10. Amiraraghi N., *et al*. "Primary otological manifestations of granulomatosis with polyangiitis: a case series". *The Journal of Laryngology and Otology* 129 (2015): 179-182.