tomography (CT) and magnetic resonance imaging (MRI) may show no changes or reveal nonspecific abnormalities, usually cerebral atrophy, diffuse subcortical abnormality, and nonspecific subcortical focal white matter abnormality.[2]

Treatment of patients with HE is based on the use of corticosteroids. Most commonly, intravenous IV methylprednisolone (500–1000 g/day) is given for three to five days followed by oral prednisone taper.[4] When deciding optimal treatment strategy regarding corticosteroid regimen, proper balance between patient's condition and multiple factors contributing to serious side effects of corticosteroid use (e.g., age, nutritional status, comorbidity) should be taken into consideration. Antithyroid antibodies levels do not correlate well with the clinical picture of HE and do not reflect response to treatment.[4] Therefore, when evaluating patient's condition, physicians must rely solely on patient's symptoms and clinical course monitoring. In the presented case, it is likely possible that immunosuppression was induced by corticosteroid treatment. Unfavorable outcome in HE is not entirely unexpected but there is a lack of published data demonstrating which risk factors are more associated with it.

Conclusion

This case provides more insight in the diversity of clinical manifestations of HE. Acute rigidity and markedly elevated levels of CK could share common pathological process in HE. It is still unclear and yet to be determined whether the symptomatology of HE is consequence of abnormal autoimmune response. Further investigations are required to establish the optimal therapeutic approach for patients with HE.

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References

1. Brain L, Jellied EH, Ball K. Hashimoto's disease and encephalopathy. Lancet. 1966;2:512-14.
2. Chong JY, Rowland LP, Utiger RD. Hashimoto encephalopathy: syndrome or myth? Arch Neurol. 2003;60:164-71.
3. Castillo P, Woodruff B, Caselli R, et al. Steroid-responsive encephalopathy associated with autoimmune thyroiditis. Arch Neurol. 2006;63(2):197-202.
4. de Holanda NC, de Lima DD, Cavalcanti TB, Lucena CS, Bandeira F. Hashimoto's encephalopathy: systematic review of the literature and an additional case. J Neuropsychiatry Clin Neurosci. 2011;23(4):384-90.
5. Ryan SA, Kennedy C, Harrington HJ. Steroid-responsive encephalopathy associated with autoimmune thyroiditis presenting as confusion, dysphasia, and myoclonus. Case Report Med. 2012;2012:782127.

To the Editor

Recovery from Deafness in the Contralateral Ear of Surgery in NF 2 Patient

Hearing loss due to vestibular schwannoma (VS) is one of the significant morbidities in patients with neurofibromatosis type 2 (NF 2). Advances in surgical techniques and intraoperative neurophysiological monitoring have enabled hearing preservation in surgeries for VS. Rarely, recovery from deafness can happen in some cases. Most of the reports for hearing improvements have occurred in the same area of the surgical procedure and were limited to cases with sudden hearing loss.[1] In this report, we present an unusual case of hearing improvement in the contralateral side of surgery in a NF 2 patient with bilateral vestibular schwannoma and deafness in both ears.

Case Report

A 23-year-old woman diagnosed with neurofibromatosis type NF 2 presented to our department at National Neuroscience Institute (NNI) in Saudi Arabia with a two month history of progressive bilateral hearing loss, gait unsteadiness, and blurred vision. On physical examination, there was right eye ptosis and partial ptosis of the left eye. She had grossly diminished hearing in both ears, with normal tympanic membranes. Brain and spine magnetic resonance imaging (MRI) showed findings consistent with NF 2. There were bilateral VSs, multiple intracranial and spinal meningiomas, and cauda equina schwannomas (Figure 1;A,B). The right large VS was causing a mass effect on the fourth ventricle with consequent third and bilateral ventricular dilatation.

Preoperative audiology assessment showed normal middle ear pressure and compliance. Ipsilateral and contralateral acoustic stapedial reflexes were absent bilaterally. Distortion product otoacoustic emissions (DPOAE) testing was normal in both ears. Pure tone audiometry (PTA) revealed moderate to severe sensorineural hearing loss in the left ear and profound sensorineural hearing loss in the right ear (Figure 2A). Speech recognition thresholds were consistent with the pure tone findings bilaterally. Although the patient can hear the tones, her speech recognition ability was significantly impaired in the left ear.

Patient underwent right suboccipital craniotomy and excision of the right VS with external ventricular drain (EVD) insertion. Her somatosensory evoked potentials, auditory brainstem responses and facial nerve function were monitored intraoperatively. Histopathology of the tumor revealed a benign schwannoma. After surgery, the patient was transferred to the intensive care unit. Postoperative brain MRI (Figure 1;C,D), showed normal postoperative findings and near total resection of right VS. The left VS and other intracranial lesions remained unchanged. Subjective improvement in hearing from the left ear (contralateral side) was noted immediately by the patient.

https://doi.org/10.1017/S0317167100017741 Published online by Cambridge University Press

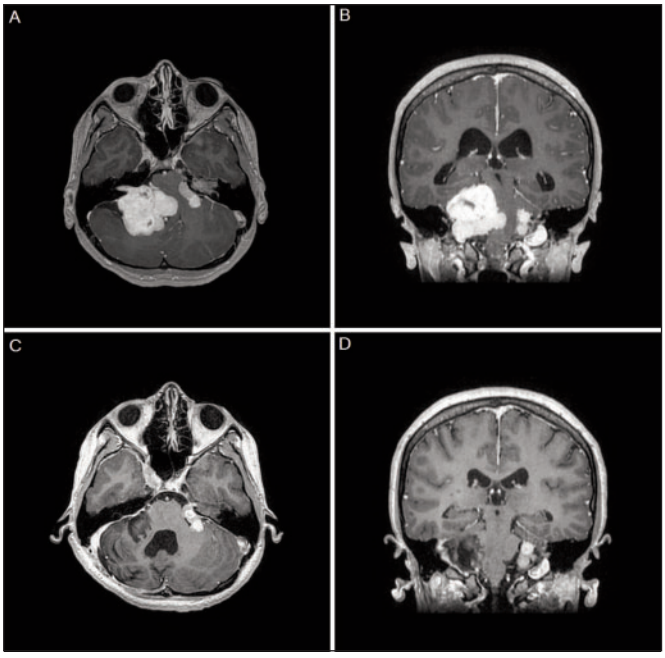

Figure 1: *Axial (A) and coronal (B) enhanced T1-weighted magnetic resonance imaging scans demonstrating the presence of two cerebellopontine angle tumors (4.4 x 4.2 cm on the right compared with 2.3 x 1.8 cm on the left). (C) and (D), postoperative scans showing near total resection of the right vestibular schwannoma with no evidence of complications.*

After two months, PTA demonstrated considerable improvement in hearing sensitivity at 1000 Hz (Figure 2 B). Moreover, assessments at six month follow-up showed significantly better hearing at all frequencies above 500 Hz and improved speech recognition abilities (Figure 2 C). Her postoperative audiometry results for the right ear were almost similar to the preoperative findings during all follow-up visits.

Discussion

NF 2 is a rare autosomal dominant genetic disorder that affects 1 in 33,000 persons at birth. It is characterized by the presence of multiple schwannomas, meningiomas, and ependymomas in the nervous system. The hallmark of the disease are bilateral VSs. They can present with symptoms caused by intracranial, spinal or cutaneous tumors. These multiple tumors can lead to cranial nerves dysfunction, myelopathies, and brainstem compression. Although there is a clear germline genotype-phenotype associations, NF 2 is a heterogonous disease in clinical practice and symptomatology varies greatly. The Manchester diagnostic criteria is a useful method to diagnose NF 2 clinically.[2]

The course of hearing loss in NF 2 population is particularly important because bilateral deafness is often inevitable. Most patients with bilateral VS present with progressive unilateral hearing loss in the second and third decades of life. No direct genotype–phenotype association for hearing loss was found in NF 2, and no predictors for the hearing status have been identified. However, hearing in the unoperated ear is likely to remain stable for up to two years.[2]

The exact mechanism underlying hearing loss in VS is still uncertain. Vascular insult or mechanical compression of the cochlea and the eighth nerve are possible causative factors. Sudden or fluctuating hearing losses can be attributed to compression or spasm of the labyrinthine artery, an end artery, or its branches in the internal auditory canal (IAC). Beside sudden ischemic events, cochlear aperture obstruction leading to elevated intralabyrinthine protein and impaired cochlear nerve axonal transport is a recent explanation of the slow progressive hearing loss in NF 2.[3]

Hearing improvement after VS surgery is possible. Most of the improvement' reports in the literature occurred in the ipsilateral ear of surgery.[1] It is rare to see this event in the contralateral ear in cases of bilateral VS. Samii and his colleagues[4] reported only 2.4% of "some hearing" on the nonoperated side in their series of 120 VS in patients with NF 2.

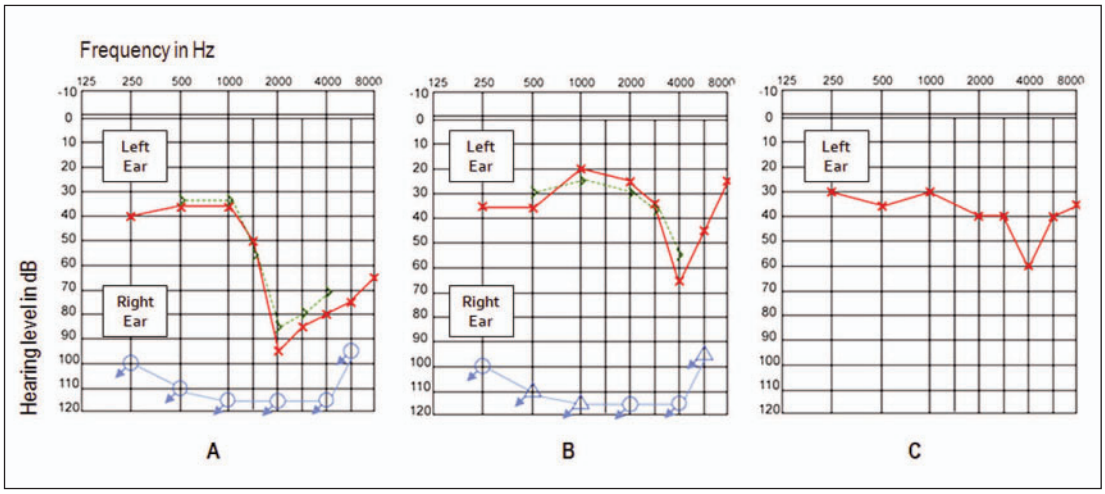

Figure 2: *Pure tone audiometry. (A) Preoperative. (B) Two and (C) six months postoperatively.*

https://doi.org/10.1017/S0317167100017741 Published online by Cambridge University Press

Our patient recovered hearing in the contralateral ear and had better PTA results after resection of the large right VS that was causing a mass effect on the adjacent structures. To our knowledge, there is only one report of an analogous case in the English literature. Farrell et al[5] reported an immediate post-operative improvement of 45 dB in the contralateral ear of a 32-year-old female who had similar baseline auditory functions to our case. However, their case did not include long term follow-up. We believe that surgery played a major role in our patient's recovery as a result of indirect decompression of the cochlear fibers. Decrease in cerebrospinal fluid volume and resolution of endolymphatic hydrops after surgery is another possible explanation.

Management of tumors in NF 2 remains challenging and controversial. Surgical removal of symptomatic lesions is the mainstay of management. However, this option frequently leads to iatrogenic hearing loss in NF 2 cases. This is due to the multifocal nature of VS in NF 2. Radiosurgery and chemotherapy are available alternatives for poor surgical candidates. Definitely, bilateral VS in NF 2 patients requires a multidisciplinary approach and must be tailored to the individual patient.

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References

1. Kawaguchi T, Tanaka R, Kameyama S, Yamazaki H. Full recovery from deafness after removal of a large acoustic neurinoma associated with neurofibromatosis 2: case report. Surg Neurol. 1994;42(4):326-9.
2. Asthagiri AR, Parry DM, Butman JA, et al. Neurofibromatosis type 2. Lancet. 2009;373(9679):1974-86.
3. Asthagiri AR, Vasquez RA, Butman JA, et al. Mechanisms of hearing loss in neurofibromatosis type 2. PLoS One. 2012;7(9): e46132.
4. Samii M, Matthies C, Tatagiba M. Management of vestibular schwannomas (acoustic neuromas): auditory and facial nerve function after resection of 120 vestibular schwannomas in patients with neurofibromatosis 2. Neurosurgery. 1997;40(4): 696-705; discussion 705-696.
5. Farrell ML, Harries ML, Baguley DM, Moffat DA. Bilateral acoustic schwannoma: post-operative hearing in the contralateral ear. J Laryngol Otol. 1991;105(9):769-71.

https://doi.org/10.1017/S0317167100017741 Published online by Cambridge University Press