*Rev Bras Otorrinolaringol.*
*V.70, n.6, 795-9, nov./dec. 2004*

**ORIGINAL ARTICLE**

# Vestibular schwannoma presenting as sudden hearing loss

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Key words: schwannoma, sudden deafness, neurinoma, magnetic resonance imaging.

## Summary

The Sudden Hearing Loss (SHL) is a symptom caused by more than 60 different diseases, including Vestibular Schwannoma (VS). Shaia & Sheehy (1976) presented a study with 1,220 cases of SHL reporting 1% incidence of VS. There is no specific characteristic for the diagnosis of VS and Magnetic Resonance Imaging (MRI) is the gold standard exam to diagnose such disease. Aim: To determine the real incidence of VS presenting as SHL performing MRI in all patients with SHL. Study design: Transversal cohort. Material and Method: Prospective study in which MRI with gadolinium was performed in all patients with SHL in the Emergency Service of Sao Paulo Hospital from April 2001 through May 2003. Results: MRI was performed in 49 patients with symptoms of SHL, with three cases (6.1%) of VS found. Conclusion: The real incidence of VS presenting as SHL may be greater than that mentioned in conventional reports probably because MRI had not been performed in all patients with SHL.

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Study presented as oral presentation at 3o Congresso Triológico de Otorrinolaringologia, Rio de Janeiro, 2003.
Article submited on May 25, 2004. Article accepted on October 14, 2004.

## INTRODUCTION

Sudden hearing loss (SHL) should be considered a symptom and not a clinical entity since it could be caused by more than 60 different diseases. It is a dramatic symptom both for the patient and doctor providing health care and, regardless of all new developments of diagnostic methods, it is still quite challenging to confirm its etiology and therapeutic possibilities.

Incidence of sudden hearing loss (SHL) is five (5) out of forty-seven (47) in one thousand patients per year in USA, with incidence increasing with age[1]. In Brazil the reported frequency is 9 cases out of 10,000 urgent care cases[2]. The mean age of affection ranges from 40 to 44 years. [1]

Vestibular Schwannoma (VS) is among several causes of Sudden Hearing Loss (SHL). The incidence of SHL ranges from 3 to 26%[3,4] in Vestibular Schwannoma patients. This high variation could be related to the fact that such incidence will be higher if the patient is asked about previous episode of sudden deafness, since there is discrepancy between symptom awareness by the patient and confirmation of clinical symptom, mainly regarding the function of 8th cranial pair[5].

If we analyze it through a different perspective or, considering patients with SHL, VS is reported as a cause in 1% of the cases[2-4,6,7]. The major challenge is to find out, among such patients, which one of them resulted from VS, since there are no clinical and pathognomic characteristics able to easily guide us in to the diagnosis.

The objective of the present study was to evaluate the real incidence of VS in our setting through MRI exams performed in all patients with symptom of sudden hearing loss in our service.

## MATERIAL AND METHOD

Prospective study carried out at the Department of ENT and Head and Neck Surgery and at the Department of Imaging Diagnosis, Federal University of Sao Paulo, from April 2001 to May 2003.

Patients with complaints of sudden hearing loss or loss whose onset was within 72 hours underwent audiometry and immittanciometry. We considered as diagnosis of sudden hearing loss unilateral sensorineural hearing loss levels equal or higher than 30 dB in at least three contiguous audiometric frequencies. Individuals with hearing loss with different characteristics than those above mentioned were excluded from the study. The study included patients from both genders with no age restriction.

Investigated patients underwent temporal bone and brain MRI performed with Philips Gyroscan NT 1.5 tesla. Images were acquired in T1 Turbo Spin Eco (TSE) concentrated sequence in axial and coronal plans with and without contrast and sections of 2.5 mm thick. T2 images were also acquired in coronal plan in TSE with 2.5 mm thickness and in 3D through 0.7 mm reconstruction. The exam was complemented with cranial cross-sections in "Fluid Attenuation Inversion Recovery" (FLAIR) in order to evaluate the brain.

## RESULTS

Sixty-one patients were included in the study. Twelve out of those 61 discontinued the study or were not submitted to MRI for different reasons. The age ranged from 15 to 91 years (mean age was 45.4 years), 23 (46.9%) were male and 26 (53.1%) were female. In twenty-three patients (46.9%) the involvement was on the left side and twenty-six (53.1%) had it on the right side. Regarding ethnic group 34 (69.3%) of the patients were Caucasian, 10 (20.4%) were Native Brazilian *pardos*, 2 (4.1%) were African-descendents, and 3 (6.1%) were Asian-descendents.

Three out of forty-nine of the patients submitted to MRI (6.1%) had VS, and 2 of them (4%) had intracanalicular affection.

*Case 1*

NCS, 43 years old, Female, Caucasian. Patient mentioned left ear sudden hearing loss for one day, associated with tinnitus, dizziness and ear fullness. Symptoms were related to an infectious event of the upper airways. Patient had systemic arterial hypertension and hypercholesterolemia under control and with normal physical evaluation. Input audiometry presented thresholds of 60 dB in1 kHz, 50 dB in 1.5 kHz and 45 dB in 2 kHz, the remaining frequencies were normal. Laboratory tests were also normal. Patient underwent MRI that evidenced of 8$^{th}$ nerve intrameatal enhancement. Initial treatment was 60 mg/day prednisone and 400 mg penthoxyphiline every 8 hours. Patient presented full recovery after the 4$^{th}$ day of treatment with normal audiometric results. Patient had a second episode of left ear sudden hearing loss after 8 months with clinical and audiometric results practically equal to those of the first crisis. Medication was introduced again as previously prescribed with full recovery and normal audiometry occurring after thirty-five days of treatment. Patient was submitted to surgery for removal of the VS, which measured approximately 8 mm, originating from the Superior Vestibular Nerve. Patient evolved to profound hearing loss in high sounds (but with 92% discrimination) and no peripheral facial palsy.

*Case 2*

JAJ, 55 years old, Male, Caucasian. Patient with history of left ear sudden hearing loss for two days, followed by tinnitus. He had high blood pressure under control with captopril and his physical examination was normal. Audiometry showed 60 dB threshold in 1 kHz and 40 dB in

1.5 kHz. MRI was performed and left-side intracanalicular Vestibular Schwannoma was found. Patient had subtle improvement with clinical treatment and refused to undergo surgery, remaining under follow-up.

*Case 3*

NOS, 42 years old, Male, Brazilian Native. Patient complaint of right-ear sudden hearing loss for fifteen days followed by tinnitus, dizziness, ear fullness and headache. History of previous diabetes mellitus. His audiometry showed thresholds of 45 dB in 2 kHz, 70 dB in 3 kHz, 4 kHz, 6 kHz and 8 kHz. MRI showed tumor of approximately 30mm at the cerebellopontine angle. Patient underwent surgery to remove the tumor and evolved to hearing loss and grade II peripheral facial palsy on the right.

## DISCUSSION

Vestibular Schwannoma is a benign tumor, which can present itself in several ways from asymptomatic to dramatic neurological symptoms. The overwhelming majority of VS present auditory (95%) and vestibular (61%) symptoms, which may also include trigeminal (9%) and facial (6%)[5] nerves. SHL can be an initial symptom of VS in a variable percentage of cases. Higgs (1973) reported 10% of VS cases with symptoms of sudden hearing loss as its first manifestation, Morrison (1975) reported 17%, Hirsch & Anderson (1980), 7.3%, Pensac et al. (1985), 14.5%, Berg (1986), 13%, Thomsen & Tos (1988), 7%, Harder (1988), 5%, Odkvist (1988), 4%, Ogawa (1991), 22%, and Moffat (1994), 10.2% (3). Berrentini (1997) mentioned a variation ranging from 3 to 26% in the literature regarding cases of VS with sudden hearing loss, and explained it as the result of questions asked or not to patients about previous hearing loss events [4]. Another factor that also collaborated to this large variation in results was that many times patients did not notice the onset of the symptoms or the severity of the auditory symptom, with major discrepancy between auditory feeling and audiometry results[5]. The same applies to facial and trigeminal function [5].

The etymological mechanisms of Sudden Hearing Loss and VS have not been completely explained yet. Berenholz (1992) explained that SHL occurs in VS due to tumor compression of the 8th nerve[1]. The vestibule-cochlear nerve has a transition zone in myelinization known as Obersteiner-Redlich zone. This is a region with variable length in which the myelin sheath of Schwann cells meets with olygodendrocytes. The transition zone of the vascular component is also located there. Peripheral vascular supply occurs longitudinally, whereas the glial segment does not have a regular standard, with few anastomoses. Therefore, there is high potential for lesions in transition regions due to low vascularization and myelinization [8]. The compression of this area could lead to reversible neuropathy through blockage of nervous stimulus to the 8th nerve [1].

Yanagihara (1993) and Moffat (1994) mentioned tumor compression of internal auditory artery (IAA), or even intracanalicular intratumoral hemorrhage as a cause of Sudden Hearing Loss in patients with VS [,9,10]. According to Berrentini (1997), however, if this hypotheses were true, cochlear and vestibular dysfunction could be expected, since blood supply to this systems is a common one[4,]; however, SHL is rarely associated with acute vestibular dysfunction [11]. Inoue (2000) did not support the hypothesis of vascular compression leading to SHL in VS, since vascular occlusion would be associated with permanent cochlear loss, and ischemia of basal and middle gyrus of the cochlea should affect the apical gyrus, since vascularization of the apical gyrus comes from the middle gyrus [7]. The increased variability of audiometric results of VS also does not comply with the standard of vascular distribution of cochlea[12]. Ischemia, however, could occur both in peripheral vessels and inside the stria vascularis itself. Therefore, angiospasm and hemorrheological phenomena inside vascular stria could lead to random and reversible patterns of cochlear abnormalities. [7]

Probably VS causes sudden hearing loss due to the combination of such factors in different levels, which could explain the variation of loss patterns, which range from cochlear to retrocochlear [7].

The size of the tumor had already been related to SHL, with higher frequency of SHL occurring in smaller tumors[9]. Nevertheless if only the size was fundamental to SHL pathogenic factors, all large tumors should have SHL symptom even in smaller sizes. Most importantly than tumor size is its location (in or out of the internal auditory canal). Moffat (1994), in his case study carried out in 11 patients with SHL resulting from VS, reported that there was no significant statistical difference regarding distribution per size, however all SHL cases due to VS presented lateral tumors, that is, inside the IAC. None of the medial tumors in his case studies presented SHL. Three patients in our study presented SHL resulting from VS, only one patient (33.3%) had extracanalicular tumor of approximately 30 mm. The other two patients had intracanalicular tumors.

Identification of those patients with SHL that resulted from VS still remains a major clinical challenge. Unfortunately, there are no pathognomonic signs of SHL caused by VS. There are no clinical evidences that allow us to safely tell which SHL cases are resulting from VS from those that are not. In epidemiological terms, the younger the patient is, the higher the likelihood of it being consequence of VS[13]. Sudden Hearing Loss due to VS also tends to be milder than those caused by other causes (viral, CVA, fistulas, acute labyrinthitis and head trauma)[13]. The improvement of SHL, with or without treatment, does not exclude VS diagnosis. The improvement of Sudden Hearing Loss caused by VS is reported as rare [12,14], but partial to total improvement may occur with steroid therapy [1,4,12,14-17]. Steroid drugs may act

on the edema and ischemia resulting from inflammation, leading to the reduction of the intracanalicular tumor mass through absorption of intratumor fluids. [1]In this case study two (66.6%) out of three patients developed Sudden Hearing Loss due to VS and achieved partial to complete improvement with steroid therapy.

There are not many fully reliable methods for diagnostic exclusion of VS. Conventional VS audiometry demonstrated mild to moderate loss with low level of discrimination. Several studies however do not show conventional audiometric pattern[1,12,14,18]. Owing to several likely etiopathogenic mechanisms VS may cause SHL of varied intensity, both cochlear and retrocochlear losses, in high, medium and low frequencies, with or without stapedian reflex. Therefore, it is very difficult to differentiate through audiometry patients with Vestibular Schwannomas among those with SHL.

Audiometry brainstem response audiometry (ABR) is viewed as the most sensitive and reliable electrophysiological exam for diagnosis of VS, with 5% or less false-negative rates[19,20]. Chaimoff (1999), however, found 15% false-negative rate, making it unfeasible to use this exam as screening method to exclude VS in SHL. [13]

The exam of choice for VS diagnosis is MRI[1,13,21]. It is the most sensitive and specific exam for diagnosis of VS, therefore MRI is recommended for all patients with SHL to obtain better etiological analysis [,13,21,22]. Case 1 is a good example of the impossibility of excluding the presence of VS with conventional investigation methods, placing MRI as the required exam for diagnostic investigation of SHL.

The rate of VS in cases of SHL found in literature was 1% [6]. In our practice, upon performance of MRI in all patients with SHL the incidence found was 6.1%. We believe such higher rate found in this study was due to MRI tests performed in all patients, including those with improved symptoms, since this fact does not exclude the presence of VS[1]. MRI is not applied as a routine exam of patients with audiovestibular symptoms due to its high cost, but Schick (2001) recommended MRI limited to IAC and cerebellopontine angle as screening method in patients with vestibular cochlear symptoms. [21,22].

The current literature corroborates the results found in this study [23]. Nageris & Popovtzer (2003) investigated 67 patients with SHL using MRI and found VS in 36% of the cases[23]. In our opinion, this incidence could be overestimated, due to some factors. Firstly, Nageris & Popovtzer considered SHL as sensorineural losses of only 10 dB in 2 contiguous frequencies, whereas we used the minimum limit of 30 dB in at least 3 contiguous frequencies. Still, a defined limit of days for the onset of the disease was not set in his study as it was in the present study (3 days). It leads to inclusion of milder cases of hearing loss, not always of sudden onset, with likely increase of VS incidence. The authors mentioned middle or external ear diseases, endolymphatic hydropsia, perilymphatic fistula and systemic diseases as exclusion criteria. The exclusion of such patients also increased VS incidence in their cases, leading to overestimated results. Regardless of such overestimation, the results also showed that probably the actual incidence of VS is higher than the 1% classically rated and described in literature. It also shows the importance of MRI for the diagnosis of VS in patients with SHL.

Assuming that the current incidence of VS in SHL is higher than that conventionally described, and considering that MRI is the only reliable exam for exclusion diagnosis of VS, the authors of the present study recommend the performance of MRI in all patients with SHL, since its high cost would be justified by the increased frequency of VS cases diagnosed.

## CONCLUSION

The incidence of Vestibular Schwannomas in the sample of Sudden Hearing Loss is higher than that described in literature. Such discrepancy is related to underdiagnosis and lack of routine use of MRI in such cases. In the opinion of the authors of the present study, MRI should be carried out in all patients with Sudden Hearing Loss.

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