Granulomatosis with Polyangiitis Presenting with Bilateral Hearing Loss and Facial Paresis

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Granulomatosis with polyangiitis (GPA) is a rare vascular inflammatory disease that affects the upper and lower respiratory tract and kidneys. Although the etiology of GPA is unknown, it is thought to be triggered by environmental events among patients with genetic susceptibility.1 Due to the frequency of upper respiratory tract involvement (70%-100%), otorhinolaryngologic symptoms may be the first clinical manifestation of disease.1

Case Presentation

We present a case of a patient who uniquely presented with bilateral otitis externa, with subsequent hearing loss and unilateral facial paresis as the initial signs of GPA. The Temple University School of Medicine Institutional Review Board determined that this case report is exempt from review. A 68-year-old woman with a history of bilateral otitis externa and septal perforation following cocaine use initially presented with hearing loss and aural fullness of 2 weeks’ duration. Her examination revealed purulent otorrhea, external auditory canal edema, and erythema consistent with acute otitis externa. She was treated and subsequently lost to follow-up. A month later, she presented to the emergency room with left facial paresis and bilateral hearing loss. There, head computed tomography (CT) showed partial opacification of the left middle ear and mastoid and absence of the nasal septum and turbinates (Figure 1). Chest CT showed bilateral pulmonary nodules. She presented again to the otolaryngology service. At that time, no mass was present on nasopharyngoscopy, and her facial function was normal. On otologic examination, she had developed left serous otitis media and mixed hearing loss. Myringotomy and tube placement were performed. Cultures of mucopurulent material grew methicillin-sensitive Staphylococcus aureus, which was treated with an extended course of antibiotics but did not resolve. Laboratory evaluation for sarcoidosis was negative.

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Follow-up imaging showed an apical pleural mass, bilateral pulmonary nodules, and renal and breast masses. Core biopsy of the breast and incisional breast biopsy revealed microabscesses and poorly formed granulomas with giant cells. Immunostains (CD20, CD3, CD5, CD43, CD10, bcl-2, CD21, S-100, CD68, and CD1a) supported a reactive process. Polymerase chain reaction studies for B-cell clonality and stains for acid-fast and fungal organisms were negative. Serum myeloperoxidase antibody was slightly elevated, as was proteinase 3 antibody. Subsequent lung wedge biopsy was indicative of GPA, including areas of geographic necrosis, granulomatous inflammation, and capillaritis (Figure 2). She was subsequently treated with rituximab and prednisone for 2 weeks and is currently maintained on azathioprine and prednisone with supplemental Prolia (denosumab), calcium D, and cholecalciferol for severe steroid-induced osteoporosis.

Discussion

Patients with GPA frequently present with a number of head and neck complaints, including epistaxis, rhinorrhea, nasal obstruction, and spontaneous septal perforation.\textsuperscript{2} Otologic manifestations, including serous otitis media, chronic otitis media, hearing loss, vertigo, and mastoiditis, are found among 35% of patients with GPA. GPA-induced otitis media can be associated with facial palsy and hearing loss (either sensorineural or conductive) and does not resolve with antibiotic therapy or surgery.\textsuperscript{3} Spontaneously resolving facial paralysis, as noted in this case, has not previously been described.

Nonenhanced CT images can indicate the extent of the disease, but findings are not specific for GPA.\textsuperscript{4} The nasal septum may be perforated and the turbinates shortened, and the paranasal sinuses demonstrate mucosal thickening and opacification.\textsuperscript{4} Subglottic stenosis occurs frequently.

Inflammation of the temporal bone results in soft tissue density in the mastoid and middle ear cavities, typically bilaterally.

Diagnosis of GPA is confirmed by histology and serologic testing, including elevated myeloperoxidase- and proteinase 3–ANCA levels. Head and neck biopsies are often nondiagnostic unless obtained from the paranasal sinuses (45% vs 84%).\textsuperscript{5} Pathognomonic biopsy characteristics include geographic necrosis, poorly formed granulomas, scattered giant cells, and microabscesses.\textsuperscript{4,5} Geographic necrosis is described as basophilic patches of tissue with serpiginous borders surrounded by granulomatous inflammation with histiocytes or giant cells (Figure 2). Vascular changes include fibrinoid necrosis of blood vessel walls and granulomatous inflammation. Renal biopsy typically reveals active glomerulonephritis.\textsuperscript{1}

Untreated severe GPA can lead to 90% mortality within 2 years, with death due to renal or respiratory failure.\textsuperscript{1} With treatment, 5-year survival is 75% to 88%. Classically, cyclophosphamide and glucocorticoids have been utilized to control severe GPA; however, rituximab has been approved by the Food and Drug Administration as an alternative to cyclophosphamide since 2011. Less severe disease may be treated with trimethoprim-sulfamethoxazole or dapsone. Treatment duration is tailored to control of symptoms, and maintenance of remission is continued for 18 months. Relapses are common (up to 93%).\textsuperscript{1} Permanent otolaryngologic sequelae include saddle nose and nasoseptal deformities and hearing loss.\textsuperscript{2,4,5}

Complex systemic illnesses can present with otolaryngologic manifestations and may present solely with head and neck complaints. This case highlights the importance of maintaining a wide differential during the evaluation of these patients.

Author Contributions

Taha Mur, first author, drafting, design of the work, final approval, accountability for all aspects of the work; Marian Ghrabi, drafting, data analysis, final approval, accountability for all aspects of the work; Jasvir S. Khurana, drafting, interpretation of the data, final approval, accountability for all aspects of the work; Pamela C. Roehm, corresponding author, conception and design of the work, drafting, final approval, accountability for all aspects of the work.

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