# *B-ENT*, 2015, 11, 309-312 Incidental findings of a Vestibular Schwannoma on Brain MRI

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**Abstract.** *Incidental findings of a Vestibular Schwannoma on Brain MRI*. Brain magnetic resonance imaging (MRI) is the gold standard for diagnosing a vestibular schwannoma (VS). The prevalence of VS ranges from 0.02-2.4%; the incidence is 20 per million per year. This case report describes two cases of VS identified on brain MRIs that were not related to the purpose of the MRI examination. A literature search was performed to find existing audiometric-based protocols for guiding MRI referrals. Asymmetric sensorineural hearing loss appeared to be the most important indication for excluding VS. However, no consensus was found on the significance of asymmetry or on the severity required to refer every patient with audiovestibular symptoms for a brain MRI, despite the view that the low number of incidental findings may not justify the additional cost.

## Introduction

Acoustic neuromas are more appropriately termed vestibular schwannomas (VSs); however, the two terms are interchangeable in the literature.<sup>1</sup> The reported prevalence of VS was 0.02-0.2% in retrospective brain magnetic resonance imaging (MRI) studies<sup>2-4</sup> and 2.4% in temporal bone studies.<sup>4</sup> An MRI of the brain is considered the primary diagnostic tool for VS.5 However, it has become apparent that current protocols used for MRI referral are not cost-effective; they are generally based on auditory brain stem responses (ABRs) or criteria, like asymmetric other audiologic sensorineural hearing loss (ASHL).5-8 In addition, current protocols may miss up to 10% of tumors, which may lead to clinical and medico-legal implications.7 On the other hand, the fear of litigation that results from missing a diagnosis of VS often compels ear, nose, and throat (ENT) specialists to refer patients for brain MRI more often than not, despite the current climate of financial restraints.<sup>5</sup> It is estimated that only 20% of patients in an ENT department are potential candidates for VS screening, but up to 70% of new patients with audiovestibular symptoms are referred for brain MRIs.5

Despite difficulties with current referral protocols, it should be stressed that timely diagnosis of VS is of paramount importance. Any delay in diagnosis can influence the average tumor size at the time of diagnosis. In turn, tumor size can influence the outcome of a potential surgery or radiotherapy applied for hearing preservation.<sup>6,7</sup>

Herein, two cases are described, where a VS was discovered incidentally in patients that underwent MRI for reasons other than audiovestibular symptoms.

## **Case reports**

## Case report 1

A 35-year-old male patient with epilepsy underwent a brain MRI as part of a standard seizure workup. The MRI showed a VS  $(12 \times 9 \times 9 \text{ mm})$  on the left side. The patient had never experienced any kind of hearing loss, tinnitus, or vertigo. A pure tone audiometry showed normal thresholds in both ears. The patient did not receive treatment, but he was followed by the neurology department.

## Case report 2

A 65-year-old female patient underwent a brain MRI after experiencing left-sided facial paralysis. The paralysis completely resolved with corticosteroids. The brain MRI revealed a right-sided VS of  $20 \times 14$  mm. Pure tone audiometry showed a bilateral sensory-neuronal hearing loss at high frequencies. She also showed an asymmetry of

10 dB on the right side at both 4 and 8 kHz. The patient was treated with gamma knife surgery, and she was followed by the radiotherapy department.

#### Discussion

This study described two cases of VS discovered incidentally in patients that underwent a brain MRI for reasons other than audiovestibular symptoms.

Recently, Vernooij et al. found four cases of VS in a retrospective analysis of 2000 brain MRI scans (0.2%) of asymptomatic volunteers.9 No pathological confirmation of the presumed brain tumors was obtained. Note that one of the patients with VS in that study later reported that he had undergone an earlier CT scan to investigate hearing loss, which he had not mentioned beforehand; however, the CT scan did not show any abnormalities. No audiometric data were available in that study. Also, interestingly, the MRIs in that study were performed without contrastenhancement; therefore, the prevalence of incidental findings could have been an underestimation.<sup>2</sup> Also, in a systematic review and meta-analysis, Morris et al. found five cases of VS in 16 studies, which included a total of 19,559 patients.<sup>3</sup> The prevalence of incidental VS in that study was 0.03% (number needed to scan = 3333). Those authors concluded that the incidental findings were too infrequent to justify screening healthy asymptomatic patients.<sup>3</sup> Finally, in a search of a radiologic database that included 46,414 patients, Lin et al. found a 0.02% prevalence of incidental VS cases,<sup>4</sup> comparable to the 0.03% reported by Morris et al. Also, Lin et al. indicated that the incidental detection of VS may be less prevalent than previously reported in temporal bone studies, where rates as high as 2.4% were cited.<sup>4</sup> All eight incidental VS cases reported in the study by Lin et al. were discovered on contrastenhanced MRIs. Among those eight patients, audiometric data were available for 7 patients, and asymmetric hearing loss was diagnosed in 3 patients.<sup>4</sup> Taken together, these studies indicated that the prevalence of incidental VS varies among different studies. Prevalence estimates have ranged from  $0.02\%^8$  to  $0.2\%^{2-4}$  in brain MRIs and were as high as 2.4% in temporal bone studies.<sup>4</sup>

In contrast, the incidence of VS was reported to be 20 per million (or 2 per 100,000) individuals per year.<sup>69</sup> This estimate was 100-fold lower than the prevalence of 2 per 1000 reported by Vernooij *et al.*<sup>2</sup> This discrepancy might be explained by the fact that up to 70% of VSs are non-growing<sup>9,10</sup> and up to 15% of patients with VSs have normal or symmetric hearing, which makes diagnosing a VS quite challenging.

Several diagnostic tools are currently used for VS screening. Contrast-enhanced brain MRI is currently considered the primary diagnostic tool for excluding VS.<sup>5</sup> Contrast-MRI is capable of detecting tumors as small as 4 mm, and it does not expose the patient to radiation.<sup>6</sup> Diagnostic yields of 100% are reported, with no false-positive or false-negative results.<sup>6,7</sup> Computed tomography (CT) is also commonly used. CT is able to detect acoustic tumors as small as 2 cm. However, because CT cannot detect intracanalicular tumors, it is an inadequate diagnostic tool for excluding VS.7 Studies that implemented CT with gas cisternography improved the sensitivity of detecting intracanalicular tumors, but they cited up to 22% false-positive findings, which resulted in unnecessary surgical explorations.7 Finally, ABR testing was initially thought to be a promising screening tool for VS;7 however, ABR has a notable shortcoming: it cannot obtain accurate results when a patient's hearing threshold is >80 dB at 4 kHz.<sup>7</sup> The sensitivity of ABR for detecting VS ranges from 63-95%; its sensitivity was 100% for tumors >2 cm, but the sensitivity decreased to 58% for tumors  $\leq 1 \text{ cm}$ .<sup>7</sup> Therefore, ABR is an inadequate screening tool for detecting VS.

Currently, brain MRI is the gold standard tool for diagnosing patients with suspected VS.6 Among patients that presented with audiovestibular symptoms, brain MRIs have detected VS in 0.5-1.4%<sup>5,6,8</sup>; however, audiovestibular symptoms associated with VS vary greatly among patients; thus, it is difficult to make correct MRI referrals. Patients with VS typically present with unilateral hearing loss or ASHL<sup>6</sup>; however, as reported by Saleh *et al.*<sup>11</sup> and Stucken *et al.*,<sup>7</sup> these symptoms are not always present. In those studies, 3-12.5% of VSs were found in patients with a normal pure tone audiogram,<sup>11</sup> and 1-15% were found in patients with normal or bilaterally symmetric hearing.7 Furthermore, Saleh et al. found no significant difference in tumor size among patients with normal hearing compared to those with hearing loss.<sup>11</sup> It should be noted that, in the study by Saleh et al., among the 16 patients that had a VS, despite a

normal pure tone audiogram, 6 had received a contrast-CT scan prior to the MRI. In two patients, the CT scan showed no abnormalities, which illustrated why MRI is considered the gold standard diagnostic tool for VS.

The most common complaints among the patients with VS, despite a normal audiogram, were tinnitus, disequilibrium, sudden hearing loss that recovered, and subjective hearing loss.<sup>11</sup> Furthermore, poor speech discrimination, particularly when disproportionate to the results from pure tone audiometry, could be an indicator of VS.7 Thus, VS is associated with a wide range of audiovestibular and other symptoms. The decision of whether to conduct an MRI is currently based on different referral protocols. In a study on the diagnostic yield of MRI for audiovestibular dysfunction with contemporary referral criteria, Vandervelde et al. included the following criteria for MRI referral: unilateral or asymmetric audiological clinical findings; clinical audiovestibular dysfunction with asymmetric audiometry; or central vestibular clinical findings.5 However, no consensus could be reached on the degree of asymmetry required for a MRI referral.<sup>5</sup> The degree of asymmetry in the study by Vandervelde et al. ranged from "any" asymmetry to > 10 dB at three frequencies on puretone audiometry.<sup>5</sup> Newton et al. attempted to design a referral protocol for brain MRI based on audiometric data in cases of ASHL.6 Those authors tested two protocols that had previously existed (the Oxford and the Charing Cross protocols) on 132 patients with ASHL and 30 patients that had been diagnosed with VS. The Oxford protocol suggests a referral for brain MRI when a minimum of 15 dB interaural difference is detected at any two average frequencies between 250 Hz and 8000 Hz; the Charing Cross protocol suggests screening for VS when, at two adjacent frequencies, an interaural difference is detected that is >15 dB for a patient with unilateral hearing loss and > 20 dB for a patient with bilateral asymmetric hearing loss. Newton et al. found that 13% of VSs were missed when applying the Oxford protocol and 23% were missed when applying the Charing Cross protocol. No protocol could identify 10% of patients with a proven VS.6 It is also worth mentioning that the Oxford protocol was the only to suggest an upper age limit of 70 years, on the grounds that operating on aged patients would not necessarily improve their health status.6

In addition, it seems to be difficult to find a protocol for determining the presence of ASHL based on a pure-tone audiogram.<sup>12</sup> Margolis et al. attempted to create an algorithm that could identify asymmetric hearing loss among a database of audiograms. Five expert clinicians were asked to classify 199 audiograms as symmetric or asymmetric.<sup>12</sup> They found no agreement on asymmetry among the judges in almost one quarter of the cases. When the algorithm was applied to the same data, it provided more accurate diagnoses than the consensus of the judges. The algorithm identified ASHL in 55% of all patients, and up to 78% in patients with severe hearing losses.<sup>12</sup> Since asymmetry in hearing is so common, it is virtually impossible to make it a sound protocol for referral for brain MRI to exclude VS.

The test-retest reliability of pure tone audiometry is rather poor. Nearly 87% of thresholds on the retest are within 5 dB of the first test,<sup>13</sup> and 10 dB differences occur roughly 10% of the time.<sup>13</sup> Thus, when thresholds for six frequencies are assessed in each ear, 14% of patients tested would be expected to have a difference  $\geq$  15 dB at more than one threshold.<sup>13</sup> This inaccuracy further complicates the application of audiometric based protocols.

Regarding the two cases brought forward in this article, none of the referral protocols mentioned above would have provided a correct identification in the first case. The first patient had no audiovestibular complaints and a normal, symmetric pure-tone audiometry. In contrast, one referral protocol could have correctly identified the second case. The protocol by Urban *et al.*<sup>14</sup> defined ASHL as an interaural difference in pure-tone thresholds  $\geq 10$  dB at two frequencies, or  $\geq 15$  dB at one frequency.<sup>14</sup> The second patient had an ASHL of 10 dB on two adjacent frequencies

#### Conclusion

Brain MRI is considered the primary diagnostic tool for VS<sup>5</sup>. However, it has become apparent that current protocols used for MRI referrals, based on ABRs or other audiologic criteria, are not costeffective.<sup>5-8</sup> Current protocols may result in missing up to 10% of tumors. However, a timely VS diagnosis is crucial for promoting a positive outcome after surgery or radiotherapy for hearing preservation. Consequently, physicians often refer more patients for MRI than strictly necessary, due to the inaccuracy of current referral protocols, and a fear of litigation due to late diagnoses. Additionally, current referral protocols are complicated by the fact that no consensus has been achieved among referring doctors on the degree of asymmetry required for a referral for brain MRI. Moreover, pure tone audiometry is quite subjective, and test-retest reliability is poor. The overall prevalence of ASHL in an audiometric database was 55% among all patients and up to 78% among patients with severe hearing losses, making it extremely challenging to design an accurate protocol for referral for brain MRI to exclude VS. In light of these difficulties, the author suggests that, despite the view that the low number of incidental findings may not justify the additional cost, it may be better to refer every patient with audiovestibular symptoms for a brain MRI.

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