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Sensorineural Deafness and Facial Palsy: A Rare Presentation of Granulomatosis with Polyangiitis in a Saudi Patient – Case Report with Review

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1. ABSTRACT

Granulomatosis with polyangiitis (GPA) is a rare autoimmune vasculitis typically affecting the respiratory tract and kidneys. This case report describes a 33-year-old Saudi female presenting with persistent otitis media, progressive hearing loss, and facial nerve paralysis unresponsive to conventional treatment. She later developed systemic symptoms, including oral ulcers, gastrointestinal bleeding, and a cecal mass. Histopathological analysis of the middle ear and cecal tissue confirmed GPA, despite the absence of renal or pulmonary involvement. Serology revealed elevated PR3 antibodies. The patient showed marked improvement following immunosuppressive therapy with steroids and rituximab. This case underscores the need to consider GPA in patients with atypical, treatment-resistant otologic symptoms and highlights the importance of early recognition and multidisciplinary management to prevent irreversible complications.

Keywords: Wegener's granulomatosis; Granulomatosis with polyangiitis; Otitis media; Facial nerve palsy; Oral ulcer

2. ABBREVIATIONS

GPA	Granulomatosis with Polyangiitis
WG	Wegener's Granulomatosis
PR3	Proteinase 3
ANA	Antinuclear Antibody
ANCA	Antineutrophil Cytoplasmic Antibody
OME	Otitis Media with Effusion
NSAIDs	Nonsteroidal Anti-Inflammatory Drugs
ER	Emergency Room
CT	Computed Tomography
PNS	Paranasal Sinuses
CSF	Cerebrospinal Fluid
TB	Tuberculosis
CHL	Conductive Hearing Loss
BC	Bone Conduction
MHL	Mixed Hearing Loss
CND	Cannot Be Determined
MRI	Magnetic Resonance Imaging
VT	Ventilation Tube
IV	Intravenous
SRT	Speech Reception Threshold
SDS	Speech Discrimination Score
PTA	Pure Tone Audiometry
WBC	White Blood Cell Count
BUN	Blood Urea Nitrogen
AKI	Acute Kidney Injury
EEG	Electroencephalogram
MRV	Magnetic Resonance Venography
HSV	Herpes Simplex Virus
AFP	Alpha-Fetoprotein
ICA	Internal Carotid Artery
IJV	Internal Jugular Vein
GI	Gastrointestinal
BVAS	Birmingham Vasculitis Activity Score
CAP	Community-Acquired Pneumonia
OD	Once Daily
SNHL	Sensorineural Hearing Loss
ESRD	End-Stage Renal Disease
C-ANCA	Cytoplasmic Anti-Neutrophil Cytoplasmic Antibodies
CT PNS	Computed Tomography of Paranasal Sinuses
HBS	House-Brackmann Scale
SSNHL	Sudden Sensorineural Hearing Loss

3.0 INTRODUCTION

Granulomatosis with polyangiitis (GPA), formerly known as Wegener's granulomatosis (WG), is a rare, multisystem autoimmune disease characterized by necrotizing granulomatous inflammation primarily affecting the upper and lower respiratory tract and kidneys. The estimated annual incidence is approximately ten cases per million population.[1] While its exact etiology remains unknown, considerable evidence supports an autoimmune pathogenesis.[2] The clinical presentation of GPA is highly variable, often leading to delayed diagnosis and treatment. Early symptoms may be nonspecific or mimic more common conditions, contributing to diagnostic challenges and the risk of irreversible organ damage or death if untreated.[3] Although sinonasal, pulmonary, and renal involvement are the classical manifestations of GPA, initial presentation with isolated otologic or ophthalmologic symptoms is rare and often misdiagnosed.[4]

We report a case of GPA in a 33-year-old Saudi female who presented with a two-month history of persistent otitis media, progressive hearing loss, and left-sided facial nerve paralysis in the absence of classical pulmonary or renal features. Despite repeated antibiotic treatments and supportive care, her condition progressed, eventually leading to the development of gastrointestinal symptoms and a cecal mass. Histopathological analysis confirmed the diagnosis of GPA. Serological evaluation revealed negative antinuclear antibody (ANA) and antineutrophil cytoplasmic antibody (ANCA) results, but a markedly elevated proteinase 3 (PR3) titer of 134 IU/mL, further supporting the diagnosis. This case underscores the importance of early recognition of atypical presentations of GPA and highlights the role of a multidisciplinary approach in achieving timely diagnosis and effective treatment.

4.0 CASE REPORT

This was a case of a 33-year-old Saudi female, not known to have any medical issues, with a history of acute left ear pain associated with ear fullness and progressive hearing loss for two months. She presented to another hospital and underwent left ear Grommet tube insertion as a case of otitis media with effusion (OME). The patient started to have a persistence of acute left ear pain and discharge (watery and sometimes yellowish discharge). Ear pain was described to radiate to the jaw and neck. She also experienced severe headaches that improved with frequent intake of NSAIDs. Symptoms mildly improved after Grommet tube insertion, in a matter of pain only. However, the discharge has persisted and progressively increased. The bothersome otalgia resulted in multiple ER visits. She also received about nine types of antibiotics (orally and intravenously) without improvement. There was no history of ear surgery, trauma, other neurological symptoms, no other associated otological symptoms. She was referred to our center as a case of persistent otitis media and a suspected tegmen defect seen on a CT scan in the primary center. A CT scan of the paranasal sinuses (PNS) was requested and is pending, as nasal symptoms and findings are among the most common presentations of Wegener's granulomatosis (GPA). This evaluation is crucial to assess sinonasal involvement, which is often a key diagnostic feature of GPA.

Upon examination, the patient was in acute pain. The left ear examination revealed an inflamed, non-stenotic external auditory canal filled with watery, yellowish fluid, obstructing the Grommet tube. The tympanic membrane appeared inflamed and pulsating, with no evidence of granulation tissue or keratin. Notably, cerebrospinal fluid (CSF) leakage was excluded by the absence of a halo sign on fluid testing. Examination of

the right ear showed mild clear fluid in the canal and an inflamed tympanic membrane without obvious perforation. A nasal examination showed signs of allergic rhinitis. The tuning fork test showed Weber to the left, whereas Rinne's test showed the left ear negative and the right ear positive. Multiple cultures from the ear fluid were taken, and all of them were negative for bacteria, viral, fungi, and tuberculosis (TB. Figure 1 shows the left ear endoscopic examination). First visit pure tone audiometry evaluation showed mild to moderate conductive hearing loss (CHL) on the right ear where bone conduction (BC) masking cannot be detected due to over-masking with excellent speech discrimination score, whereas a severe mixed hearing loss (MHL) with excellent speech discrimination score was noted with the left ear. Tympanometry cannot be determined (CND) due to excessive discharges from both ears. (Figure 2)

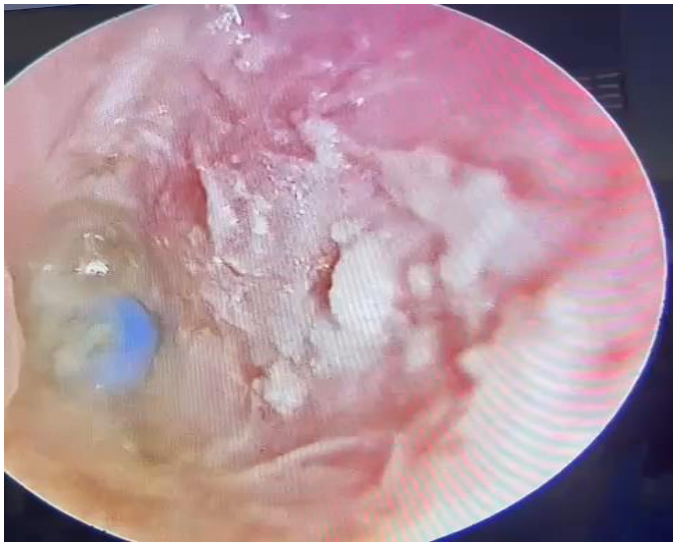

Figure 1: Left ear Endoscopic examination

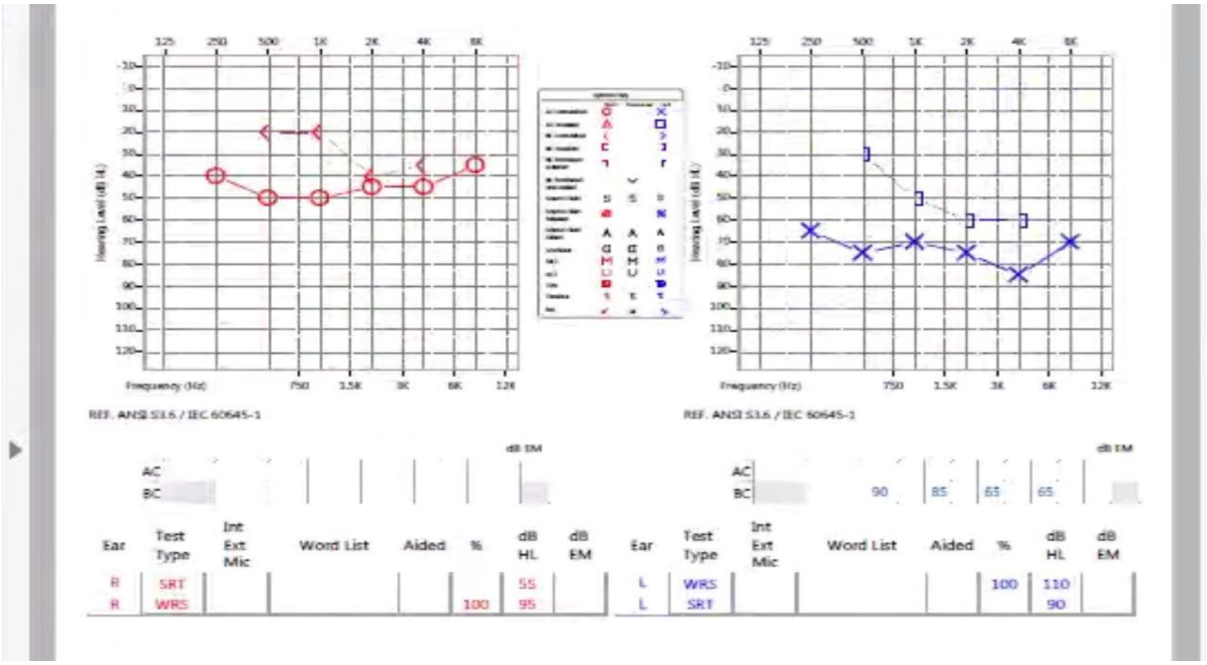

Figure 2: First visit pure tone audiometry evaluation.

On follow-up, the patient came post CT and MRI, which showed bilateral ear otomastoiditis with left small suspected cholesteatoma, and the initial report suspected a small tegmen defect. The patient showed mild

improvement compared to the last visit. Culture came negatively. The disease started in the other ear but was still improved after a single dose of steroids. A high suspicion of autoimmune and rheumatological disease was entertained. The culture was repeated. A review of the radiological images showed no picture of cholesteatoma with no clear skull base defect. She was managed with a tapering dose of steroid and analgesia and was requested to follow up in two weeks.

A week later, she again presented to the ER with acute right ear pain, as the disease progressed to the contralateral ear. Right ear myringotomy was done, and no pus came out. A trial of ventilation tube (VT) insertion was done. A swab was taken from the right external auditory canal. She was admitted as a case of right acute mastoiditis for IV antibiotics and further investigation. She presented with right eye swelling and was seen by the ophthalmology team. She was started on IV vancomycin and tazocin as per the suggestion by the infectious disease team. Audiological assessment showed a speech reception threshold (SRT) of 60 dBHL on the right ear and 70 dBHL on the left ear. The speech discrimination score (SDS) of the right ear showed 100% at 100 dBHL and 80% at 100 dBHL on the left ear. Tympanometry showed Type B flat with normal ear canal volume on the right ear and CND due to a recent Grommet on the left ear. PTA showed moderate sloping to severe MHL of the right ear and mild sloping to profound MHL of the left ear. (Figure 3) This time patient had severe hearing loss and was only able to communicate through writing on a piece of paper or by sign language.

During admission, the patient started to have an oral cavity ulceration on the left molar area. The maxillofacial surgery team was consulted. She also had two episodes of bloody vomiting and was seen by the medical team, for which steroids were stopped. She continued to have severe otalgia. Laboratory investigation showed a WBC of 11.8, negative for antinuclear antibody (ANA), and antineutrophil cytoplasmic antibodies (ANCA), but positive for proteinase 3 (PR3), and C-reactive protein was high at 220 mg/dL. She also had three episodes of passing dark red blood per rectum, not associated with stool. Furthermore, she had anorexia, nausea, and epigastric pain. Her blood urea nitrogen (BUN) was 5.3 mg/dL, and her serum creatinine was 173 umol/L. She was referred to the nephrology team, who suspected a multifactorial AKI as she had poor oral intake with active loss from vomiting and diarrhea. The nephrology team also entertained the possibility of acute interstitial nephritis with a history of significant use of NSAIDs or antibiotics. They suggested keeping the patient on intravenous fluids and avoiding nephrotoxic medications, as well as using contrast media with medication adjustments from the pharmacist.

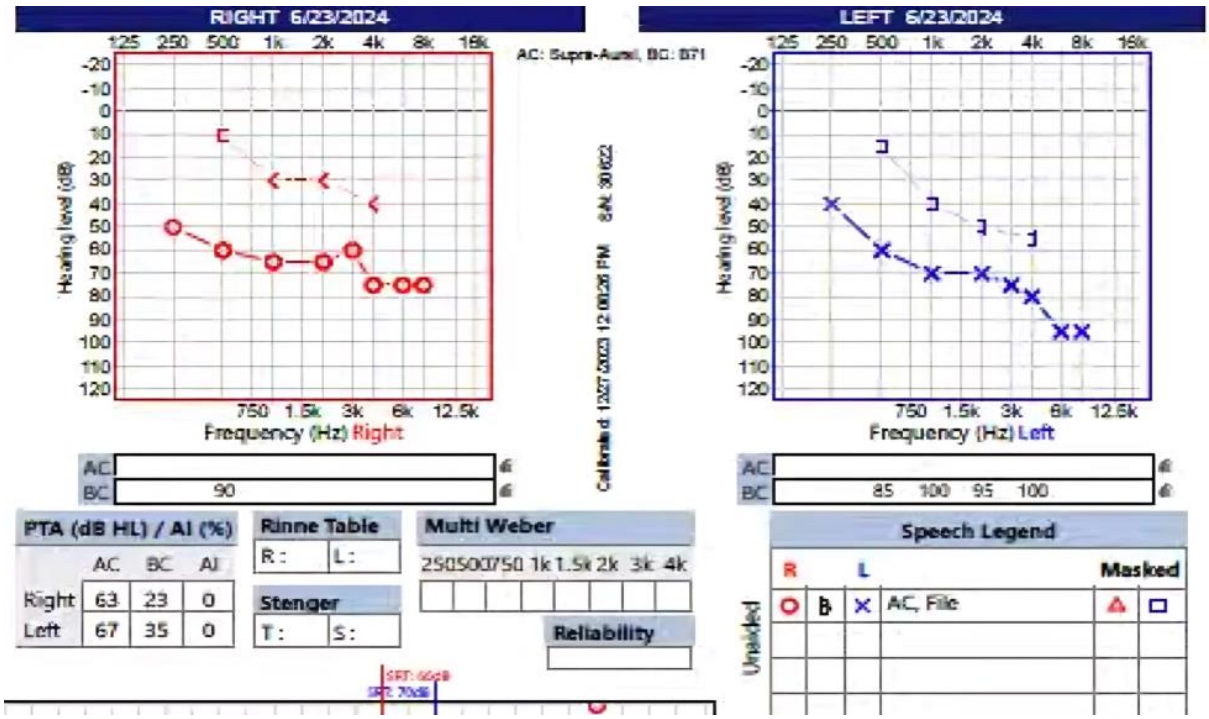

Figure 3: Follow-up pure tone audiometry evaluation.

On her 9th hospital day, she started to complain of jerky head and limb movement with spasms, hand numbness, and unsteady gait. Lower limb skin rash was also noticed. Her left facial nerve palsy Grade IV was managed with intra-tympanic steroid injection with minimal improvement. Ear bacterial and fungal culture and sensitivity came out negative, as well as a negative stool culture. A multispecialty plan was made available. The gastroenterology team suggested an endoscopy if the hemoglobin level was <7.0 g/dL. The neurology team suggested a lumbar puncture, which came out negative. A biopsy from the oral cavity ulcer was suggested and taken by the maxillofacial team, and a full serology panel from the rheumatology team was ordered.

An urgent electroencephalogram (EEG) and brain magnetic resonance imaging (MRI)/magnetic resonance venography (MRV) with nephrology clearance. The oral and maxillofacial team was also consulted on that same hospital day and suggested to be referred to the dental clinic for a possible biopsy to rule out vasculitis or an autoimmune disease. Furthermore, the infectious disease team was reconsulted for the refractory process of her disease and minimal improvement on the intravenous antibiotics. Lumbar puncture was suggested for CSF staining and culture, HSV PCR, AFP, TB PCR, Brucella, and fungal culture. She was switched from Tazocin to a meropenem meningitis dose. CT scan of the temporal bone showed complete opacification of both the right and left middle ear and mastoid. The left middle ear cavity showed focal hazy calcifications, whereas the bilateral scutum, tegmen tympani, mastoideum, and internal auditory canals were intact. MRI of the internal auditory canal showed evidence of heterogenous signal intensity completely filling the bilateral mastoid air cells. The middle ear showed a small focus of diffuse restriction, keeping with a cholesteatoma focus. There was also a small bony defect of the tegmen mastoideum. The impression based on the MRI was a cholesteatoma within the background of severe left otomastoiditis with moderate to severe contralateral right otitis media and mastoiditis. MRI of the brain showed a complicated acute bilateral otomastoiditis with extension along the eustachian tubes and extensive involvement of the deep upper neck spaces, moderate to severe narrowing of the

encased upper cervical segment of the left internal carotid artery (ICA) as well as the internal jugular vein (IJV), and a suspected skull base involvement along the undersurface of the left petrous apex concerning for focal skull base osteomyelitis. There was also a suspected intracranial extension through the left foramen ovale with focal dural thickening along the anterior aspect of the left Meckel's cave, an abnormal enhancement along the left cochlear basal turn denoting acute labyrinthitis, and a subtle abnormal facial nerve enhancement, especially on the left side.

After clearance from all the involved multispecialty teams, the patient was booked for left ear exploration. Intra-operative findings showed the distorted middle ear cavity anatomy where the cavity is filled with inflammatory polypoidal-like mucosa, no secretions or pus, no bony defects, no skull base defect, and the tegmen both tympani and mastoideum are intact. The facial canal was intact with no bony dehiscence. The diseased chorda tympani was also intact, as well as all ossicles. Middle ear mucosa and mastoid bone specimens were sent for histopathological analysis. Furthermore, the GI team suggested that a colonoscopy be done to rule out possible involvement of a likely GPA, which revealed a fungating mass arising from the cecal base. A biopsy was taken from the cecal mass for histopathology. Both cecal mass and middle ear mucosa and bone showed granulomatous inflammation, which was suggestive of granulomatosis (GPA)/ Wegener's GPA. Her Birmingham Vasculitis Score (BVAS) was 23. She was started on steroids and rituximab (1 cycle, 1st dose 1 gm, 2nd dose 1 gm, then 1 gm every 6 months.

Three months later, she was again admitted to our institution with pleuritic chest pain for one week and was admitted as a case of community-acquired pneumonia (CAP); she completed the 7-day antibiotic course. She was started on prednisone 25mg, then methylprednisolone 25 mg, shifted to oral prednisone 60 mg OD every three days until a baseline dose of 25 mg once daily. Blood, urine, and sputum cultures all came back negative. Computed tomography pulmonary angiogram showed features of pulmonary edema with bilateral ground glass opacification and infiltration suggestive of pulmonary edema, and mild pericardial fluid but no major pulmonary embolism, pleural effusion, or pneumothorax. Labs were unremarkable. She was discharged on her 6th hospital day with significant improvement, appeared healthy, was vitally stable, and was able to communicate freely. However, there was still an apparent facial asymmetry due to the destructive effects and manifestations of her GPA-WG.

At her recent follow-up visit, the patient reported being pain-free, with significant improvement in her ear discharge and a slight improvement in hearing since the initial presentation. Examination of the left ear revealed a dry canal with no active discharge, and the tympanostomy tube was still present in the canal. The right tympanic membrane appeared dry and clean. While the facial nerve persisted, it improved to grade 3 on the House-Brackmann grading system. The patient can now close her eye with minimal effort and has mild drooping of the left corner of the mouth, although no asymmetry was noted at rest on the 3rd month of follow-up after initiating GPA medications. of this writing, she is continually followed up as an outpatient in our clinic with repeated Audiology.

5.0 DISCUSSION

GPA, formerly known as Wegener's granulomatosis, is a rare autoimmune vasculitis primarily affecting small vessels, with typical involvement of the upper and lower respiratory tracts and kidneys.[2] However, atypical presentations can pose significant diagnostic challenges, leading to delays in appropriate treatment.[4] This case of GPA has been confirmed and presents a unique combination of clinical features, including sensorineural hearing loss, facial nerve paralysis, ocular complications, a cecal mass due to vasculitis, and large vessel vasculitis, all of which showed significant improvement following the administration of suitable immunosuppressive treatment.

5.1 Facial Nerve Paralysis in GPA-WG: Possible Mechanisms

Facial nerve paralysis in GPA is a rare but recognized manifestation, occurring through multiple potential mechanisms. In our case, the patient developed left-sided facial nerve palsy, likely resulting from either secondary inflammation due to severe left otitis media, compression of the internal auditory canal due to adjacent inflammatory processes, or primary vasculitic involvement of the facial nerve. While primary facial nerve vasculitis has been reported in GPA,[2] it remains an uncommon presentation, making secondary compression due to extensive inflammation a more plausible explanation in this case.

Facial nerve paralysis in GPA is uncommon, often resulting from secondary nerve compression rather than direct vasculitic involvement. According to Iannella et al. (2016), facial nerve palsy typically occurs as the disease progresses, rather than at initial presentation.[4] While necrotizing vasculitis of the "vasa nervorum" is considered a key mechanism, our case lacked destructive bone changes, supporting inflammation-induced compression as the primary cause.[4]

Pagnoux and Villa-Forte (2023) note that GPA predominantly affects small vessels, commonly involving the respiratory tract, lungs, and kidneys.[5] Facial nerve paralysis occurs in 5–10% of cases, usually associated with otomastoiditis. In our patient, chronic bilateral otomastoiditis likely contributed to facial nerve dysfunction, reinforcing the secondary compression hypothesis.

Differentiating between primary vasculitis and inflammatory compression is crucial for treatment. Inflammatory compression, rather than direct vascular injury, is more responsive to surgical interventions like myringotomy and ventilation tube insertion. Our patient showed partial recovery with systemic immunosuppression (steroids and rituximab), improving her HBS from Grade IV to III over three months. This aligns with previous studies where facial nerve function improves variably with treatment, emphasizing the need for early diagnosis and targeted management.[4]

5.2 Hearing Loss and Audiological Manifestations in GPA

Hearing loss is a recognized but variable manifestation of GPA, occurring in approximately 8–65% of cases.[6] Most studies describe sensorineural hearing loss (SNHL) as the predominant type; however, MHL and CHL have also been reported.[7] The pathophysiology of GPA-related hearing loss remains unclear but is thought to be multifactorial, involving both direct vasculitic damage and secondary effects from chronic otitis media or mastoiditis.[6]

Our patient exhibited MHL, conductive in the right ear and mixed in the left, suggesting a mechanical component such as chronic otitis media. This contrasts with the findings of Rahne et al. (2017), who identified predominantly SNHL in GPA patients. Their study noted that many previous investigations failed to measure bone-conduction thresholds, limiting differentiation between SNHL and CHL.[7] Our case, supported by a comprehensive audiological assessment, underscores the importance of recognizing mixed and conductive components in GPA-related hearing loss.

The prevalence and mechanisms of hearing loss in GPA have been a topic of debate. Vainutienė et al. (2024) conducted a detailed analysis of audiological manifestations in GPA patients and found that 60.7% had hearing impairment, with SNHL being the most common (32.1%), followed by MHL (21.4%) and CHL (7.1%).[6] These findings align with other studies indicating SNHL as the predominant form of hearing impairment in GPA.[7] However, our case differs in that MHL was a significant feature, reinforcing the need for individualized audiological evaluation.

In contrast, studies such as Rahne et al. (2017) suggested that conductive components in GPA-related hearing loss might be underreported due to incomplete audiological assessments. Their study demonstrated a significant air-bone gap in GPA patients, suggesting CHL as a contributing factor, likely due to middle ear pathology.[7] Our patient exhibited similar findings, with imaging confirming otomastoiditis, which likely contributed to CHL. Another key study by Batinović et al. (2023) described a 36-year-old male with otologic symptoms, including hearing loss and facial palsy, as the initial presentation of GPA.[8] Like our patient, this case involved significant middle ear involvement, with profound hearing loss and tympanic cavity opacification. This reinforces the role of chronic inflammation and granulomatous disease in GPA-related CHL.

Cochlear involvement in GPA can lead to irreversible SNHL, particularly in untreated or progressive cases.[9] Some studies suggest that cochlear implantation may be an option for GPA patients with profound SNHL, but there is no consensus on the optimal management approach.[9] The literature highlights that delayed diagnosis and therapy negatively impact hearing prognosis, making early recognition essential.[6] Our patient's hearing loss was partially reversible with immunosuppressive therapy, suggesting an inflammatory rather than a purely vasculitic etiology. This aligns with findings from Gupta et al. (2023) and Koenen et al. (2022), where patients initially misdiagnosed with otitis media later developed facial nerve palsy and were ultimately diagnosed with GPA.[10,11] Their hearing outcomes varied, with some improvement following systemic immunosuppression but persistent hearing deficits in severe cases.

Hearing loss in GPA can occur alongside other systemic manifestations. Liang et al. (2019) reported that 50% of cases with both otologic and ophthalmologic involvement initially presented with peripheral ulcerative keratitis and suppurative otitis media.[12] Our case follows this pattern, where hearing loss was a primary manifestation, underscoring the importance of multi-system evaluation in suspected GPA cases. Dermatologic symptoms are often early indicators of GPA but are not pathognomonic. Kihiczak et al. (1994) highlighted that skin manifestations can persist for months or even years before severe multi-organ involvement.[13] In our case, while the patient's primary presentation was otologic, the presence of a lower limb skin rash further supported the suspicion of GPA. Dermatologic manifestations, though not always present, can be an early indicator of systemic vasculitis. This reinforces that GPA should be considered even in patients initially presenting with

isolated otologic symptoms, particularly when accompanied by additional systemic signs such as cutaneous involvement.

GPA, formerly known as Wegener's granulomatosis, presents significant diagnostic and therapeutic challenges, particularly when otologic manifestations are the primary or early clinical features. In this case series, all three patients demonstrated resistance to initial antibiotic and corticosteroid therapy, consistent with prior reports of GPA-related otologic disease.[8,10,11] Similar patterns of treatment failure have been noted in the literature, emphasizing the necessity of considering GPA in cases of recalcitrant otitis media and progressive SNHL.[14, 15]

Hearing loss in GPA is multifactorial, predominantly sensorineural, and attributed to vasculitic involvement of the cochlear vasculature.[6,16] Our findings align with those of Vainutienė et al. (2024), who identified SNHL as the most common audiometric pattern in GPA, affecting approximately 32.1% of cases.[6] Other studies have reported varying degrees of conductive, mixed, or SNHL.[17] Notably, our case supports prior observations that GPA-induced otologic disease often presents with persistent serous otitis media and mastoiditis, which fail to resolve with standard antimicrobial therapy.[14]

Facial nerve involvement, while uncommon in GPA, has been documented in 5–10% of cases, often as a consequence of segmental vasculitis or nerve compression due to granulomatous inflammation.[18,19,20] The delayed diagnosis in our patient underscores the diagnostic complexity of GPA, particularly in cases where systemic manifestations such as renal involvement and cutaneous lesions emerge late in the disease course. This is in contrast to cases described by Gupta et al. (2023) and Koenen et al. (2022), where facial nerve paralysis was an early and defining feature, leading to more timely intervention.[10,11]

Furthermore, our case highlights the evolving role of cochlear implantation in GPA-related hearing loss. While cochlear implantation has shown promise in restoring hearing function and reducing reliance on long-term immunosuppressants,[9] there remains a lack of standardized guidelines for patient selection and surgical timing. Studies by Lee et al. (2021) and Vainutienė et al. (2024) suggest that cochlear implantation may offer functional auditory rehabilitation, even in patients with systemic autoimmune disease.[6,9] However, the potential for fluctuating hearing thresholds and disease progression warrants further investigation into long-term audiologic outcomes.

The findings of this study reinforce the need for heightened clinical suspicion of GPA in patients presenting with persistent otologic symptoms, particularly those unresponsive to conventional therapy. Diagnostic delay, as observed in our case, can contribute to irreversible SNHL and systemic complications. Early recognition through comprehensive audiometric evaluation, serologic testing, and histopathologic confirmation is critical in guiding appropriate immunosuppressive therapy.[15,16]

5.3 Large Vessel Involvement: A Rare Feature in GPA

GPA is primarily characterized by necrotizing vasculitis that predominantly affects small to medium-sized vessels, with granulomatous inflammation commonly targeting the upper respiratory tract, lungs, and kidneys.[21] Large vessel involvement, including the ICA and IJV, remains a rare and unusual manifestation of the disease. The pathophysiological mechanisms underlying this phenomenon are not fully understood but likely involve secondary perivasculitis due to extensive localized inflammation, possible direct extension of

inflammation from the Eustachian tube into adjacent vasculature, or an exceptionally rare co-occurrence of both small and large vessel vasculitis.

While small vessel vasculitis is a hallmark of GPA, large vessel involvement has been documented in limited case reports and literature reviews. A study by Ozaki et al. (2017) reviewed 24 cases of large vessel involvement in GPA, identifying luminal stenosis, wall thickening, and aneurysmal formations as common vascular manifestations.[22] The most frequently affected vessels were the abdominal aorta, thoracic aorta, and subclavian arteries, with ICA involvement reported in only four cases. These findings highlight the unusual nature of large vessel involvement in GPA and suggest that it may be underrecognized due to its rarity.

Anti-proteinase 3 (anti-PR3) and antineutrophil cytoplasmic antibody-C (c-ANCA) antibodies are frequently present in the blood and are typically detectable in GPA-WG cases.[21] However, we have not investigated the presence of antibodies in our series of examinations since any of the organs showed typical GPA-WG findings. Classic GPA typically presents with small vessel vasculitis affecting capillaries, venules, and arterioles, leading to manifestations such as necrotizing glomerulonephritis, pulmonary nodules, and upper airway granulomas.[21] In contrast, our case exhibited involvement of the left ICA and IJV, representing an atypical vascular distribution for GPA. This aligns with findings from Ozaki et al. (2017), where large vessel involvement was noted in a minority of cases, often presenting with periaortitis or arterial wall thickening rather than the classic small vessel pathology.[22] While GPA is traditionally classified as a small- to medium-vessel vasculitis, rare cases of large vessel involvement have been reported.[23]

In our patient, the involvement of large vessels could be due to either of two mechanisms: (1) a direct extension of inflammation from the Eustachian tube into the surrounding vasculature, leading to secondary vasculitis, or (2) the coexistence of both large and small vessel vasculitis, which has been documented in a small subset of GPA patients.[24]

Large vessel involvement in GPA can present as aortitis, aneurysms, or stenotic lesions, although these manifestations are more characteristic of Takayasu arteritis or giant cell arteritis rather than GPA.[23] However, recent studies suggest that overlapping vasculitic syndromes may exist, necessitating careful vascular imaging and serologic evaluation. Booth et al. (2003) reported a 5-year retrospective study in which 28% of patients with ANCA-associated vasculitis developed end-stage renal disease (ESRD), emphasizing the prognostic significance of systemic vasculitic involvement.[25] The presence of anti-proteinase 3 (PR3) and cytoplasmic anti-neutrophil cytoplasmic antibodies (c-ANCA) is a hallmark of GPA and aids in its diagnosis.[26] However, in our case, we did not investigate these markers due to the absence of classic small vessel GPA findings. This differs from most documented cases where PR3-ANCA positivity was observed in the majority of large vessel GPA cases, reinforcing the importance of serological testing in atypical presentations.[22]

5.4 Computed Tomography (CT) Scan of Paranasal Sinuses (PNS) and Sinonasal Involvement in GPA

Sinonasal involvement is a hallmark of GPA, occurring in approximately 85% of cases, with some studies reporting involvement in up to 100% of patients presenting with upper respiratory tract symptoms.[24]

However, our patient displayed an atypical presentation, initially lacking significant sinonasal symptoms, which delayed suspicion of GPA. Given the high prevalence of sinonasal disease in GPA, computed tomography (CT) of the paranasal sinuses (PNS) plays a critical role in evaluating disease extent and complications.

CT imaging of sinonasal structures in GPA reveals a spectrum of findings, including mucosal thickening (87.7% of cases), bony destruction (59.9%), and septal erosion (59.4%).[27] These features, although not pathognomonic, are highly suggestive of GPA when present in the appropriate clinical context. While our patient's CT PNS findings are still pending, the absence of sinonasal symptoms does not preclude a significant disease burden on imaging. In contrast, the systematic review by D'Anza et al. (2017) noted that most GPA-related sinonasal abnormalities on CT are incidental and often identified before clinical symptoms manifest.[27] This discrepancy underscores the importance of imaging in the early identification of GPA-related sinonasal involvement, particularly in cases where symptoms are minimal or absent.

Biopsy confirmation is another key diagnostic tool in GPA, often obtained from nasal or sinus tissues.[24] However, in our case, the diagnosis was confirmed via a biopsy of a cecal mass, which is a rare initial presentation. This deviation from the standard diagnostic pathway highlights the heterogeneous nature of GPA and the need for a multidisciplinary diagnostic approach. Furthermore, while sinonasal biopsies frequently demonstrate granulomatous inflammation, they can also yield nonspecific inflammatory changes, necessitating correlation with clinical and radiologic findings.[27] The reliance on non-sinonasal biopsy in our case aligns with the findings by Lynch et al. (2018), who emphasized the necessity of broad diagnostic strategies when conventional sites fail to provide definitive histopathologic confirmation.[23]

5.5 Left Eyelid Swelling

Ocular involvement in GPA is well documented, typically presenting as scleritis, episcleritis, uveitis, or orbital granulomatous masses.[28] However, isolated left eyelid swelling, as observed in our patient, is an uncommon manifestation, raising the possibility of an alternative etiology such as orbital cellulitis, idiopathic orbital inflammation, or thyroid-associated orbitopathy.

Most cases of orbital GPA present with proptosis and vision impairment, resulting from granulomatous inflammation extending into the orbit.[24,28]

In contrast, our patient exhibited no visual changes or proptosis, making a direct GPA-related orbital process less likely. MRI evaluation remains crucial in distinguishing between orbital involvement of GPA and other potential etiologies. In a review of imaging findings, D'Anza et al. (2017) reported that MRI often demonstrates retro-orbital masses with variable enhancement, indicative of granulomatous inflammation, yet in some cases, diffuse orbital infiltration can mimic other inflammatory or neoplastic conditions.[27]

In a case study reported by Khare and Ramachandran (2025), a 32-year-old male with ocular GPA developed a choroidal granuloma, which responded well to immunosuppressive therapy. While our case lacks such definitive granulomatous findings, the presence of eyelid swelling necessitates further investigation. The importance of prompt and targeted treatment in preventing irreversible ocular complications cannot be overstated. Given that corticosteroids alone may be insufficient in managing orbital involvement, the use of Rituximab or cyclophosphamide should be considered in cases with confirmed GPA-related orbital disease.[23]

Our case presents notable deviations from the conventional clinical course of GPA. While sinonasal involvement is nearly ubiquitous in GPA, our patient initially lacked significant sinonasal symptoms, highlighting the importance of imaging in detecting subclinical disease. In contrast, studies by D'Anza et al. (2017) underscore the high prevalence of sinonasal pathology in GPA, often preceding systemic symptoms.[27] Additionally, the atypical presentation of isolated left eyelid swelling further complicates the diagnostic picture. Most cases of orbital GPA manifest with more pronounced ophthalmologic symptoms, including proptosis and vision impairment, making isolated eyelid swelling an unusual finding.[28] This underscores the need for comprehensive imaging and interdisciplinary evaluation.

5.6 Diagnostic Challenges and Importance of Early Identification

GPA is classically diagnosed based on renal and pulmonary involvement, with respiratory and kidney manifestations serving as hallmark features of the disease. However, in this case, the absence of these typical findings led to a delay in diagnosis. Instead, the diagnosis was confirmed via biopsy of a fungating cecal mass, an unusual presentation for GPA. This highlights the need for a broader diagnostic approach in atypical cases where the classic organ involvement is absent.

Serological testing played a crucial role in confirming the diagnosis, with positive C-ANCA and anti-PR3 findings supporting the autoimmune etiology. While these serological markers are highly specific for GPA, their presence alone is insufficient for diagnosis without corresponding clinical or histopathologic evidence. In most cases, diagnosis relies on findings from the respiratory or renal systems, making this case distinct in requiring an intestinal biopsy for confirmation. The initial uncertainty surrounding the diagnosis led to delays in treatment initiation, emphasizing the importance of maintaining a high index of suspicion for GPA in patients with unusual presentations.

5.7 Treatment and Prognosis

The patient demonstrated a positive response to intravenous methylprednisolone, which provided rapid symptomatic relief. However, given the systemic nature of GPA, Rituximab was initiated as part of the long-term management strategy.[23,24,25] The initiation of Rituximab was delayed due to concerns regarding infection risk, underscoring the importance of balancing immunosuppression with infection control in GPA management.

Facial nerve function showed partial improvement, progressing to HBS Grade III, along with slight hearing recovery. Hearing outcomes in GPA are highly variable and largely depend on the timing of diagnosis and intervention. In this case, early immunosuppressive therapy contributed to the stabilization of symptoms, aligning with existing literature indicating that systemic corticosteroids and biologic agents can improve hearing function if initiated promptly.[8] However, complete recovery of hearing is uncommon, and residual deficits may persist despite aggressive treatment.

The standard treatment regimen for GPA typically includes cyclophosphamide and corticosteroids. However, due to the severity of systemic involvement in this case, Rituximab was preferred, consistent with evolving treatment paradigms that favor biological therapies in refractory or severe disease.[23] The effectiveness of Rituximab in inducing remission while minimizing the long-term adverse effects of cyclophosphamide makes it

a suitable option, particularly in patients with relapsing disease or those at risk for cyclophosphamide-related toxicity.[24]

Diagnosing GPA in patients presenting primarily with hearing loss remains particularly challenging due to the overlap with more common otologic conditions such as chronic otitis media and sudden sensorineural hearing loss (SSNHL). This case underscores the need for a high index of suspicion in patients with persistent or progressive auditory symptoms unresponsive to conventional therapies. Systemic immunosuppression remains the mainstay of treatment for GPA-related hearing loss, with corticosteroids and Rituximab demonstrating efficacy in stabilizing disease progression and partially restoring hearing function.

Surgical interventions such as ventilation tube insertion, mastoidectomy, or cochlear implantation may be required in cases of chronic otitis media, ossicular erosion, or profound SNHL. However, their role in GPA remains debated due to concerns about disease progression affecting surgical outcomes.[9]

In this case, medical management alone was sufficient to achieve partial hearing recovery, avoiding the need for surgical intervention.

The prognosis of untreated GPA is extremely poor, with a mean survival of only five months and a mortality rate nearing 90% within two years.[25] Early diagnosis and aggressive immunosuppressive treatment significantly improve survival rates. Long-term therapy with low-dose cyclophosphamide and prednisolone has been associated with an 80% five-year survival rate and a 75% remission rate.[23,24] With the emergence of biologic agents such as Rituximab, the treatment landscape has shifted toward targeted therapies that offer improved safety and efficacy in long-term disease control.

6.0 CONCLUSION

This is a confirmed GPA case with a rare presentation of sensorineural deafness, facial nerve palsy, ocular involvement, cecal vasculitic mass, and a large vessel vasculitis that responded significantly to appropriate immunosuppressive therapy. This underscores the need for early intervention to prevent irreversible damage. Given the high mortality of untreated GPA, prompt diagnosis and immunosuppressive therapy significantly improve outcomes. A multidisciplinary approach is essential for managing complex cases and ensuring timely and effective treatment.

Conflict of Interest Statement:

The authors declare that there is no conflict of interest regarding the publication of this case report.

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