Subtotal Petrosectomy and Cochlear Implant Placement in Otologic Presentation of "Wegener's Granulomatosis"

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Citation

Elmas F, Shrestha BL, Linder TE. Subtotal Petrosectomy and Cochlear Implant Placement in Otologic Presentation of "Wegener's Granulomatosis". *Kathmandu Univ Med J* 2017;57(1):93-7.

ABSTRACT

Granulomatosis with polyangitis is a rare granuloma forming necrotizing vasculitis, which involves mainly the respiratory tract and renal system. Otologic involvement may occur primarily as chronic serous otitis media and chronic silent mastoiditis with conductive hearing loss and may rarely lead to sensorineural hearing loss requiring cochlear implantation.

This case describes a patient with granulomatous poylangitis with profound sensorineural hearing loss who underwent subtotal petrosectomy with cochlear implantation.

KEY WORDS

Cochlear implant, granulomatosis with polyangitis, subtotal petrosectomy, necrotizing vasculitis

INTRODUCTION

Wegener granulomatosis or as today named granulomatosis with polyangitis is a rare disease that is characterized by an idiopathic vasculitis of medium and small arteries leading to necrotizing granu lomatous inflammation of the respiratory tract and the renal system. Typically it presents with nasal and paranasal lesions, pulmonary and renal involvement.[1] It can present with otologic involvement causing serous otitis media, sensorineural hearing loss and vertigo.[2] In cases of severe and persistent sensorineural hearing loss, patients may qualify for a cochlear implantation (CI). This surgery faces two challenges: 1) finding the proper access to the round window or cochleostomy through a chronically inflamed mucosa with continuous bleeding and 2) implanting a foreign body (CI) into an inflamed environment risking tympanic membrane perforations as the disease may not be controllable. These patients need careful planning of the CI procedure and an adequate operation technique that eradicates the middle ear disease, avoids recurrence, prevents meningitis and allows secure placement of the cochlear implant electrodes.[3] We favor the Fisch technique of subtotal petrosectomy for cochlear implantation in this case as it provides all above mentioned properties and allows for safe and efficient implantation.[4]

CASE REPORTS

A 29 year old male presented in our outpatient clinic with a history of recurrent otitis media with purulent otorrhea and hearing loss. Clinical examination showed a thickened tympanic membrane with loss of anatomic landmarks

and a microperforation with discharge of secretion on valsalva maneuver. Flexible nasopharyngolaryngoscopy examination showed bilateral inflamed nasal mucosa with crusting. The audiogram revealed a progressive sensorineural hearing loss on both sides (Fig. 1a and b). The patient was treated initially with intravenous antibiotics and dexamethason. The bacteriology culture of the middle ear fluid showed no growth, especially no mycobacterium tuberculosis.

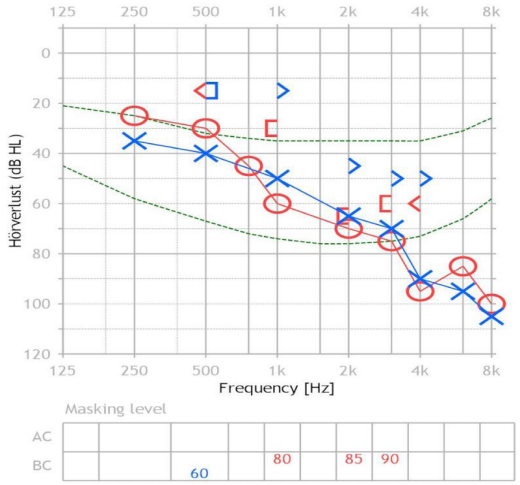

Figure 1a. Audiogram showing the hearing loss at first presentation.

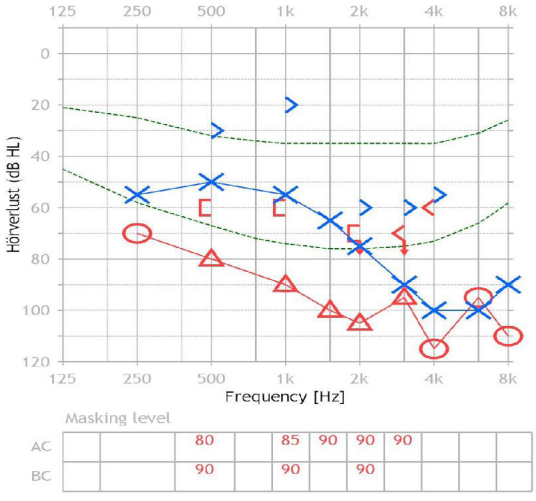

Figure 1b. Audiogram showing the progression of hearing loss 1 month after the first presentation.

Since the chronic suppurative otitis media and the endonasal crusting did not improve, the blood was investigated with suspicion of Wegener's granulomatosis. The results confirmed elevated cytoplasmic antineurophil cytoplasmic antibodies (cANCA) and Anti-PR3 (18 IU/ml.) antibodies. The chest X-ray and a urine investigation were normal. The rheumatologic assessment confirmed the diagnosis of a granulomatous polyangitis. A treatment with methotrexate was started. Meanwhile a subglottic stenosis was confirmed during endoscopic examination. The pneumologic assessment showed a light inspiratory stridor, the pulmonary function test was normal without a restriction or obstruction and with normal blood gas and CO_2 diffusion capacity. The patient improved during his hospital stay and was discharged.

After three months, the patient was again admitted with severe vertigo, spontaneous nystagmus to the left and positive Halmayagi test on the right. The right sided vestibular paresis was confirmed in the video head impulse test with saccades and reduced gain. The pure tone audiogram revealed a mixed hearing loss with progression of sensorineural component (fig. 2a and b). Treatment with dexamethason and betahistine was started and brought some relief of his vertigo symptoms.

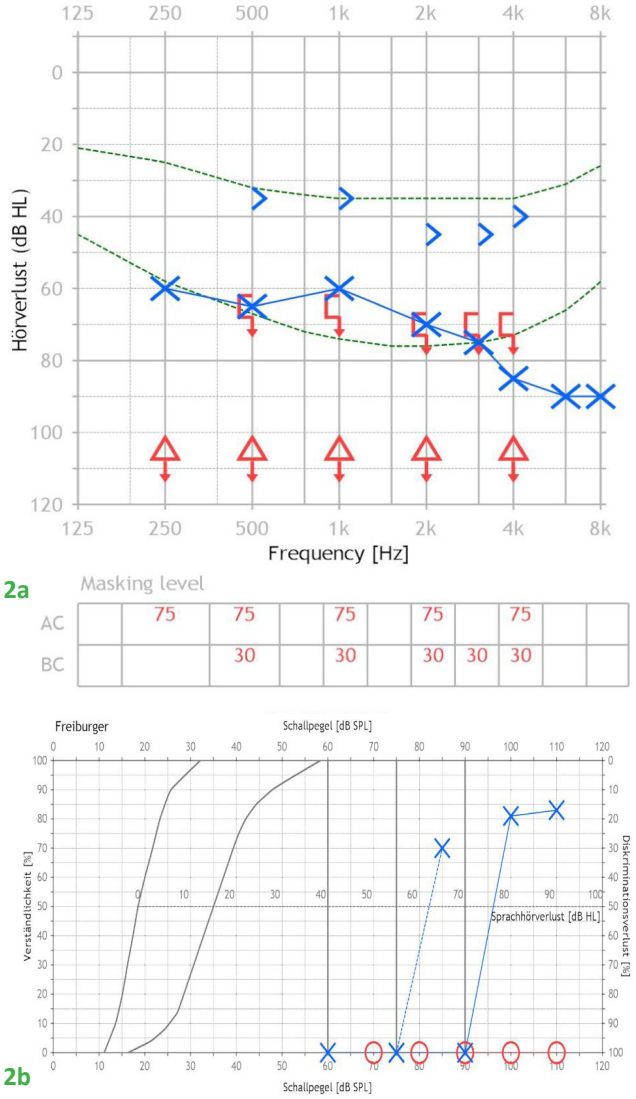

Figure 2a and b. Pure tone audiogram and speech audiometry showing profound sensorineural hearing loss right ear prior to cochlear implantation.

A high resolution CT scan of temporal bone showed the opacity in middle ear yet normal antrum and normal bony labyrinth (fig. 3). The gadolinium enhanced magnetic resonance imaging (MRI) further revealed an inflammatory lesion within the right middle ear and mastoid. The cochlea remained normal and fluid filled (fig. 4 and 5).

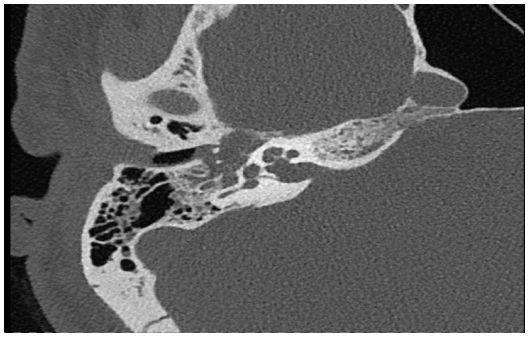

Figure 3. HRCT scan early in the disease process showing the opacity in middle ear and yet limited extension to the antrum

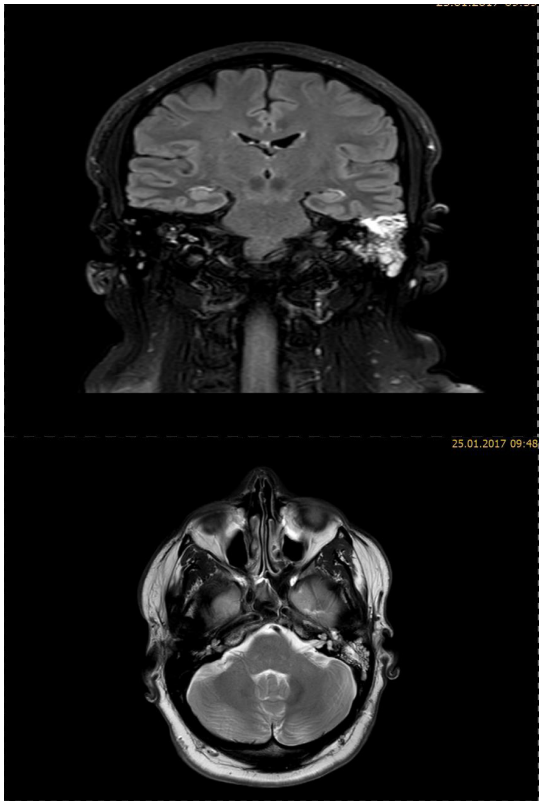

Figure 4 and 5. The gadolinium enhanced magnetic resonance imaging showing an inflammatory lesion within the right middle ear and mastoid. The cochlea remain normal and fluid filled.

A nasopharyngoscopy with biopsy of the nasal mucosa was performed which showed a granulomatous inflammation with necrosis in the septum mucosa, and focal inflammation of the nasopharynx. A Treponema pallidum hamagglutination assay (TPHA) was negative. The rheumatologist increased the dose of the steroid therapy and started a therapy with infliximab. Due to the rapid deterioration of his hearing bilaterally, the patient was scheduled for a right side CI. A conventional anterior mastoidectomy with posterior tympanotomy would lead to difficulties at surgery with continuous bleeding, insufficient landmarks and possible improper placement of the electrodes. The continuously draining perforation was a risk for infection to the implanted ear even after a myringoplasty due to the chronicity of the polyangitis. Therefore the patient underwent a right sided subtotal

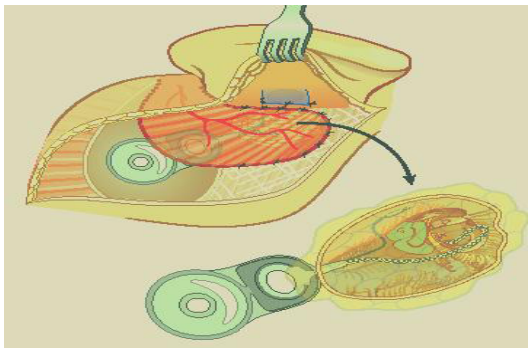

Figure 6. Schematic Figures depicting Subtotal Petrosectomy with Cochlear implant.

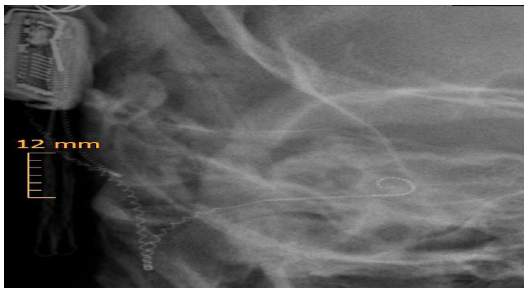

Figure 7. Post-operative X-ray Stenvers view showing good placement of cochlear implant electrode.

petrosectomy with abdominal fat obliteration and simultaneous cochlear implantation (Cochlear nucleus slim modiolar CI532) (fig. 6 schematic figures). The post-operative course was uneventful. The post-operative x-ray stenver's view showed a correctly placed cochlear implant as shown in fig 7. As early as 2 months after implantation, the patient regained successful speech understanding and is still under rehabilitation (fig. 8a and b).

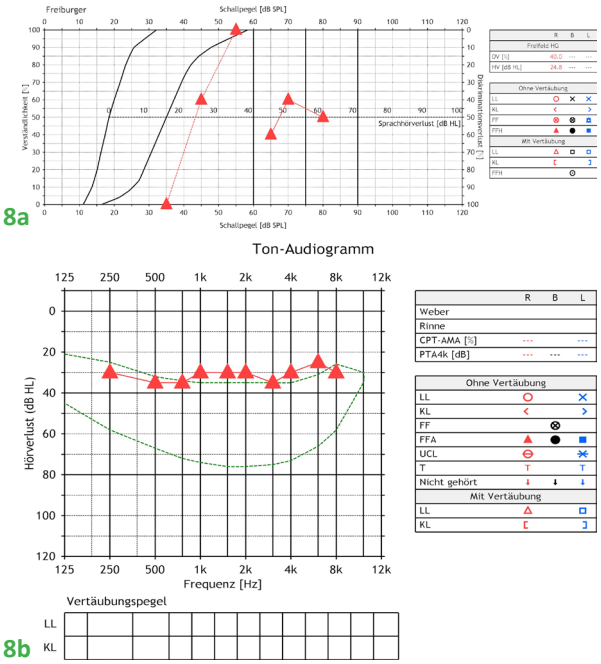

Figure 8a and b Post-operative audiogram and speech audiometry showing hearing recovery with cochlear implantation on the right side.

DISCUSSION

Wegener's granulomatosis or as today named granulomatosis with polyangitis is a rare disease that is characterized by an idiopathic vasculitis of medium and small arteries leading to necrotizing granulomatous inflammation of the respiratory tract and the renal system. The disease is serious and can lead to renal or lung failure with a mean survival time of 5 months.

Typically it presents with nasal and paranasal lesions, pulmonary and renal involvement. Otological manifestations are present in nearly half of the cases and consist mostly of serous otitis media and eustachian tube blockage with nasopharyngeal involvement, mastoid opacification on CT scan by granulomatosis of the mucosa and may lead to sensorineural hearing loss due to inflammation of cochlear vessels.[1,5]

Wegener's granulomatosis is diagnosed by clinical features and investigations such as high erythrocytic sedimentation rate, positive cytoplasmic antineutrophil cytoplasmic antibodies (C-ANCA) test, proteinuria, abnormal serum creatinine level, anemia and biopsy from nasal mucosa, middle ear mucosa or even renal biopsies. Diagnosis is made based on the Criteria of the American College of Rheumatology. Treatment typically consist of two phases. In the induction phase a combination of systemic corticosteroid and immunosuppressant is used and the maintenance phase consist of supplementation with corticosteroids and azathioprine/methotrexate.[6,7]

Patients with severe hearing loss/near deafness without recovery under these treatments should be evaluated for possible cochlear implantation. The surgery in these cases is challenging due to the ongoing inflammation, tympanic membrane perforations, severe and continuous bleeding from the inflamed mucosa. In addition, a foreign body will be implanted into the inflamed environment. Recurrent tympanic membrane perforations or ascending nasopharyngeal infections carry the risk of implant infection and biofilm formation. Also the need of opening the inner ear through a cochleostomy or via the round window membrane creates potential routes of spread of infection to subarachnoid spaces and meningitis. Therefore cochlear implant candidates with Wegener's granulomatosis as any other patient with chronic otitis media require special attention and management. The primary goal of surgery besides hearing rehabilitation must be the eradication of the middle ear disease to avoid recurrenceand meningitis and prevent implant infection.[8,9]

Different techniques for cochlear implantation in patients with inflamed or infected middle ear where described in the literature as summarized by Szymanski et al.[3] Some authors suggest to wrap the electrodes with patients own tissues to prevent extrusion. Other authors suggest reconstruction of the tympanic membrane and posterior canal wall.[10,11] A different approach tries to prevent extrusion by putting the electrode insertion away from the diseased middle ear. This was achieved by a transcanal approach, subfacial approach or a bony groove drilled into the posterior canal wall.[11-13] But all of these techniques don't solve the underlying problem, the ear still remains inflammatious and therefore has the risk of recurrence and implant infection. Only the subtotal petrosectomy technique as described by Fisch and Mattox provides a maximum exposure of the temporal bone for elimination of the middle ear disease and therefore minimal risk of recurrent disease.[4] It involves eradication of all accessible pneumatic spaces in the temporal bone, removal of the middle ear mucous membrane, tympanic membrane, skin of the external ear canal with closure of eustachian tube and external ear canal. It is used during all infratemporal fossa approaches A-C to remove skull base pathologies, but also serves as a treatment for persistent chronic otitis media or temporal bone tumors. In our case it allows to perform the implantation of the electrodes into an inflammation free surgical field. The use of subtotal petrosectomy for CI was first described by Bendet et al. for patients with chronic discharging ear and profound hearing loss.[13] Regarding CI performance patients with CI and subtotal petrosectomy have the same functional outcome as healthy CI patients.[14]

Wegener's granulomatosis is a rare disease that can lead to severe sensorineural hearing loss. For this entity cochlear implantation with subtotal petrosectomy is a safe and efficient treatment option. It provides hearing rehabilitation and cleaning of the inflamed middle ear at the same time.

REFERENCES

1. Srouji IA, Andrews P, Edwards C, Lund VJ. Patterns of presentation and diagnosis of patients with Wegener's granulomatosis: ENT aspects. *J Laryngol Otol.* 2007 Jul; 121(7):653-8.
2. Kim SH, Jung AR, Kim SI, Yeo SG. Refractory Granulomatosis with Polyangiitis Presentingas Facial Paralysis and Bilateral Sudden Deafness. *J Audiol Otol.* 2016;20(1):55-8.
3. Szymański M, Ataide A, Linder T. The use of subtotal petrosectomy in cochlearimplantc and idates with chronicotitis media. *Eur Arch Otorhinolaryngol.* 2016 Feb;273(2):363-70.
4. Fisch U, Mattox D. Micro surgery of the Skull Base. Stuttgart – New York: Georg Thieme Verlag; 1988.
5. Takagi D, Nakamaru Y, Maguchi S, Furuta Y, Fukuda S. Otologicmanifestations of Wegener's granulomatosis. *Laryngoscope.* 2002 Sep;112(9):1684-90.
6. Wojciechowska J, Krajewski W, Krajewski P, Kręcicki T. Granulomatosis With Polyangiitis in Otolaryngologist Practice: A Review of Current Knowledge. *Clin Exp Otorhinolaryngol.* 2016;9(1):8-13.
7. Erickson VR, Hwang PH. Wegener'sgranulomatosis: current trends in diagnosis and management. *Curr Opin Otolaryngol Head Neck Surg.* 2007 Jun;15(3):170–6.
8. Schlöndorff G, Hermes H, Weck L. Cochlearimplants in patients with radicalcavity. *HNO*. 1989 Oct; 37(10):423-5.

9. Manrique M, Cervera-Paz FJ, Espinosa JM, Perez N, Garcia-Tapia R. Cochlearimplantation in radicalcavities of mastoidectomy. *Laryngoscope.* 1996 Dec; 106(12):1562-5.

10. Tamura Y, Shinkawa A, Ishida K, Sakai M. Cochlearimplant after reconstruction of the external bonycanal wall and tympaniccavity in radically mastoidectomized patients withc holesteatoma. *Auris Nasus Larynx.* 1997 Oct; 24(4):361-6.

11. Olgun L, Batman C, Gultekin G, Kandogan T, Cerci U. Cochlearimplantation in chronicotitis media. *J Laryngol Otol.* 2005 Dec; 119(12):946-9.

12. Colletti V, Fiorino FG, Carner M, Pacini L. Basal turn cochleostomy via the middle fossa route for cochlearimplantinsertion. *Am J Otol.* 1998 Nov; 19(6):778-84.

13. Bendet E, Cerenko D, Linder TE, Fisch U. Cochlearimplantation after subtotal petrosectomies. *Eur Arch Otorhinolaryngol.* 1998; 255(4):169-74.

14. Vincenti V, Pasanisi E, Bacciu A, Bacciu S, Zini C. Cochlearimplantation in chronicotitismedia and previous middle ear surgery: 20 years of experience. *Acta Otorhinolaryngol Ital*. 2014 Aug; 34(4):272-7.