Transient profound hearing loss and severe facial nerve palsy in schwannomas within the internal acoustic canal: A case report

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Abstract. Vestibular schwannoma is the most common intracranial schwannoma and constitutes ~8% of all intracranial tumors in adults with an estimated incidence rate of ~1.3/100.000. Facial nerve schwannomas and cochlear nerve schwannomas are rare, and information regarding incidence rates of these is still missing in the literature. All three variants of nerve origin present most frequently with unilateral hearing loss, unilateral tinnitus and disequilibrium. Facial nerve palsy is a common finding in facial nerve schwannomas but a rare finding with vestibular schwannomas. The symptoms are normally persistent and often progress over time, leading to therapeutic interventions that predispose to quality of life-reducing morbidities, e.g., deafness and/or imbalance. The case report describes a 17-year-old male who, during a 1-month period, presented with profound unilateral hearing loss and severe facial nerve palsy followed by complete remission. An MRI scan showed a 5x8-mm schwannoma within the internal acoustic canal. Profound hearing loss together with concomitant severe peripheral facial nerve palsy in small schwannomas within the internal acoustic canal may show spontaneous and total remission within weeks of symptom debut. This knowledge, as well as possible remission of objective findings, should be considered before interventions with potential severe morbidity are suggested.

Introduction

A schwannoma is a rare benign tumor that is caused by pathological hyperproliferation of the myelin sheath producing Schwann cells, which can be found alongside the nerves of the peripheral nervous system and cranial nerves. A typical localization is alongside the pathway of the eighth cranial nerve and more rarely alongside the seventh cranial nerve. The vestibular schwannoma (VS) is the most frequent variant and constitutes ~8% of all intracranial tumors in adults (1). The facial nerve schwannoma (FNS) and the cochlear nerve schwannoma (CS) are so rare, that the incidence rates are still not well described (2,3). The incidence rate of VS is estimated to approximately 1.3/100.000 (4) but differs between subpopulations, including age, which is strongly associated with a higher risk, as individuals above the age of 50 have a rate of 2/100.000 or more, while individuals below 20 years of age have a rate of 0.05-0.1/100.000 (5,6). The prevalence is estimated to be much higher, as one study found incidentalomas in 2 out of every 10.000 (7). VSs either occur sporadic (in approximately 95% of cases with unilateral presentation except for a few known cases), or are associated with neurofibromatosis type 2 (NF2) (in ~5% of cases), which include different presentation case histories at the time of diagnosis and are most commonly bilateral (8). NF2 is caused by alterations in the NF2 gene on chromosome 22q, which is inherited as an autosomal dominant genetic disorder or due to de novo mutations. The phenotypical presentation depends on the genetic variant and epigenetics. NF2 patients present with VS at a younger age compared to sporadic VS, often during their adolescence, and young patients with sporadic VS seem to have a higher risk of genetic predisposition (9).

Symptoms occur as the tumor expands and induces pressure to surroundings. The order in which the symptoms present differs between the site of origin. With VS, the most frequently presenting symptom is ipsilateral sensorineural hearing loss (70-90%) and more than half of these progress further and a few experience sudden hearing loss, followed by tinnitus (50-80%), vestibular disequilibrium (15-75%) including vertigo and imbalance, and headache (10-20%) (10-15). A VS usually arises alongside the internal acoustic canal (IAC), but can protrude into the cerebellopontine angle (CPA) and potentially cause compression to the surrounding structures including the cerebellum, the fourth ventricle (resulting in hydrocephalus), and the fifth cranial nerve (resulting in trigeminal disturbances) (16,17). Large VSs are associated with facial nerve disturbances, as the eighth and seventh cranial nerves share a common pathway (18). Affection of the fifth and seventh cranial nerves are relatively rare, 9 and 6%, respectively (18). The correlation between tumor size and level of hearing loss

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is ambiguous as previous studies have shown contradictory results (19-21). However, the general opinion seems to be that size is not related to the severity of hearing loss (19-21).

Symptoms of FNS include 1) unilateral hearing loss, which can be sensorineural, conductive or mixed (50-80%), 2) facial nerve palsy (40-85%), 3) unilateral tinnitus (20-50%), 4) vertigo (20-50%), 5) facial spasm (5-25%), 6) headache (0.5-20%), parotid mass (20%) and hypesthesia (1-15%) (22-25). Vertigo and affected vestibular function are related to localization in the IAC (26). The age of debut seems younger than in VS, as ~30% of patient are below 30 years of age (27).

CS is extremely rare, and therefore the clinical picture may not be fully understood. In described cases, CS seem to usually present with both unilateral sensorineural hearing loss and tinnitus, but can present with one of these solely (3,28,29). The progression of the hearing loss in CS seem substantial compared to initial tumor size (29). Vertigo and imbalance are also described but infrequently.

Contemporary management of schwannomas includes intervention or a more conservative 'wait-and-scan' strategy. Recommendation and selection of either one of these strategies remain controversial, as inventions may cause a higher degree of morbidity than the tumor itself. The conservative strategy is often chosen with asymptomatic patients or when symptoms are sparse, because the growth patterns of schwannomas are known to be divers, as both non-growth, slow- and fast growth rates, and even tumor regression are well described (11,22). Approximately one half of all tumors do not grow following diagnosis (10,30), and particularly small tumors (below 10 mm) are associated with low growth potential (30).

Case report

A 17-year-old male, without any previous otological or vestibular disease(s), was admitted to the emergency department (ER) at Region Hospital North Jutland Thisted, Denmark, July 2016, with a 1-day history of episodes of rotational vertigo with accompanying nausea and vomiting. The vertigo worsened with head movements and ceased at rest. Symptoms included 1) unilateral left-sided tinnitus which worsened with head rotations to the left and 2) a 3-day history of hyperacusis.

Initial clinical examination, including a gross neurological assessment, revealed impaired hearing on the left side following finger rubbing in near proximity of both ears. A left-sided Dix-Hallpike-test supposedly evoked a minor positional nystagmus without any complaints of vertigo. The positional nystagmus observed was unfortunately not described in any further details. Routine blood testing was normal (blood counts, electrolytes, CRP, liver-, and kidney function test). The initial examinations and diagnostics were compromised in terms of vestibular- and otoneurologic examinations because no neurotologist and no advanced diagnostic equipment was available at the ER of the regional hospital.

The initial tentative diagnoses included vestibular neuritis and benign paroxysmal positional vertigo. Because these diagnoses were considered harmless, the patient was discharged from the ER the same day and scheduled for an appointment with a private otorhinolaryngologist (ENT) the following day for further examinations and targeted treatment. At the private ENT clinic, the objective neurotologic examination revealed a spontaneous right-beating nystagmus as well as pathological saccades with video head impulse testing of the left lateral semicircular canal. The patient was diagnosed with a left-sided vestibular neuritis and treatment with 50 mg of prednisone was prescribed for the following 10 days. After 4 days, the patient experienced progressive left-sided facial weakness, progressive ipsilateral hearing loss and echo sensation, and 2 days later the patient was admitted to the Department of Otorhinolaryngology, Head & Neck Surgery at Aalborg University Hospital, Denmark. The patient now complained of mild pain in left ear when yawning or biting his teeth together, vertigo was not a complaint anymore and ipsilateral tinnitus was intermittent. Otoneurological examinations found a negative Rinne test on the left side and a Weber test with lateralization to the right. No spontaneous- or gaze-evoked nystagmus was observed. Head shake test showed a pathological horizontal right-beating nystagmus. Left-sided peripheral facial nerve palsy was present and classified as a House-Brackmann (HB) grade 3 with progression to a grade 5 2 days later with accompanying Bell's phenomenon. The sensibility of the face was normal. Audiometry done 10 days after initial symptom debut, refer to Fig. 1A, showed a left-sided profound hearing loss in the frequency range between 250-2,000 Hz and a severe hearing loss with the remaining frequencies. Ipsilateral speech discrimination score (DS) was 0%. The hearing on the right side was within the normal range (better than or equal to 20dB).

Cerebral MRI showed an 8x5-mm tumour in the left IAC near the porus with minimal protrusion into the CPA. Several neuroradiological experts and specialists have analysed the MRI images, and the joint conclusion was that the observed tumor was a schwannoma, but the specific nerve origin had been impossible to determine. The three possible origins of the tumor included 1) the face nerve, 2) the vestibular nerve, and 3) the cochlear nerve.

Neither neuromatic nor neurofibromatotic stigmatae were found, nor were any known familiar predispositions encountered.

The patient was discharged with persisting symptoms 5 days following admittance and was scheduled for a follow-up 9 days later, corresponding to 24 days after initial symptom debut and 12 days after debut of facial nerve palsy. At the scheduled follow-up, the clinical examinations concluded that there was a complete remission of the left-sided hearing loss (pure tone audiometry thresholds were all within the normal range) except for a discrimination score (DS) of 70%, as seen in Fig. 1B, and complete remission of facial nerve palsy, vertigo, and tinnitus.

At the next follow-up 20 days later, the DS was within the normative range too, as seen in Fig. 1C, and the audiometry was also within the normative range 6 months later as shown in Fig. 1D. Annual follow-ups with clinical examination, audiometry, vestibular examination, and MRI has been done for 6 consecutive years, and neither relapse of symptoms nor tumor growth has been reported.

Discussion

Schwannomas of the different nerves in the IAC induce similar main symptoms, though the order in which they present as

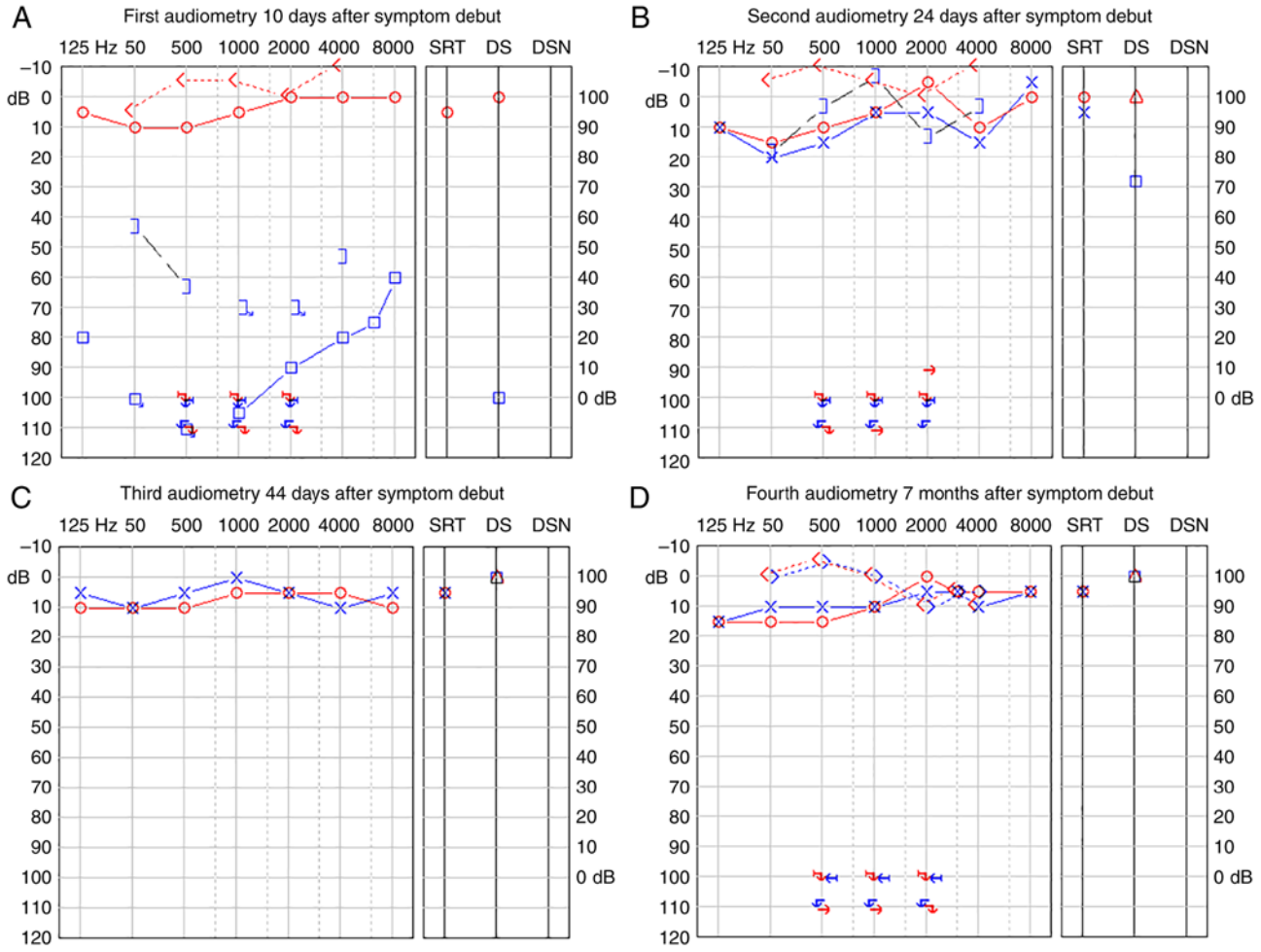

Figure 1. Consecutive standard tone- and speech audiometries. (A) Standard pure tone- and speech audiometry 10 days following symptom debut showed asymmetric hearing with left-sided profound sensorineural hearing loss with the frequencies ranging between 250 and 2,000 Hz, severe inner ear hearing loss in the higher and lower frequencies and accompanying DS of 0. (B) After 2 weeks (24 days after symptom debut), subsequent standard pure tone- and speech audiometry showed symmetrical pure tone hearing thresholds within the normal range of 20 dB. However, the speech audiometry was still asymmetrical with a 70% DS on the left side. (C) Approximately 1 month after the first audiometry indicated a left-sided profound hearing loss, the DS was normal too. (D) Audiometry continued to be normal half a year later. x-axis, frequencies of stimuli in Hz; y-axis, sound intensities in dB. Red, right side; blue, left side. Solid line, air conduction thresholds (unmasked left, X; unmasked right, O); dotted line, bone conduction thresholds (unmasked left, >; unmasked right, <); masked air conduction thresholds (masked left, Δ; masked right, □); masked bone conduction thresholds (masked left,]; masked right, [). Horizontal arrows indicate stapedial reflex thresholds ipsi- and contralaterally. Arrows without a perpendicular line at the end indicate contralateral thresholds and arrows with a perpendicular line at the end indicate ipsilateral thresholds. The arrows pointing downwards indicate no stapedial reflex. DS, discrimination score; SRT, speech recognition threshold; DSN, discrimination score in noise (was not examined with this patient).

well as the probability seem to differ. Inner ear hearing loss is the most frequent symptom in all IAC schwannomas, and it is usually gradually progressive and persisting. Sometimes, the hearing loss presents as sudden deafness, also referred to as sudden hearing loss, which is a loss of sensorineural hearing of at least 30 dB involving at least three neighbouring frequencies within a 72-hour period. Sudden deafness occurrence with VS is reported to be 12-19% (15,31,32). Spontaneous remission, however, has not been evaluated systematically in the included literature, although it has been known to occur for more than 30 years (33). Anacusis is more rarely seen and even more rarely a passing event. Clinical presentation with anacusis is reported to be 9% with FNS (34) and 4% with VS (31), but none of these studies described any transient course. Fluctuating symptoms with VS has been reported to be 6%, but the level of hearing loss was not evaluated (15). Other studies found spontaneous remission of sudden hearing loss in 17-24% of VS patients within 1 month (35,36). These studies, however, define sudden hearing loss as a loss of hearing of at least 10 dB in two consecutive frequencies in a few days-a definition, which must be considered obsolete cotemporally. Two studies have reported the age range of their population with sudden deafness, and both these studies had 20 years of age as the minimum age (31,37). Thus, adolescents presenting with anacusis is extremely rare.

To some extent, there seems to be consensus regarding the fact, that the degree of hearing loss is not directly correlated to tumor size, and even small tumors (less than 10 mm) may cause substantial hearing loss and sudden deafness (32,37,38). Anacusis has been associated with larger FNS tumors (20), but existing literature evaluating this is sparse. In addition, progression of sensorineural hearing loss does not seem to correlate with tumor growth in VSs (29). It has been proposed that the hearing loss is caused by a multifactorial pathogenesis,

comprising of compression of both cochlear nerve fibres and vascular supply, and conduction block due to environmental and chemical changes (31,37,38).

VS is the most frequent schwannoma in the IAC, and facial nerve palsy is only seen in up to 6% of cases (18). The House-Brackmann grading system is used to grade the severity of facial paresis (39), and severe paresis (HB≥5) is only seen in about 10% of cases with facial nerve palsies (22). The size of VS is known to be associated with the presence of peripheral facial nerve palsies (40). Thus, the probability of a small VS (less than 10 mm) causing a severe facial nerve palsy (above or at HB 5 score) is very low.

Contrary to VS, facial nerve palsy is one of the main symptoms of FNS, as up to 85% and generally more than half of these patients experience this (22-24). The number of patients that experience only temporary palsy is reported to be 6-18% in two studies with 56 and 53 cases (23,34), many of which were patients who did not experience any facial nerve palsies at the time of inclusion but retrospectively described specific symptoms. Approximately half of the patients presented with some degree of paresis, and none of these were transient in the follow-up period. One might possibly think, that patients who described a transient course of their palsy did not experience moderate to severe facial nerve palsies, as this most likely would have resulted a wish to seek medical evaluation.

The origin of the involved nerve in this case report was not able to be verified by the neuroradiologist, and therefore, parameters like symptomatology, size and site may contribute to determination of the most likely nerve origin. When comparing the above-mentioned symptomatology and probabilities, the most likely nerve origin with this case report seems to be the facial nerve. This is primarily based upon the fact that facial nerve palsies are most commonly found in FNS regardless of size. With VSs, on the contrary, facial nerve palsy is predominantly related to larger size tumors and rarely seen. The characteristics of the hearing loss do not seem to differ conspicuously between VS and FNS. To the best of the authors' knowledge, no existing CS literature report facial nerve palsy or spontaneous remission of hearing loss.

In conclusion, sudden profound hearing loss, together with concomitant severe peripheral facial nerve palsy in small schwannomas in the IAC, may have a transient course with spontaneous and complete remission within weeks of initial onset of symptoms. Thus, inner ear schwannomas should always be considered whenever patients present with ipsilateral inner ear hearing loss and peripheral facial nerve palsy no matter the severity and duration, especially if combined with accompanying symptoms of ipsilateral tinnitus and disequilibrium. The current knowledge of possible spontaneous and complete remission of severe symptoms and objective findings should be considered when interventions with potential severe morbidity is considered.

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Availability of data and materials

All data generated or analysed during this study are included in this published article.

Authors' contributions

MHT was responsible for data curation, formal analysis, investigation, methodology, project administration, visualization and writing of the original draft. DDH was responsible for conceptualization, funding acquisition, resources, supervision, and review and editing of the entire article. MHT and DDH confirm the authenticity of all the raw data. Both authors have read and approved the final manuscript.

Ethics approval and consent to participate

Ethical review and approval were not required for the study on human participants in accordance with the local legislation and institutional requirements. Written informed consent for participation was not required for this study in accordance with the national legislation and the institutional requirements. The study follows the Declaration of Helsinki.

Patient consent for publication

The authors have received written informed consent from the patient included in this case report prior to submission when the patient was 22 years old.

Competing interests

The authors declare that they have no competing interests.

References

1. Ostrom QT, Gittleman H, Liao P, Vecchione-Koval T, Wolinsky Y, Kruchko C and Barnholtz-Sloan JS: CBTRUS statistical report: Primary brain and other central nervous system tumors diagnosed in the United States in 2010-2014. Neuro Oncol 19 (suppl_5): v1-v88, 2021.
2. McRackan TR, Wilkinson EP and Rivas A: Primary tumors of the facial nerve. Otolaryngol Clin North Am 48: 491-500, 2015.
3. Shin SH, Chun YM and Lee HK: A cochlear schwannoma presenting with sudden hearing loss. Eur Arch Otorhinolaryngol 265: 839-842, 2008.
4. Tos M, Stangerup SE, Cayé-Thomasen P, Tos T and Thomsen J: What is the real incidence of vestibular schwannoma? Arch Otolaryngol Head Neck Surg 130: 216-220, 2004.
5. Gal TJ, Shinn J and Huang B: Current epidemiology and management trends in acoustic neuroma. Otolaryngol Head Neck Surg 142: 677-681, 2010.
6. Kshettry VR, Hsieh JK, Ostrom QT, Kruchko C and Barnholtz-Sloan JS: Incidence of vestibular schwannomas in the United States. J Neurooncol 124: 223-228, 2015.
7. Lin D, Hegarty JL, Fischbein NJ and Jackler RK: The prevalence of 'incidental' acoustic neuroma. Arch Otolaryngol Head Neck Surg 131: 241-244, 2005.
8. Carlson ML and Gompel JJ: Multiple unilateral vestibular schwannomas: Segmental NF2 or sporadic occurrence? J Neurol Surg Rep 77: e106-e108, 2016.
9. Sadler KV, Bowers NL, Hartley C, Smith PT, Tobi S, Wallace AJ, King A, Lloyd SKW, Rutherford S, Pathmanaban ON, *et al*: Sporadic vestibular schwannoma: A molecular testing summary. J Med Genet 58: 227-233, 2021.
10. Tschudi DC, Linder TE and Fisch U: Conservative management of unilateral acoustic neuromas. Am J Otol 21: 722-728, 2000.
11. Shigihara S: Natural history of acoustic neuroma. Practica Oto-Rhino-Laryngologica 106: 668-689, 2013 (In Japanese).

12. Kentala E and Pyykkö I: Clinical picture of vestibular schwannoma. Auris Nasus Larynx 28: 15-22, 2001.
13. Moffat DA, Jones SE, Mahendran S, Humphriss R and Baguley DM: Referral patterns in vestibular schwannomas -10 years on. Clin Otolaryngol Allied Sci 29: 515-517, 2004.
14. Goldbrunner R, Weller M, Regis J, Lund-Johansen M, Stavrinou P, Reuss D, Evans DG, Lefranc F, Sallabanda K, Falini A, *et al*: EANO guideline on the diagnosis and treatment of vestibular schwannoma. Neuro Oncol 22: 31-45, 2020.
15. Morrison GAJ and Sterkers JM: Unusual presentations of acoustic tumours. Clin Otolaryngol Allied Sci 21: 80-83, 1996.
16. Al Hinai Q, Zeitouni A, Sirhan D, Sinclair D, Melancon D, Richardson J and Leblanc R: Communicating hydrocephalus and vestibular schwannomas: Etiology, treatment, and long-term follow-up. J Neurol Surg B Skull Base 74: 68-74, 2013.
17. Liu P, Liao C, Zhong W, Yang M, Li S and Zhang W: Symptomatic trigeminal neuralgia caused by cerebellopontine angle tumors. J Craniofac Surg 28: e256-e258, 2017.
18. Matthies C and Samii M: Management of 1000 vestibular schwannomas (acoustic neuromas): Clinical presentation. Neurosurgery 40: 1-9; discussion 9-10, 1997.
19. Wagner JN, Glaser M, Wowra B, Muacevic A, Goldbrunner R, Cnyrim C, Tonn JC and Strupp M: Vestibular function and quality of life in vestibular schwannoma: Does size matter? Front Neurol 2: 55, 2011.
20. Day AS, Wang CT, Chen CN and Young YH: Correlating the cochleovestibular deficits with tumor size of acoustic neuroma. Acta Otolaryngol 128: 756-760, 2008.
21. Nadol JB Jr, Diamond PF and Thornton AR: Correlation of hearing loss and radiologic dimensions of vestibular schwannomas (acoustic Neuromas). Am J Otol 17: 312-316, 1996.
22. Carlson ML, Deep NL, Patel NS, Lundy LB, Tombers NM, Lohse CM, Link MJ and Driscoll CL: Facial nerve schwannomas: Review of 80 cases over 25 years at mayo clinic. Mayo Clin Proc 91: 1563-1576, 2016.
23. McRackan TR, Rivas A, Wanna GB, Yoo MJ, Bennett ML, Deitrich MS, Glasscock ME and Haynes DS: Facial nerve outcomes in facial nerve schwannomas. Otol Neurotol 33: 78-82, 2012.
24. Li Y and Dai C: A retrospective study on facial nerve schwannomas: A disease with a high risk of misdiagnosis and hearing loss. Eur Arch Otorhinolaryngol 274: 3359-3366, 2017.
25. Bartindale M, Heiferman J, Joyce C, Balasubramanian N, Anderson D and Leonetti J: The natural history of facial schwannomas: A meta-analysis of case series. J Neurol Surg B Skull Base 80: 458-468, 2019.
26. Chung JW, Ahn JH, Kim JH, Nam SY, Kim CJ and Lee KS: Facial nerve schwannomas: Different manifestations and outcomes. Surg Neurol 62: 245-252; discussion 452, 2004.
27. Wiggins RH 3rd, Harnsberger HR, Salzman KL, Shelton C, Kertesz TR and Glastonbury CM: The many faces of facial nerve schwannoma. AJNR Am J Neuroradiol 27: 694-699, 2006.
28. Işıldak H, Ibrahimov M, Yilmaz M, Enver O and Albayram S: A purely intracanalicular cochlear schwannoma presenting with progressive hearing loss. Ear Nose Throat J 90: 481-488, 2011.
29. Leonetti JP: Cochlear neuromas. Am J Otol 19: 499-502, 1998.
30. Battaglia A, Mastrodimos B and Cueva R: Comparison of growth patterns of acoustic neuromas with and without radiosurgery. Otol Neurotol 27: 705-712, 2006.
31. Moffat DA, Baguley DM, Von Blumenthal H, Irving RM and Hardy DG: Sudden deafness in vestibular schwannoma. J Laryngol Otol 108: 116-119, 1994.
32. Yanagihara N and Asai M: Sudden hearing loss induced by acoustic neuroma: Significance of small tumors. Laryngoscope 103: 308-311, 1993.
33. Sterkers JM and Hocsman E: Unusual circumstances of disclosure of acoustic neurinoma. Ann Otolaryngol Chir Cervicofac 108: 204-207, 1991 (In French).
34. McMonagle B, Al-Sanosi A, Croxson G and Fagan P: Facial schwannoma: Results of a large case series and review. J Laryngol Otol 112: 1139-1150, 2008.
35. Nageris BI and Popovtzer A: Acoustic neuroma in patients with completely resolved sudden hearing loss. Ann Otol Rhinol Laryngol 112: 395-397, 2003.
36. Popovtzer A, Nageris BI and Bahar G: Resolved sudden hearing loss as a presenting symptom of retrocochlear lesion. J Basic Clin Physiol Pharmacol 12 (2 Suppl): S101-S107, 2001.
37. Inoue Y, Kanzaki J and Ogawa K: Vestibular schwannoma presenting as sudden deafness. J Laryngol Otol 114: 589-592, 2000.
38. Remenschneider AK, Gaudin R, Kozin ED, Ishai R, Quatela O, Hadlock TA and McKenna MJ: Is the cause of sensorineural hearing loss in patients with facial schwannomas multifactorial? Laryngoscope 127: 1676-1682, 2017.
39. House JW and Brackmann DE: Facial nerve grading system. Otolaryngol Head Neck Surg 93: 146-147, 1985.
40. Swensson RC, Swensson RP, Caramante Pizzini FE, Robson Boldorini P and Jorge Júnior JJ: An uncommon presentation of an VIII nerve tumor. Rev Bras Otorrinolaringol 74: 628-631, 2008.