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CASE REPORT

Cochlear implantation in an adult woman with Takayasu's arteritis: a case report

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Abstract

Background: Takayasu's arteritis (TAK) is an idiopathic large-vessel vasculitis. Sensorineural hearing loss is a rare complication in patients with TAK. In this study, we report an adult woman with hearing loss associated with TAK who underwent cochlear implantation (CI).

The Case: The case was a 28-year-old hearing-impaired woman with TAK who underwent unilateral CI. The surgery improved the patient's speech perception and perceived sound quality. However, her auditory and speech performances changed over time.

Conclusion: The present case report highlights the importance of monitoring auditory and speech performance of CI patients with TAK.

Keywords: Cochlear implantation; Takayasu arteritis; hearing loss

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Introduction

Takayasu's arteritis (TAK) is an idiopathic large-vessel vasculitis, which predominantly involves the aorta and its proximal branches. Its overall incidence has been estimated to be 1−2 per million people and mostly occurs in young or middle-age Asian women [1,2]. Clinical manifestations of the TAK are various, depending on the time point along the disease course, including cardiovascular (e.g. hypertension), cutaneous (e.g. subcutaneous edema or ulcers), gastrointestinal (e.g. abdominal pain), ocular (e.g. retinal damage), or neurological (e.g. headache or seizure) [2]. Sensorineural hearing loss (SNHL) has been reported as a rare neurological complication in patients with TAK. It has been suggested that hearing loss following TAK may be persistent or fluctuating, or can be manifested as gradual or sudden SNHL [3-5]. Cochlear implant (CI) is widely used to treat patients with bilateral severe-to-profound SNHL who no longer benefit from hearing aids. It has been demonstrated that CI individuals typically experience improved speech intelligibility and sound localization abilities [6,7]. In this study, we report an adult woman with hearing

Table 1. Audiological characteristics of a woman with Takayasu's arteritis across different time intervals before cochlear implantation

Parameter	Time of assessment			
	On admission (26.12.2016)	**3-m post-admission (3.3.2017)**	**6-m post-admission (20.6.2017)**	**12-m post-admission (7.1.2018)**
Chief complaint	Hearing loss, tinnitus	Hearing loss, tinnitus, nausea/vomiting	Hearing loss, tinnitus, true vertigo	Hearing loss, tinnitus
Tympanogram	Type An	Type An	Type An	Type An
PTA (dB HL)				
	RE: 64.34	RE: 86.33	RE: 89.71	RE: 88.55
	LE: 72.13	LE: 78.65	LE: 85.13	LE: 90.14
WRS (%)				
	RE: 76	RE: 64	RE: 68	RE: 60
	LE: 72	LE: 68	LE: 64	LE: 60

m; month, PTA; pure tone average, RE; right ear, LE; left ear, WRS; word recognition score

loss associated with TAK who underwent unilateral cochlear implantation.

Case presentation

This is a case report approved by the Ethics Committee of Ahvaz Jundishapur University of Medical Sciences, Ahvaz, Iran (Ethics code: IR.AJUMS.1400.239). The patient was a 28-year-old woman with TAK who received consultation in our otology clinic on December 18, 2016, complaining of high-pitched tinnitus and hearing loss in both ears in the past three months. Audiological assessments showed a bilateral severe SNHL, and a slight difficulty in word recognition ability. We found no evidence of joint disease. An ophthalmologic examination demonstrated no evidence of uveitis or interstitial keratitis. Visual acuity was within a normal range. The TAK in this patient was diagnosed according to American College of Rheumatology criteria [8] and based on the onset of asthenia, asymmetry of radial artery pulsation, sideropenic anemia, and amaurosis fugax. Laboratory examinations showed increased erythrocyte sedimentation level (42 mm/h) and C-reactive protein (3.29 mg/dL) markers. The plasma levels of IgG (1356 mg/dL), IgM (104 mg/dL) and IgA (267 mg/dL) were within a normal range. Rheumatoid (Rh) factor and other autoantibodies were negative. Syphilis serology was also negative. The epiaortic ultrasound test revealed the significant stenosis of the right subclavian artery and left common carotid artery and the nonsignificant stenosis of the mesenteric artery and abdominal aorta. Diagnosis of TAK was also confirmed by the magnetic resonance angiography approach. High-dose methotrexate (15 mg/day) and steroid therapy (50 mg/day prednisone) were administered. A progressive stabilization of the clinical and laboratory findings was then observed during a 4-year follow-up. The patient reported improvement in hearing after the first admission session, but her hearing thresholds were worsened gradually during the follow-up period. Table 1 shows the audiological results during the admission and follow-up periods.

Cochlear implant mapping

The patient underwent unilateral cochlear implantation (MED-EL Mi1200 Synchrony) in May 2018 and participated in regular follow-up sessions. The CI system breaks the input signal into different frequency bands, each attributed to a channel. The acoustical dynamic range (DR) in each band is mapped into the electrical DR characterized by two reference electrical thresholds associated with a corresponding channel, known as the electrical threshold (THR) and the most comfortable level (MCL). The accurate estimation of the THR and MCL is very important for a

Table 2. Cochlear implant mapping parameters at different time intervals after cochlear implant surgery

		Channel number											
Date of CI mapping	**Parameter**	**1**	**2**	**3**	**4**	**5**	**6**	**7**	**8**	**9**	**10**	**11**	**12**
1-month post-CI (19.6.2018)	**Impedance**	OK	OK	OK	OK	OK	OK	OK	OK	OK	OK	OK	OK
	MCL (qu)	99.87	86.41	88.29	93.67	93.23	98.4	75.61	99.16	105.1	111.6	108.0	109.5
	THR (qu)	9.87	8.64	8.83	9.37	9.32	9.84	7.56	9.91	10.52	11.16	10.81	10.95
	CF (Hz)	162	303	502	778	1153	1693	2430	3475	5214	7302	0	0
	Chanel Status	Enab	Enab	Enab	Enab	Enab	Enab	Enab	Enab	Enab	Enab	**Disab**	**Disab**
6-month post-CI (8.12.2018)	**Impedance**	OK	**High**	OK	OK	**High**	OK	OK	OK	OK	**High**	OK	OK
	MCL (qu)	79.08	40.39	66.23	76.78	39.88	78.3	78.51	78.74	86.36	34.97	53.13	100.2
	THR (qu)	7.91	4.04	6.62	7.68	3.99	7.83	7.85	7.87	8.64	3.50	3.41	10.02
	CF (Hz)	189	0	405	793	0	1412	2447	4158	7230	0	0	0
	Chanel Status	Enab	**Disab**	Enab	Enab	**Disab**	Enab	Enab	Enab	Enab	**Disab**	**Disab**	**Disab**
9-month post-CI (5.3.2019)	**Impedance**	OK	**High**	OK	OK	OK	OK	OK	OK	OK	OK	OK	OK
	MCL (qu)	89.08	42.39	66.23	76.78	42.31	78.3	78.51	78.74	86.36	34.97	65.49	106.5
	THR (qu)	8.91	4.23	6.62	7.68	4.23	7.83	7.85	7.87	8.64	3.50	6.54	10.65
	CF (Hz)	155	0	516	769	0	1692	2429	3485	5315	7375	0	0
	Chanel Status	Enab	**Disab**	Enab	Enab	**Disab**	Enab	Enab	Enab	Enab	Enab	**Disab**	**Disab**
12-month post-CI (12.6.2019)	**Impedance**	OK	OK	OK	OK	OK	OK	OK	OK	OK	OK	OK	OK
	MCL (qu)	87.83	70.26	71.79	76.16	75.80	82.4	77.88	83.07	96.27	99.19	104.9	104.7
	THR (qu)	8.78	7.03	7.18	7.62	7.58	8.24	7.79	8.31	9.63	9.92	10.49	10.47
	CF (Hz)	142	277	479	752	1133	1605	2444	3356	5265	7462	0	0
	Chanel Status	Enab	Enab	Enab	Enab	Enab	Enab	Enab	Enab	Enab	Enab	**Disab**	**Disab**
18-month post-CI (29.2.2019)	**Impedance**	OK	OK	OK	OK	OK	OK	OK	OK	OK	OK	OK	OK
	MCL (qu)	95.06	79.08	80.80	85.72	85.32	92.7	87.65	93.49	98.35	99.64	94.93	106.33
	THR (qu)	9.51	7.91	8.08	8.57	8.53	9.28	8.77	9.35	9.83	9.65	9.43	10.63
	CF (Hz)	142	277	479	760	1133	1692	2429	3474	5214	7302	0	0
	Chanel Status	Enab	Enab	Enab	Enab	Enab	Enab	Enab	Enab	Enab	Enab	**Disab**	**Disab**

CI; cochlear implant, OK; channel impedance ≤ 20 KΩ, MCL; maximum comfortable level, THR; electrical threshold, CF; center frequency, Enab; enabled (active) channel, Disab; locally disabled channel, High; channel impedance > 20 KΩ

successful CI fitting. Table 2 shows the CI programming parameters at different time points.

At the first post-operative CI mapping session, all channels showed normal impedances, and the

Table 3. Changes in clinical characteristics of a woman with Takayasu's arteritis after unilateral cochlear implantation

	Time of cochlear implant mapping				
Parameter	**1-month post-CI**	**6-month post-CI**	**9-month post-CI**	**12-month post-CI**	**18-month post-CI**
HISQUI$_{19}$	67	48	53	73	78
WRS (%)	72	36	42	64	76

CI; cochlear implant, HISQUI; hearing implant sound quality index, WRS; word recognition score

THR and MCL (except for two channels) were within a normal range. However, 6 and 9 months after CI mapping, we noticed a change in the THR associated with TAK such that 5 and 4 channels had been disabled, respectively. The number of disabled channels reduced to 2 when the patient was referred to the clinic again in the following months (Table 3).

Subjective assessment of the cochlear implant function

The speech perception ability was measured by an open-set test using monosyllabic words in a sound-field acoustic room. The test was conducted without lip-reading, at 65 dB SPL in a quiet environment, and with a signal-to-noise ratio of 10 dB SPL lower than the signal level. The signals were presented through a calibrated loudspeaker at 0-degree azimuth, placed at 1-m distance from the patient. The hearing implant sound quality index (HISQUI19) questionnaire was used to measure the patient's auditory performance in everyday listening situations [9]. The HISQUI19 contains 19 items scored on a 7-point Likert scale from 1 = Never to 7 = always. The total score ranges from 19 to 133, where higher scores indicate better performance in using the CI device.

Our findings suggested that the number of active (enabled) channels has a significant impact on the CI function. At the first post-operative session, the speech perception score was 72% which was reduced to 36% and 44 %, when 5 and 4 channels of the CI system were disabled, respectively. The total HISQUI19 score at the first post-operative session was 67, indicating perceived moderate sound quality. However, the score was reduced to 48 and 53 (perceived poor sound quality) when 5 and 4 channels of the CI system were disabled, respectively.

Discussion

In the current study, we reported a woman with confirmed TAK disease who had undergone unilateral cochlear implantation. Before surgery, she had a bilateral severe-to-profound SNHL complaining of fluctuating hearing loss. Cochlear implantation led to obvious improvement in her speech and language skills; however, this improvement was not stable and she frequently reported a detorioration or improvement in her listening skills over time. Our results showed that by the increase in the number of disabled channels, the speech perception and perceived sound quality were deteriorated substantially. Her perceived sound quality was at a moderate level at the first post-operative session, but it became poor when 5 and 4 channels of the CI device were disabled.

It has been indicated that SNHL following TAK commonly occurs in women in their fourth and fifth decades of life, which is usually manifested as a gradual hearing impairment in both ears [10]. The etiology of hearing loss associated with TAK is still unclear. Different mechanisms have been proposed to explain the pathogenesis of TAK-related hearing loss. It has been shown that TAK influences medium- and small-size arteries and induce thrombosis in the labyrinthine artery [4,10]. Murofushi et al. [11] reported that intratympanic injection of steroids in TAK subjects might result in vasodilation along with increased

microvascular blood flow in the cochlea. They suggested that the increased cochlear blood flow can prevent or reduce irreversible alterations in the inner and outer hair cells' function, and lead to hearing recovery [11]. However, temporal bone histopathological findings of Nomura and Kitamura [12] did not support this claim. They observed that blood vessels in the internal auditory meatus and cochlea were normal in a patient with TAK. Another proposed mechanism for TAK-related hearing loss is the immune-mediated inflammatory response in the membranous labyrinth [10,13,14]. This suggestion is supported by the fact that corticosteroid therapy can improve hearing sensitivity in patients with TAK. According to the results, it can be speculated that the combination of autoimmune (elevation of serum immune complexes) and vascular factors can be responsible for the pathogenesis of hearing loss in TAK patients. The current case report is the first study that provides information about the alterations in CI function of TAK patients. The results can be useful in otology and audiology for monitoring the auditory function of affected patients.

Conclusion

The sensorineural hearing loss resulted from Takayasu's arteritis (TAK) may be progressive and fluctuating during the treatment course, and a severe-to-profound degree of hearing loss following the disease may persist in spite of medical therapy. The cochlear implant can improve the speech perception and perceived sound quality in patients with TAK. However, the auditory and speech performances may prominently change over time. The current case report highlights the importance of monitoring auditory function in cochlear implant patients with TAK.

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Conflict of interest

The authors declare no conflicts of interest.

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