False Negative Auditory Brainstem Response Findings in Vestibular Schwannoma: Case Reports

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Abstract
The false negative rate of the auditory brainstem response (ABR) for the detection of vestibular schwannoma (VS; acoustic neuroma) has been reported to range from 2 to 9 percent. The introduction of magnetic resonance with gadolinium contrast (Gd MRI) may be expected to raise the false negative rate, because smaller tumors can now be detected. We present two cases that illustrate some of the complex issues raised by the detection of small asymptomatic VS by Gd MRI that were missed by ABR. The role of ABR testing in the diagnosis and management of VS remains well established and will continue to evolve.

Key Words: Acoustic neuroma, auditory brainstem response (ABR), false negative, vestibular schwannoma

Over the past 2 decades, clinicians have included auditory brainstem response (ABR) testing in their test battery to help identify vestibular schwannomas (VS; acoustic neuroma). The reported "hit rate" in detecting tumors of the eighth cranial nerve (CN VIII) associated with ABR is as high as 96 to 100 percent (Hall, 1992). Although ABR testing is a powerful clinical tool, clinicians should be aware of the possibility of obtaining false negative results. In the present report, a false negative ABR outcome refers to a normal ABR for a patient with a VS.

There have been a few reports documenting false negative findings associated with ABR testing (Bockenheimer et al, 1984; Legatt et al, 1988; Telian et al, 1989; Grabel et al, 1991). These observations have raised some concern among clinicians with respect to the sensitivity of the ABR for the detection of these tumors. At present, gadolinium contrast magnetic resonance imaging (Gd MRI) is regarded as the definitive technique for identifying VS (NIH Consensus Development Conference Consensus Statement, 1991). The following are two case reports that illustrate the possibility of obtaining a normal ABR in patients with VS, as identified by Gd MRI.

CASE REPORTS

Case 1

Patient 1 is a 53-year-old male with a 3.5 year history of hearing loss and tinnitus on the right side. In 1989, the patient underwent an MRI scan of the head that initially showed an 8 mm right intracanalicular lesion consistent with VS (Fig. 1 A and B). The patient was followed with serial MRI scans from 1989 to 1991. The lesion showed no change in size during the first year. During the second year, the lesion increased in size to fill the internal auditory canal and extended 2 mm medial into the porus acusticus and cerebellopontine angle (CPA). Although his 1991 audiogram showed a slight improvement, the patient continued to complain of tinnitus and aural fullness on the right side.
Preoperative audiometric testing (Fig. 2) showed a mild, high-frequency sensorineural hearing loss at 3000 Hz and above in the right ear and normal hearing sensitivity in the unaffected left ear. Word recognition scores were excellent bilaterally (96% for the right ear and 100% for the left ear). Tympanometry and ipsilateral and contralateral stapedial reflex thresholds were normal bilaterally. There was no stapedial reflex decay noted. Auditory brainstem response testing (Fig. 3) revealed bilaterally normal interwave intervals (wave I-wave V) and wave V/wave I amplitude ratios. There was no evidence of asymmetry on caloric or rotational testing (0.01–0.64 Hz).

Subsequently, the patient underwent resection of the VS. A middle cranial fossa approach was utilized with neurophysiologic monitoring to help preserve usable hearing.

Case 2

This healthy, 62-year-old male presented to the neurology clinic with a left Bell’s palsy. At that time, a Gd MRI scan was ordered. No mass was noted on the left side; however, a 2 x 3 mm intracanalicular lesion, consistent with VS, was an incidental finding on the right side (Fig. 4 A and B). The patient was referred to the Department of Otolaryngology–Head and Neck Surgery for assessment. Audiometric testing (Fig. 5) revealed bilaterally normal hearing sensitivity. Word recognition ability was excellent bilaterally (96% in the right ear and 100% in the left ear). Immittance measurements showed normal tympanometry and ipsilateral and contralateral stapedial reflex thresholds bilaterally. There was no stapedial reflex decay. Auditory brainstem response testing (Fig. 6) revealed bilaterally normal interwave intervals (wave I–wave V) and wave V/wave I amplitude ratios. This patient is being followed with audimetric evaluations and ABR and MRI scans.

DISCUSSION

It is important to differentiate between the clinical diagnosis and clinical detection of VS. Whereas the clinical diagnosis of VS is made histopathologically, the clinical detection of VS is currently made through the use of
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there have been several reported cases of false negative ABR findings in the detection of VS (Bockenheimer et al, 1984; Legatt et al, 1988; Telian et al, 1989; Grabel et al, 1991, Wilson et al, 1992). Most of the reported “misses” consisted of intracanalicular (as opposed to extracanalicular) VS. In the present report, we have added two additional cases to the subgroup of patients with VS who demonstrate false negative findings on ABR testing. A false negative ABR may be defined as the absence of a statistically significant physiologic effect of the tumor on auditory nerve and brainstem neural transmission.

Cases 1 and 2 are instances of true false negative ABR findings for the detection of VS. In both cases, the absolute and interpeak latency values and amplitude ratios were within 2.5 standard deviations of our laboratory norms. It should be noted that for Case 2 the wave I–V interpeak latencies approached our laboratory upper limits. However, in a clinical setting, decisions regarding normalcy are based on a dichotomy of “normal” versus “abnormal” (as opposed to “borderline abnormal”). The upper limit value is used to base this decision.

Case 1 is an example in which a false negative ABR result would have misled the clinician into thinking that the patient did not have a tumor. This patient was managed nonsurgically until sudden growth of the tumor was detected by Gd MRI. This resulted in a recommendation to the patient that he consider surgery before the opportunity for a hearing-sparing operation.

Figure 3 ABR examination for Case 1. Notice that ABR interpeak latencies are bilaterally normal, despite the presence of VS on the right side.

gadolinium-DTPA MRI (Gd MRI), which serves as the “gold standard.” Gadolinium-DTPA is a paramagnetic contrast agent that greatly enhances the intensity of tumor tissue on T1-weighted MRI scans (Jackler et al, 1990). In comparison with Gd MRI scanning, the ABR is an established, lower-cost test that will detect all but a few VS. Over the past 10-year period,
was lost. He chose to proceed with surgery, and
aidable hearing was preserved. If electro-
diagnostic studies had been relied upon for
tumor diagnosis, his tumor might not have been
detected until (1) his hearing declined further;
(2) the option of a surgical approach designed to
preserve hearing would have been lost; or (3) his
tumor threatened to disrupt the function of
other neural systems. This patient, however,
presented with characteristic complaints and
audiologic signs of retrocochlear disease. The
presence of intermittent vertigo, tinnitus, and
unilateral hearing loss created a high index of
suspicion that the CN VIII was compromised,
despite the normal ABR. This finding under-
scores the need to consider the entire diagnostic
profile (medical history and audiometric tests)
when evaluating patients, instead of focusing
only on electrodiagnostic test results. With ref-
erence to the use of audiometric asymmetry as
a referral criterion, Mangham (1991) has sug-
gested that patients be referred for MRI if the
interear difference in the mean of thresholds at
1000 Hz, 2000 Hz, 4000 Hz, and 8000 Hz ex-
ceeds 20 dB.

While the incidence of clinical VS is ap-
proximately 1 case per 100,000 population per
year (NIH Consensus Development Conference
Consensus Statement, 1991), autopsy series
have demonstrated a prevalence of small, asym-
ptomatic, histologic VS in 0.03 to 2.4 percent of
the population (Leonard and Talbot, 1970; Schu-
knecht, 1976). The discrepancy between the
rates of histologic and clinical tumors suggests
that at least some of the tumors detectable by
Gd MRI may not be clinically significant. This
could be true for Case 2.

Further, a significant number of these
tumors will not progress, or will progress at an
extremely slow rate of growth. Bederson et al
(1991) followed 70 VS patients with serial com-
puted tomographic and magnetic resonance
imaging scans for periods ranging up to 26
months. Forty percent of tumors showed no
growth, and 6 percent showed an apparent
small decrease in size. The remaining 54 per-
cent of tumors grew at mean rates of 1.6 mm and
1.9 mm in the first and second years respec-
tively. A few tumors grew at much higher rates.
Some patients whose tumors did not grow were
operated on because of a progression of neurologic
symptoms or findings.

Although Gd MRI is considered the "gold
standard" for the detection of VS, it has been
suggested that this radiologic technique may be
hypersensitive, showing greater detail than cli-
nically necessary, at least for some patients (NIH
have not revealed evidence of neurophysiologic where ABR and balance function test results ing tumor removal, resulted in the preservation tated the early detection of small (0.5 cm or than a false-negative ABR. It is clear, however, (e.g., intracranial surgery) are more significant of VS, the consequences of a false-positive MRI considered the "gold standard" for the detection it is possible that Gd contrast MRI (or MRI evaluation of the patient included both T2- and T2 - weighted imaging. Crain and Dolan (1990) reported the presence of bilateral internal auditory canal enlargement from Paget's disease that appeared radiologically as bilateral VS. The radiologic evaluation of the patient included both T2- weighted MRI and air-contrast CT scan. Although these false-positive occurrences are rare, it is possible that Gd contrast MRI (or MRI alone) could "mislead" the clinician into diagnosing a VS when none exists. Since Gd MRI is considered the "gold standard" for the detection of VS, the consequences of a false-positive MRI (e.g., intracranial surgery) are more significant than a false-negative ABR. It is clear, however, that the availability of the Gd MRI has facilitated the early detection of small (0.5 cm or less), asymptomatic VS. In many of these instances, the early detection of the small tumors, coupled with auditory system monitoring during tumor removal, resulted in the preservation of useful hearing postoperatively.

In cases where VS has been identified and where ABR and balance function test results have not revealed evidence of neurophysiologic abnormality, and where the neurologic examination is normal, one might ask if the structural abnormality (e.g., the tumor) represents a significant health risk? In this regard, the identification of VS does not mandate its removal. The advisability of surgery to remove a VS and the choice of surgical approach are best determined by the patient and physician together, taking into account a number of factors. These include the size, location, and growth rate of the tumor; the presence of brainstem compression; the results of balance function testing; the progression of symptoms; age and general health; the prospects for preservation of usable hearing; and the general wishes of the patient. Thus, only a thorough analysis of all these factors can distinguish clinically important data from insignificant false negative ABR data.

REFERENCES


