Hemangioma of Internal Auditory Canal: Systemic Review

Omar Ramadan*

Department of ENT, USA

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*Corresponding author: Omar Ramadan, Independent researcher, ENT registrar, Paterson NJ, New Jersey, USA

Abstract

Objective: The objective of this study was to present a review article about internal auditory canal hemangioma.

Data Sources: Published English-language literatures.

Review Methods: PubMed and Google scholars were systematically searched using multiple search terms.

Study Selection: We included studies about internal auditory canal hemangioma.

Results: 42 studies were included in this study. The results showed that most patients were male, the age of patients varied between 5 to 69 years old. Two cases were multiple familial hemangioma, while the most other cases were sporadic. Hearing loss was the most common symptoms, 37% patients had facial palsy, and 38% patients had vestibular symptoms. Since most patients had non-serviceable hearing loss so, translabyrinthine approach was the most common surgical approach. Most serviceable hearing patients lost their serviceable hearing after surgery. Only one third of patients with facial palsy had improvement after surgery. Most cases had attachment to facial nerve and can be removed totally.

Conclusion: we should suspect IAC hemangioma in any patient with considerable hearing loss and facial palsy with small lesion in internal auditory canal.

Keywords: IAC: Internal Auditory Canal; Cavernous hemangioma; Facial spasm; Translabrynithin

Abbreviations: TL: Translabrynthine; RS: Retrosigmoid; MFA: Middle Fossa Approach; IAC: Internal Auditory Canal; TR: Total Removal; NTR: Near Total Removal; HL: Hearing Loss; FP: Facial Palsy

Introduction

Cavernous hemangioma of the internal auditory canal (IAC) is a rare disease. It comprises 10 to 20% of all central nervous vascular malformations. They are composed of large, sinusoidal, thin-walled capillary spaces that may invade the surrounding neural tissue. They can mimic the symptoms of vestibular schwannomas. We review the clinical features and the management of the IAC hemangioma [1].

Material and Methods

Literature review was conducted using PubMed (MEDLINE) and Google Scholar for English articles, the following keywords were used: internal; auditory; canal and hemangioma.

Inclusion Criteria

All internal auditory canal hemangioma articles published after 1975 were included in the study.

Results

Forty-four studies about IAC hemangioma have been reported in PubMed (MEDLINE) and Google scholars in English literatures (Table 1).

Table 1: IAC Hemangioma report cases.

| Article | Age | sex | symptoms | Surgical approach | Intraoperative Finding nerve attachment | Removal extension CPA | Postoperative Hearing profile | Postoperative Facial palsy |
|---------|-----|-----|----------|-------------------|----------------------------------------|-----------------------|-------------------------------|--------------------------|

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| Author(s) | Gender | Age | Clinical Presentation | Auditory Canal | Facial Nerve | NTR/改善 | Hearing Status | Facial Palsy |
|-----------|--------|-----|-----------------------|----------------|-------------|---------|---------------|-------------|
| Sundarsean et al. [1] | 23 M | 50 M | Progressive profound HL | RS | CN VIII | NTR No | Same | Same |
| Mangham et al. [2] | 29 M | 45 F | Progressive profound HL | TL | CN VII | Deaf | Facial palsy |
| | | | | MFA | CN VII | No | Deaf | Same facial palsy |
| | | | | TL | CN VII | No | deaf | normal |
| | | | | MFA | CN VII | No | Unchanged | No facial spasm |
| | | | | TL | CN VII | No | No facial spasm | Normal |
| | | | | MFA | CN VII | No | THL worse | Worse facial palsy |
| | | | | TL | CN VII | No | Unsteadiness improve | Delayed total FP recovered |
| Pappas et al. [3] | 26 M | 31 F | Progressive profound HL | TL | CN VII | NTR Yes | Deaf | Posterior | Deaf |
| | | | | TL | CN VII | NTR Yes | Unsteadiness improve | FP Unchanged |
| | | | | TL | CN VII | TR Yes | Unsteadiness improved | FP improved |

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|   | Age | Hearing Status | Tinnitus | Facial Nerve | Facial Function | Facial Anesthesia | Facial Palsy | Facial Palsy Improvement |
|---|-----|----------------|-----------|--------------|----------------|-------------------|--------------|-------------------------|
| 39 M | Progressive profound HL | TL | CN VII | No | THL Unsteadiness improved | Normal |
| 56 M | Progressive profound HL | TL | CN VII | NTR No | THL Unsteadiness improved | Postoperative FP improved |
| 44 M | Progressive severe HL deaf | TL | CN VII | NTR No | THL | Postoperative FP improved |
| 66 F | Acute profound HL unsteady deaf acute FP | TL | CN VII | No | THL Unsteadiness improved | IMPROVED |
| Madden et al. [4] | 36 F | Progressive profound HL Progressive FP Facial spasm | TL | CN VII Facial- Facial anastomosis | TR No | T HL Same facial nerve Improved Facial spasm |
| Bordi et al. [5] | 29 M | Progressive moderate HL PTA 60 | RS | CN VII Facial- Facial anastomosis | TR No | Deaf Facial palsy |
| Jacobson et al. [6] | 41 F | Imbalance Normal hearing | MFA | CN VII | TR No | Normal Normal |
| Cremers et al. [7] | 39 M | progressive profound HL Tinnitus Recurrent FP | TL | CN VII | TR No | T HL Worse facial palsy |
| Fujino et al. [8] | 58 M | Progressive mild to moderate serviceable HL Tinnitus Vertigo | RS | Attached CN III | TR No | Hearing worse Vertigo improved Mild Facial palsy |
| Babu et al. [9] | 36 M | Progressive severe HL PB 20 Vertigo | RS | CN VII | TR No | Improved Normal |
| Saleh et al. [10] | 44 M | Progressive severe HL PTA 70 Progressive FP | TL | CN VII | TR No | Deaf Facial palsy worse |
| Kohn et al. [11] | 44 M | Progressive profound HL Tinnitus Progressive FP | TL | CN VII NTR | STR No | Deaf Same |
| Name                  | Gender | Age | Type of Hearing Loss | Associated Symptoms | Type of Treatment | Outcome |
|-----------------------|--------|-----|----------------------|---------------------|-------------------|---------|
| Greiner - Perth et al. | M      | 32  | MILD MODERATE        | Tinnitus            | RS                | Normal  |
|                       |        |     | Sudden HL            | serviceable         |                   |         |
|                       |        |     |                      | Dizziness           |                   |         |
|                       |        |     |                      | Attached CN VII     |                   |         |
|                       |        |     |                      | CN VIII             |                   |         |
|                       |        |     |                      | TR No               |                   |         |
|                       |        |     |                      | Hearing improved    |                   |         |
|                       |        |     |                      | Vertigo improved    |                   |         |
|                       |        |     |                      | Normal              |                   |         |
| Fukuda et al.         | M      | 34  | MILD MODERATE        | Vertigo             | MFA               | Same    |
|                       |        |     | Progressive serviceable HL | CN VII   | TR No           | Vertigo improved |
|                       |        |     | Tinnitus             |                      |                   | Normal  |
| Gjuric et al.         | F      | 43  | MILD MODERATE        | Mild to moderate HL | MFA               | Same    |
|                       |        |     | serviceable          | Tinnitus            | TR No             | Normal  |
| Sasaki et al.         | F      | 39  | MILD MODERATE        | Mild progressive HL | MFA               | Same    |
|                       |        |     | serviceable          | Tinnitus            | CN VIII CN VII    | Post-operative Facial palsy improved |
|                       |        |     |                      |                      | NTR No            |         |
|                       |        |     |                      | Vertigo improved    |                   |         |
|                       |        |     |                      | Same preserve       |                   |         |
| Omjola et al.         | M      | 45  | RAPIDLY PROGRESSIVE PROFOUND HL | | TL              | Deaf               |
|                       |        |     | HL                    | Facial spasm        | CN VII            | Increase facial palsy |
|                       |        |     | Tinnitus              |                      | TR No             | Facial spasm improved |
| Roche et al.          | F      | 34  | PROGRESSIVE PROGRESSIVE PROFOUND HL | Tinnitus | TL              | Deaf               |
|                       |        |     | HL                    |                      | CN VII            | Postoperative FP |
|                       |        |     |                      | Tinnitus            | TR No             |         |
|                       |        |     |                      |                      | Deaf              |         |
| Sasaki et al.         | M      | 62  | PROGRESSIVE PROGRESSIVE PROGRESSIVE HL | Tinnitus | TL               | Deaf               |
|                       |        |     | HL                    |                      | CN VII Hypoglossal - Facial anastomosis | Postoperative |
|                       |        |     | Moderate HL           |                      | TR Yes            | F         |
|                       |        |     | Tinnitus              |                      |                   |         |
| Sasaki et al.         | M      | 23  | PROGRESSIVE PROGRESSIVE PROGRESSIVE HL | Tinnitus | RS              | Deaf               |
|                       |        |     | HL                    |                      | CN VII CN VII     | Improved FP   |
|                       |        |     | Sudden FP             |                      | TR No             |         |
|                       |        |     | Tinnitus              |                      | Deaf              |         |
| Sasaki et al.         | M      | 40  | PROGRESSIVE PROGRESSIVE PROGRESSIVE HL | Tinnitus | RS              | Deaf               |
|                       |        |     | HL                    |                      | CN VIII            | Facial palsy worse |
|                       |        |     | Tinnitus              |                      | TR No             |         |
| Sasaki et al.         | M      | 42  | PROGRESSIVE PROGRESSIVE PROGRESSIVE HL | Tinnitus | RS              | Deaf               |
|                       |        |     | HL                    |                      | CN VIII            | Facial palsy worse |
|                       |        |     | Tinnitus              |                      | TR No             |         |
| Sasaki et al.         | M      | 53  | PROGRESSIVE PROGRESSIVE PROGRESSIVE HL | Tinnitus | RS              | Deaf               |
|                       |        |     | HL                    |                      | CN VII CN VIII     | Facial palsy worse |
|                       |        |     | Tinnitus              |                      | Hypoglossal- Facial anastomosis | FacialSpam improved |
|                       |        |     |                      |                      | TR No             |         |
| Sasaki et al.         | M      | 53  | PROGRESSIVE PROGRESSIVE PROGRESSIVE HL | Tinnitus | RS              | Deaf               |
|                       |        |     | HL                    |                      | CN VII CN VIII     | Facial palsy worse |
|                       |        |     | Tinnitus              |                      | Hypoglossal- Facial anastomosis | FacialSpam improved |
|                       |        |     |                      |                      | TR No             |         |
| Sasaki et al.         | F      | 53  | PROGRESSIVE PROGRESSIVE PROGRESSIVE HL | Tinnitus | RS              | Deaf               |
|                       |        |     | HL                    |                      | CN VII CN VIII     | Facial palsy worse |
|                       |        |     | Tinnitus              |                      | Hypoglossal- Facial anastomosis | FacialSpam improved |
|                       |        |     |                      |                      | TR No             |         |

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| Reference | Age | Sex | Presentation | MRI Findings | Treatment | Outcome | Comments |
|-----------|-----|-----|--------------|--------------|-----------|---------|----------|
| Lenarz et al. [19] | 51 M | Progressive severe HL, deaf PT, progressive FP, Tinnitus, unsteadiness | TL | CN VIII CN VII Facial - Facial anastomosis | TR No | Deaf | Same FP |
| Safarova et al. [20] | 40 M | Progressive profound HL, Recurrent FP | RS | CN VIII CN VII | TR No | Dead | FP improved |
| Barrera et al. [21] | 21 M | Progressive profound HL, Tinnitus | TL | CN VIII | TR No | THL | Facial palsy |
| Aquilina et al. [22] | 29 F | Progressive profound HL, Tinnitus, Progressive Facial palsy, Multiple familial hemangioma | TL | CN VIII CN VII | TR No | Deaf | Same |
| Albid et al. [23] | 61 M | Progressive profound deaf HL, Tinnitus, Progressive FP | RS | CN VII | TR No | Same deaf | Same |
| Shaida et al. [24] | 30 F | Progressive profound HL, unsteadiness, Tinnitus, FP | TL | CN VII | TR No | Deaf Unsteadiness improved | Improved |
| Sepehrina et al. [25] | 53 M | Progressive Moderate HL, Serviceable Tinnitus, Progressive FP | RS | CN VII | TR No | Worse hearing loss | Improved |
| Zhu et al. [26] | 40 M | Progressive severe HL, Tinnitus, vertigo, Sudden facial palsy, progressive | Progressive | Progressive | Progressive | worse |
| 27 M | Progressive severe HL, Tinnitus, vertigo, progressive FP | TL | CN VII CN VIII | TR No | Deaf | Same |
| 31 M | Progressive mild HL, serviceable Tinnitus, vertigo, progressive FP | MFA | CN VII CN VIII | TR No | Deaf | Same |
| 37 F | Progressive severe HL, Tinnitus, Recurrent facial palsy | TL | CN VII CN VIII Facial - Facial Anastomosis | TR No | Deaf | Same palsy |
| 32 F | Progressive severe HL, Tinnitus, Facial spasm | TL | CN VII CN VIII Facial - Facial Anastomosis reconstruction | TR No | Deaf | Worse facial palsy Facial spasm Improved |
| Age  | Gender | Hearing | Facial Paralysis | Follow-up | Details |
|------|--------|---------|------------------|-----------|---------|
| 23 M |         |         | Mild HL, Tinnitus | Wait, scan |         |
| 34 M |         |         | Sudden profound HL, Sudden facial palsy | TL | STR Yes | Deaf | Same |
| 43 M |         |         | Progressive profound HL | TL | NTR No | Deaf | Improve |
|      |         |         | Progressive FP | | | | |
| 60 M |         |         | Progressive severe HL | TL | NTR No | Deaf | Mild postoperative FP |
| 18 M |         |         | Progressive profound HL, Progressive FP | TL | NTR No | Deaf | Improved |
| 41 M |         |         | Progressive profound HL | TL | NTR No | Deaf | Postoperative FP |
| 47 M |         |         | Moderate progressive mild HL (serviceable) | RS | NTR No | Deaf | Normal |
| 49 M |         |         | Moderate progressive mild HL (serviceable) | RS | NTR No | Deaf | Normal |
| 28 M |         |         | Mild progressive mild HL serviceable | MFA | NTR No | Deaf | Normal |
| 49 F |         |         | Progressive profound HL | TL | NTR Yes | Deaf | Normal |
| 66 M |         |         | Progressive severe HL | RS | NTR No | Deaf | Normal |
| 36 M |         |         | Sudden severe HL | TL | STR Yes | Deaf | Normal |
| 56 M |         |         | Progressive profound HL | TL | NTR Yes | Deaf | Postoperative FP |
| 22 F |         |         | Vertigo, Progressive mild HL serviceable | RS | CN VII CN VIII | NTR No | Hearing same | Mild FP |
| 47 M |         |         | Progressive profound HL, Progressive FP | TL | CN VIII | TR No | Deaf | Same |
| 45 M |         |         | Progressive Profound HL | TL | CN VIII | TR No | Deaf | Same |
| 38 M |         |         | Progressive profound HL | TL | CN VII CN VIII | STR No | Deaf | Postoperative FP |
| 41 M |         |         | Progressive Severe HL, Tinnitus Vertigo | TL | CN VIII | TR No | Deaf | Vertigo improved |
| 21 M |         |         | Moderate progressive serviceable Vertigo | RS | CN VII CN VIII | TR No | Deaf worse | Vertigo improved |

**References**

Oldenburg et al. [27]

Mastronardi et al. [28]

Hanamitsu et al. [29]

Bonforte et al. [30]

Nakashima et al. [31]

Hekmatare et al. [32]

Silveira et al. [33]

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| Reference                | Gender | Age | Symptoms                                                                 | Nerves | Site of Lesion | Hearing Status | Observation  |
|--------------------------|--------|-----|---------------------------------------------------------------------------|--------|----------------|----------------|--------------|
| Magliulo et al. [34]     | 38 F   | 58  | Progressive severe profound HL, Dizziness, Tinnitus, Progressive Facial palsy | TL     | CN VIII        | Deaf           | Improved     |
| Mahran et al. [35]       | 58 F   | 58  | Sudden profound HL, Vertigo, Facial spasm, Tinnitus                      | RS     | CN VIII CNVIII | Deaf           | Improved     |
| Refass et al. [36]       | 36 F   | 24  | Sudden deaf HL, Tinnitus                                                 | TL     | CN VIII        | Deaf           | Same         |
| Matias-Guiu et al. [37]  | 24 F   | 24  | Progressive profound HL, Tinnitus                                         | RS     | CN VIII        | Deaf           | Same         |
| Ferrante et al. [38]     | 24 F   | 24  | Acute profound HL, vertigo                                               | RS     | CN VIII        | Deaf           | Same         |
| Shao-yan et al. [39]     | 47 M   | 47  | Progressive profound HL, Progressive FP                                 | TL     | CN VIII        | Deaf           | Same FP      |
| Moore et al. [40]        | 45 M   | 45  | Progressive profound HL, Hearing loss, Imbalance, Headache                | RS     | CN VIII CNVIII | Deaf           | Transient FP |
| Deshmukh et al. [41]     | 67 M   | 67  | Sudden severe HL, Sudden FP                                              | RS     | CN VII CNVIII  | Same           | Improved     |
| 53 M                     |        |     | Sudden severe HL, Sudden FP, Multiple familial hemangioma, headache       | RS     | CN VII CNVIII  | Same           | Improved     |
| Ahmad et al. [42]        | 45 M   | 45  | Vertigo, Tinnitus, Progressive Profound HL, Facial spasm                 | TL     | CN VII CNVIII  | Same hearing   | Vertigo      |
|                          |        |     |                                                                           |        |                | Facial palsy   | Facial spasm |
| Jun Shim et al. [43]     | 5 M    | 5   | Sudden Profound HL                                                       | Observation |        | TR Yes         |              |
| Di rocco et al. [44]     | 23 F   | 23  | Progressive severe HL (60-70 dB), Acute FP                               | RS     | CN VII CNVIII  | Same           | Improved     |
Demographic

There were 81 patients of age ranged from 5 to 69 with majority of the patients between 20 to 50 year old. There were 57 males and 24 females in the study (Figures 1 & 2).

Symptoms

78 patients had hearing loss (96%). 11 patients of them had sudden hearing loss (14%), while the other 67 patients had progressive hearing loss (86%). 16 patients had serviceable hearing loss (20%), while the other 52 patients had non-serviceable hearing a loss (80%).

46 patients had tinnitus (56%), 31 patients had vestibular symptoms (38%) (16 patients had vertigo and 15 patients had unsteadiness). 30 patients had facial palsy (37%), 6 patients of them had sudden facial palsy (20%), 3 patients of them had recurrent facial palsy (10%), while the other 21 had progressive facial palsy (70%). 13 patients had facial spasm (16%). 2 patients had headache (3%), and 2 patients had familial multiple hemangioma (3%) (Table 2).

| Hearing loss     | Tinnitus | Vertigo and unsteadiness | Facial palsy | Facial spasm | Headache |
|------------------|----------|--------------------------|--------------|--------------|----------|
| 96%              | 56%      | 38%                      | 37%          | 16%          | 3%       |

Management

Table 3: Hearing outcomes after surgery.

| Normal hearing | Serviceable hearing loss | Non serviceable |
|----------------|--------------------------|-----------------|
| Patients       | 3                        | 15              | 51              |
| Improved       | 2                        | 5               | 0               |
| Percentage     | 66% preserve hearing     | 33% preserve serviceable hearing | 0% preservation |

Table 4: IAC Hemangioma management.

| Observation | MFA | RS | TL |
|-------------|-----|----|----|
| 2           | 8   | 29 | 42 |
| 3%          | 9%  | 35%| 51%|
| 3%          | 9%  | 35%| 51%|

Table 5: Facial palsy outcomes after surgery.

| Improved     | Same | worse |
|--------------|------|-------|
| 6 patients with acute FP | 4    | 1     |
| 3 patients with recurrent FP | 1    | 1     |
| 21 patients with progressive FP | 6    | 11    | 5    |
| Total        | 11 (36%) | 13 (43%) | 6 (23%) |

Table 6: Facial reconstruction types.

| Facial- Facial anastomosis | Facial-hypoglossal anastomosis | Sural nerve graft | Great auricle nerve graft |
|----------------------------|---------------------------------|------------------|--------------------------|
| 10                         | 3                               | 2                | 1                         |

Two patients were managed by observation. 79 patients had a surgery. 3 of them had normal hearing, and 15 patients of them had serviceable hearing loss, while the other 51 patients had non-serviceable hearing loss (Table 3). Only 11 articles reported the postoperative clinical progress of vestibular symptoms, and all reported patients with vestibular symptoms improved. 13 patients had preoperative facial spasm, and it disappeared in all of them postoperatively. Two patients were managed conservatively, 8 patients had MFA, and 29 patients had RS approach, while the other 42 patients had TR approach (Table 4). 30 patients with facial palsy had a surgery, 11 patients of them had facial weakness improvement (Table 5). 16 patients had facial reconstruction (20%) (Table 6). 68 patients had a limited disease to IAC, while 13 patients had disease extension.
into CPA (Figure 3). 72 articles reported the surgical removal type, 52 patients of them had a total surgical resection, and 17 patients of them had a near total resection while the other 3 patients had only partial resection (Figure 4). Hemangioma attachments were reported in 63 patients, 27 patients of them had CN VII attachment, and 21 patients of them had CN VII & CN VIII attachment, while the other 15 patients had CN VIII attachment (Figure 5).

**Results**

Most patient were male with age ranging between 20 to 50 years old. Most patients had non-serviceable hearing loss. 38% patients had vestibular symptoms, 37% patients had facial palsy, 16% patients had facial spasm. Only third patient with serviceable-hearing loss preserved their hearing abilities after surgery, third patient with facial palsy had significant improvement after surgery (acute facial palsy has a better prognosis than progressive facial palsy), and all patients with facial spasm and vestibular symptoms had a good improvement. Most cases were located primary to IAC, most cases had attachment to facial nerve and can be removed totally, and about20% of patients had facial reconstruction surgery due to advanced facial nerve involvement.

**Conclusion**

IAC hemangioma is a rare disease with poor outcomes prognosis, we should suspect this disease in patients with progressive hearing loss and facial palsy with small lesion in internal auditory canal vertigo and Tinnitus.

**Discussion**

Vestibular schwannoma is the most common tumor in IAC and the cerebellopontine angle (CPA). Other tumor lesions of CPA area include meningioma, primary cholesteatoma, facial nerve neuroma, various vascular tumors, metastatic tumors, and others. cavernous angioma was reported rarely in the IAC. Sundares et al. reported the first case of IAC cavernous angioma in 1976. These tumors were considered to originate from the capillary bed of the epineurium nerve. Vascular steal mechanism in which the blood is taken by tumor instead of the nerve causing nerve function loss even with a small size of tumor. Histopathologically, they consist of large thin-walled blood vessels that lined by flattened endothelium, which stain positive by endothelial marker CD 31, the stroma is composed of fibrous component and has mainly myofibroblast and fibroblast that stain positive for smooth muscle actin.

Depending on nerve origin and location, these tumors can cause severe progressive or sudden sensorineural hearing loss, tinnitus, sudden or progressive facial nerve palsy, facial spasm, vertigo and disequilibrium even when they are in small size. The tumor sizes usually less than 10 mm. On CT scan, IACHemangioma appear as iso- or hyper dense lesion with slight enhancement after intravenous administration of contrast, usually stippled calcifications could also be seen with enlargement of the IAC. On MRI, it appears as lobular and iso-intense in T1 and hyperintense on T2 with heterogeneous T1 post - gadolinium enhancement. The main differential diagnosis is IAC Vestibular shwannoma (Table 7). Other differential diagnosis may include meningioma, lipoma, melanoma, hamartoma, and lymphoma.

| Table 7: IAC Hemangioma V/S IAC Vestibular shwannoma. |
|--------------------------------------------------------|
| **Intra-meatual CH**                                   | **Intra-meatual vestibular shwannoma**                      |
| Non-serviceable CH                                      | serviceable hearing                                       |
| Facial palsy                                           | Normal facial function                                    |
| MRI lobular and iso-intense in T1 and hyper intense on T2 with heterogeneous T1 post - gadolinium | Round or oval And have more homogeneous gadolinium uptake |

Complete surgical resection with avoiding complications such bleeding is the goal of treatment. Radiotherapy may promote growth and hemorrhage, so it is not recommended. No symptomatic cases can be managed conservatively. Symptomatic case should be resected surgically. Surgical approach is depended on hearing deficit and tumor size. Translabyrinthine approach is recommended for patient with non-serviceable hearing loss. Middle fossa approach and retrosigmoid approach are recommended for patients with serviceable hearing loss. This lesion could be attached firmly with facial nerve. Since the tumor is benign and slow growing, it is advocated to perform a near total resection in case where the tumor is difficult to dissect from facial nerve [18,26,27].

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**Conflict of interest**

Author declared no conflict of interest
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