Granulomatosis with polyangiitis presenting as acute bilateral facial nerve paralysis

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Introduction

- Granulomatosis with polyangiitis (GPA) is an auto-immune disease characterized by granuloma formation with necrotizing vasculitis that typically presents with respiratory or renal involvement.
- More than 70% have otolaryngologic complaints at presentation.
- Bilateral facial nerve paralysis is a rare presenting symptom with 18 prior cases reported in the English literature; however, all other cases presented with additional signs and symptoms.
- We present a unique case of bilateral facial palsy as the isolated symptom with no other systemic involvement.

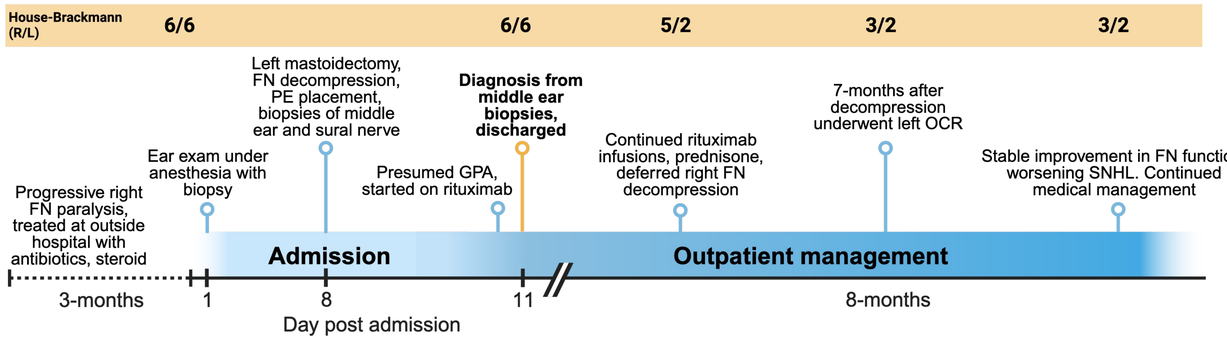
Case Presentation

- 52yo female with remote h/o right CWD mastoidectomy (>20y prior)
- Presented with bilateral facial nerve (FN) paralysis. Right FN paralysis had started 2 months earlier and left FN paralysis 3 days prior to presentation. No prior steroid treatment.

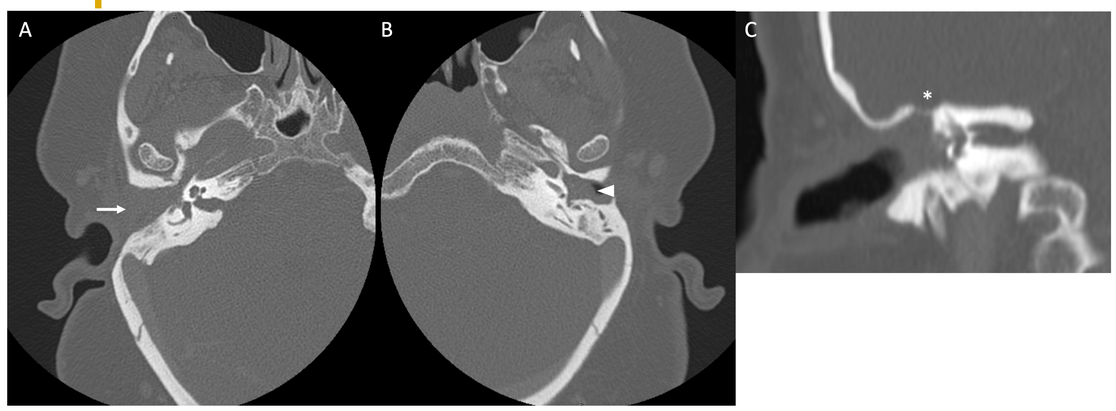
Exam findings:

- Bilateral - House-Brackmann VI.
- No sinonasal inflammation.
- Otomicroscopy: bilateral polypoid tissue occluding EAC with purulent discharge.

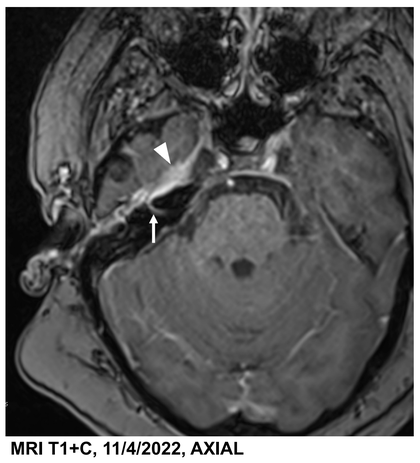
- Clinically, non-serviceable hearing bilateral. Audiogram revealed severe to profound mixed hearing loss, left worse than right (*see Audio A*).
- CT chest & renal function – WNL.
- Exam under anesthesia was performed due to patient's intolerance and biopsies were taken from polypoid tissue.
- Rheumatologic workup negative with exception of positive PR3, imaging concerning for granulomatous disease



Workup / Results



CT sinuses, 11/4/2022, AXIAL of the right ear (A) and left ear (B), coronal section, right ear (C).
Findings consistent with history of right canal-wall-down mastoidectomy with soft tissue filling the mastoid bowl (white arrow). Dehiscence of the tympanic segment of the facial nerve and a left sclerotic mastoid cavity with complete opacification. Soft tissue noted filling the left external ear canal (white arrow-head. No dehiscence of the fallopian canal along the tympanic segment (not appear in the image; thinning or dehiscence of the tegmen-tympani of the right ear, adjacent to the area of dural thickening on the MRI (white asterisk).



MRI T1+C, 11/4/2022, AXIAL
Findings: Soft tissue thickening along the external auditory canals and mastoid bowl. Post-contrast enhancement of the labyrinthine portions of the facial nerve (white arrow). Dural enhancement, more pronounced on the right, with a 5mm thickness (white arrow-head)

Pathology Results

Initial ear exam under anesthesia

FINAL DIAGNOSIS
A. LEFT EXTERNAL AUDITORY CANAL CONTENTS (BIOPSY):
- Blood and fibrinopurulent debris with focal bacterial clusters.

B. RIGHT EAR CANAL (BIOPSY):
- Skin with acute inflammation.
- No evidence of malignancy.

At time of tympanomastoidectomy / FN decompression

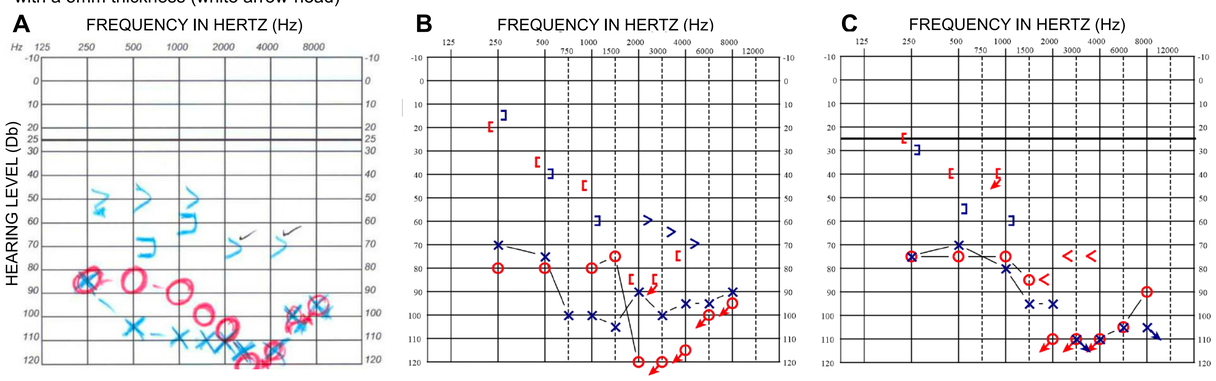
FINAL DIAGNOSIS
A. LEFT MASTOID (BIOPSY):
- Granulation tissue with marked acute and chronic inflammation.

B. LEFT MIDDLE EAR CONTENTS (EXCISION):
- Necrotizing granulomatous inflammation (see comment).

C. RIGHT MIDDLE EAR CONTENTS (EXCISION):
- Squamous epithelium with marked acute and chronic inflammation.

D. LEFT SURAL NERVE (BIOPSY):
- Peripheral nerve without evidence of acute injury and no definitive vasculitis (see comment).

Comment: The left middle ear contents consist of extensive necrotizing granulomatous inflammation. AFB and PAS special stains were performed to evaluate for the presence of mycobacteria and fungal organisms, respectively, and are negative (the controls stain appropriately). No definite vasculitis is identified. Overall, the findings are non-specific but could be compatible with granulomatosis with polyangiitis in an appropriate clinical setting. Correlation with clinical findings is recommended.



Ear	Tympanogram	SRT	WRS (%)	HL
R	Type B	90	60	110
L	Type B	105	48	110

Ear	Tympanogram	SRT	WRS (%)	HL
R	Type B	75	64	110
L	Type C	90	64	110

Ear	Tympanogram	SRT	WRS (%)	HL
R	Type B	75	36	110
L	Type B	80	24	110

Audiograms at time of presentation (A), after initial left FN decompression and tympanomastoidectomy (B) and after left OCR (C)
Initial audiogram with severe to profound mixed hearing loss (MHL) bilaterally with poor WRS. Bone conduction thresholds improved after initiation of therapy and left tympanomastoidectomy (B). The most recent audiogram showed severe to profound MHL on the right and moderate-severe to profound MHL on the left. Notably, audiogram C showed worsening WRS, suggestive of progressive vasculitis affecting sensorineural hearing.

Treatment Course

- At presentation started on 60mg daily prednisone for FN paralysis.
- 8 days after presentation left facial nerve decompression was performed.
- Findings at time of surgery:
  - Left: Dense granulation tissue filling middle ear space, adherent to ossicular chain and medial aspect of tympanic membrane. Granulation extending into round window niche. Facial nerve decompressed along tympanic segment and second genu, intact with response at 0.5mA.
  - Right: Fibrosed granulation along inferior and posterior mastoid bowl.
- Biopsies from left middle ear space resulted in diagnosis of GPA.
- Due to severe inflammatory disease, the (OCR) was deferred
- Started on rituximab 375mg/kg, continued prednisone
- Left OCR 7 months after first surgery with improved air-bone gap but gradually worsening sensorineural hearing outcomes, Right FN HB3 with synkinesis and crocodile tears. Left HB 2 at last exam

Discussion/Conclusions

- GPA is often an elusive diagnosis and can be associated with significant morbidity and mortality if diagnosis is delayed
- Bilateral facial nerve paralysis is a rare presenting symptom and in the absence of trauma should prompt workup of systemic causes of disease
- A multidisciplinary team should be utilized in workup and management

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