Acoustic Neuroma in An Adolescent Without Neurofibromatosis: Case Study

Janet P. Sells*
Raymond M. Hurley†

Abstract

When acoustic neuromas are found in younger age groups, they are typically bilateral and associated with neurofibromatosis (NF). A unilateral acoustic neuroma in a child or adolescent without NF is rare. We report audiometric, auditory brainstem response (ABR) and magnetic resonance imaging (MRI) data for a 15-year-old male with an acoustic neuroma but without associated NF. The patient was seen for audiologic assessment on three separate occasions over a period of 5 years. The first two assessments produced unremarkable audiometric and immittance data. The third assessment, subsequent to a failed school hearing screening, demonstrated a flat unilateral sensorineural hearing loss, unilateral absent acoustic reflexes, and abnormal bilateral ABR recordings. MRI with contrast demonstrated a unilateral mass extending out of the internal auditory meatus. Microscopic examination of the removed tumor confirmed a vestibular schwannoma. An enhanced MRI 3 years post neuroma removal demonstrated no evidence of tumor regrowth and no evidence of other neoplasms. ABR recordings for the uninvolved ear continued to demonstrate neural synchrony as evidenced by normal absolute wave latencies; however, the III–V and I–V interwave latencies remained extended beyond the 99th percentile. Postoperatively, the young man was fitted with a high gain in-the-ear hearing aid in the involved ear, which he has continued to wear on a daily basis for the past 3 years.

Key Words: Acoustic neuroma, acoustic reflex thresholds, auditory brainstem response (ABR), immittance measures, sensorineural hearing loss, vestibular schwannoma

While the annual incidence of unilateral acoustic neuromas (vestibular schwannomas) is approximately 1 per 100,000 in the general population, it is not an expected finding in an adolescent without neurofibromatosis (NF). Of the 2000 to 3000 acoustic neuromas diagnosed annually, the vast majority are identified in individuals 30 years of age or older (Jackler and Pitts, 1990). When acoustic neuromas do occur in children and adolescents, they are associated with one of the two types of NF. Table 1 outlines the diagnostic criteria for NF-1 and NF-2. It is estimated that approximately 2 percent of the NF-1 cases have unilateral acoustic neuromas with even fewer NF-1 cases having bilateral acoustic neuromas. Acoustic neuromas are more commonly associated with NF-2, an autosomal dominant disorder with a high degree of penetrance and an onset between 20 and 40 years of age. NF-2 accounts for 5 percent of all acoustic neuromas (Listernick and Charrow, 1990). After eliminating acoustic neuromas associated with NF, Mattucci et al (1987) identified only 12 previously reported cases of a unilateral acoustic neuroma without NF in patients under 16 years of age (Craig et al, 1954; Bjorkesten, 1957; Rushworth et al, 1984). Our more recent literature search uncovered three additional reports of these cases (Krause and McCabe, 1971; Anderson and Bentik, 1972; Graham, 1979). Thus, acoustic neuromas not associated with NF are a rare occurrence in young patients.

The classic presenting symptom of a patient with an acoustic neuroma is asymmetric hearing sensitivity and/or perception that may or may not be accompanied by unilateral or asym-

*Audiology/Hearing Conservation, Naval Hospital, Newport, RI; and †Kresge Hearing Research Laboratory, Department of Otorhinolaryngology & Biocommunications, Louisiana State University Medical Center, New Orleans, Louisana

Reprint requests: Raymond M. Hurley, Kresge Hearing Research Laboratory, Department of Otorhinolaryngology & Biocommunications, Louisiana State University Medical Center, 2020 Gravier, Suite "A", New Orleans, LA 70112-2234

Table 1 Summary of Diagnostic Criteria for Neurofibromatosis Types NF-1 and NF-2

NF-1 is characterized in Caucasians by two or more of the following:

1. Six or more cafe-au-lait spots
2. Two or more neurofibromas
3. Axillary or inguinal freckling
4. Optic glioma
5. Osseous lesion such as sphenoid dysplasia
6. Two or more Lisch nodules
7. A first-degree relative with NF-1

NF-2 is characterized by one or more of the following:

1. Bilateral VIII nerve tumors
2. A first-degree relative with NF-2 and either an acoustic neuroma or one of the following: (a) neurofibroma; (b) meningioma; (c) glioma; (d) schwannoma; (e) posterior capsular cataract; or (f) early opacity

Adapted from Acoustic Neuroma (1991).

metric tinnitus with or without disequilibrium. While these symptoms have been used to describe the quintessential acoustic neuroma patient, only 5 percent of patients with these symptoms will be found to have an acoustic neuroma (Swan, 1989; Kotlarz et al, 1992). The preponderance of acoustic neuroma patients will have a gradually progressive unilateral sensorineural hearing loss (SNHL). In a study of 66 patients, Moffat et al (1989b) found 56 percent to have a high frequency SNHL, 25.5 percent to have no residual hearing, 15 percent to have flat losses, 3 percent to have normal hearing, and 1.5 percent to have trough-shaped SNHL. Further, it is estimated that 10 to 15 percent of acoustic neuroma patients present with a sudden hearing loss rather than the typical gradual SNHL (Yanagihara and Asai, 1993). These findings are in good agreement with a previous review of 500 cases by Johnson (1977).

The growth rate of an acoustic neuroma is now known to vary from virtual dormancy to a rapid growth of about 1 cm/year (Moffat et al, 1989b; Swan, 1989; Lanser et al, 1992; Selesnick and Jackler, 1992). For a moderate growth rate of 0.5 cm/year, Swan (1989) estimates the tumor would take approximately 2 years to fill the internal auditory meatus (the intracanalicular stage), and another 3 years of growth to compress the brain stem (the cisternal stage), at which time the tumor is 2 to 3 cm in size. There is some speculation on why some tumors appear dormant or shrink in size while others rapidly increase in size. Kasantikul et al (1980) suggest that growth rates may follow different time courses based on age, the growth rate

during the first and second decades of life being accelerated. Further, Wiet et al (1989) speculate that pregnancy and puberty may cause rapid enlargement of the neuroma.

The prevalence of associated neurologic symptoms accompanying an acoustic neuroma may be affected by age. Selesnick et al (1993) argue that younger patients may have less presenting symptoms than older patients due to the ability of the cranial nerves to withstand more stretching and compressing before neurologic symptoms appear. They also suggest that the lack of reported symptoms in younger patients may be due to a tendency to ignore medical problems.

CASE REPORT

The following case study is an example of a unilateral acoustic neuroma occurring in an adolescent who was asymptomatic except for a mild unilateral hearing loss. The patient's retrocochlear dysfunction was identified at an audiologic assessment precipitated by a failed school hearing screening. Previous audiometric findings were normal. Specifically, the patient is a male Caucasian who was originally seen at 11 years of age for pure-tone audiometry and immittance testing because of a previous history of middle ear effusion. The results of this assessment are displayed in Figure 1. At 13 years of age, he was again assessed to monitor his hearing and middle ear status. Conventional audiometric and immittance testing were normal at this time and are summarized in Figure 2. At age 15, he was assessed subsequent to a failed school hearing screening, at which time he demonstrated normal hearing in the right ear and a mild sensorineural hearing loss of flat pure-tone configuration in the left ear. The speech recognition scores for NU-6 materials were 100 percent bilaterally. Immittance testing demonstrated normal tympanograms bilaterally with normal ipsilateral acoustic reflexes (ARs) and crossed ARs consistent with the hearing levels in the right ear (Silman and Gelfand, 1981) and absent ipsilateral and crossed ARs for the left ear. These test results are summarized in Figure 3. Auditory brainstem response (ABR) recordings, displayed in Figure 4, were elicited by 100 µsec clicks. The ABRs were obtained with a two-channel electrode montage, Fz-A$_1$ and Fz-A$_2$, at a bandpass filter setting of 100–3000 Hz. Each summed waveform was the result of 2000 stimulus presentations. At a presentation rate of 11.4/sec and an
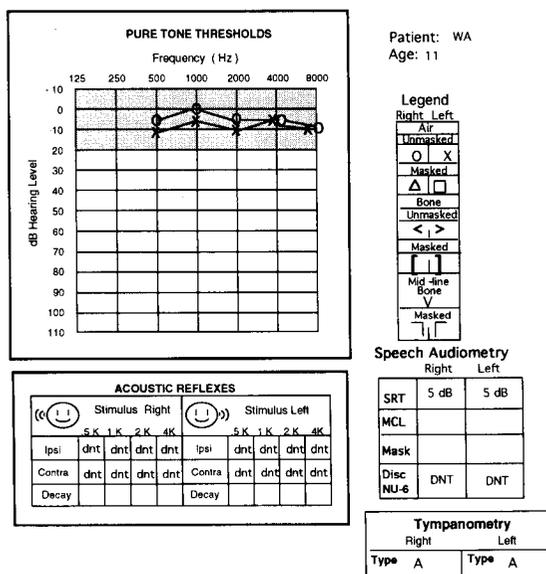
Figure 1 Audiologic test results at age 11 showing normal hearing and normal tympanograms.

Figure 2 Audiologic test results at age 12 showing normal hearing and normal immittance results.
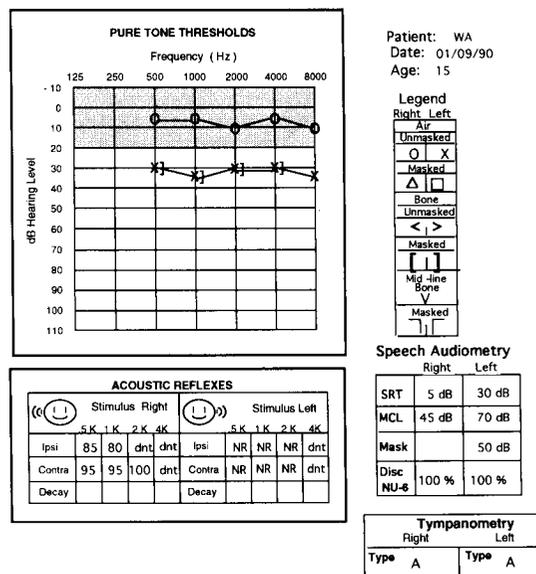

Figure 3 Audiologic test results at age 15 showing normal hearing and normal acoustic reflexes in the right ear and a mild sensorineural hearing loss and absent acoustic reflexes in the left ear.
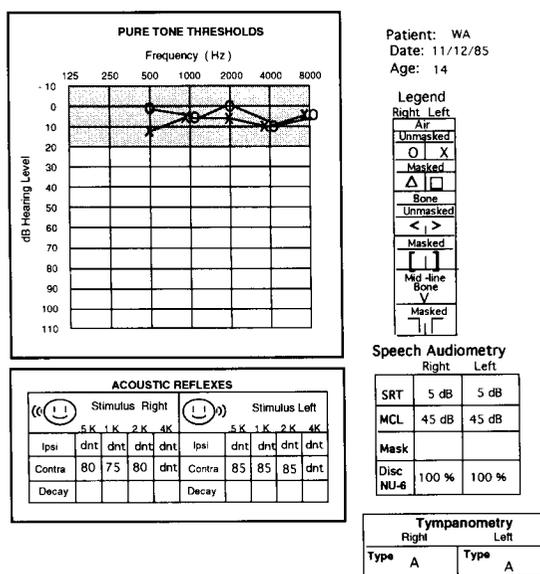
intensity level of 75 dB nHL, the right ear latency values for waves I, III, and V were 1.66 msec, 3.72 msec, and 6.16 msec, respectively, and the I–III, III–V, and I–V interpeak latencies were 2.06 msec, 2.44 msec, and 4.50 msec, respectively. While the absolute latencies for the right ear are within the 99th percentile for normal hearing ears, the III–V and I–V interpeak latencies exceed the 99th percentile for normal hearing ears (Joseph et al, 1987). At a presentation rate of 11.4/sec and an intensity

level of 95 dB nHL, the left ear latency values for waves I, III, and V were 1.60 msec, 4.30 msec, and 8.94 msec, respectively, and the I–III, III–V, and I–V interpeak latencies were 2.70 msec, 4.64 msec, and 7.34 msec, respectively. The absolute latencies of waves III and V exceed the 99th percentile for ears with SNHL as did the interpeak latencies for I–III, III–V, and I–V (Joseph et al, 1987). The above values for V and the I–V interval are based on our designation of the last wave as V, although arguably our designated V could be Po of the middle latency response complex with V being absent.

Following the ABR recordings, a gadolinium-enhanced magnetic resonance imaging (MRI) study (see Fig. 5) identified a mass extending out of the left internal auditory meatus and compressing the brain stem. Detailed medical and neurologic evaluations ruled out NF. In fact, this patient did not exhibit a single characteristic of either NF-1 or NF-2. At surgery, a 3-cm tumor was confirmed and removed using a suboccipital approach. Hearing in the left ear was not preserved. Microscopic examination of the mass identified Antoni Type A cells and confirmed an acoustic neuroma (vestibular schwannoma).

A summary of the 3-year postoperative audiometric results are displayed in Figure 6 and show a complete hearing loss in the left ear and
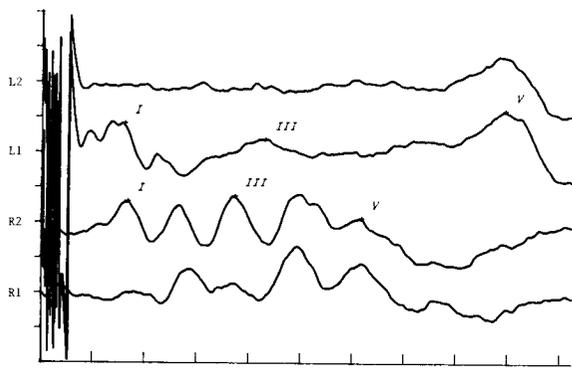
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Figure 4 Auditory brainstem responses at age 15. Tracings R1 and R2 are the contralateral and ipsilateral recordings for the right ear, respectively. Tracings L1 and L2 are the ipsilateral and contralateral recordings for the left ear, respectively. Time is 1 msec/division and amplitude is 0.31 μV/division.

Figure 5 Magnetic resonance imaging study at age 15 showing a left-sided tumor.

normal hearing in the right ear. The 3-year postoperative ABR recordings are displayed in Figure 7 showing right ear latencies for waves I, III, and V of 1.40 msec, 3.60 msec, and 5.96 msec, respectively, and I–III, III–V, and I–V interpeak latency values of 2.20 msec, 2.36 msec, and 4.56 msec, respectively. The III–V and I–V interpeak latency values continue to exceed the 99th percentile for normal hearing ears (Joseph et al, 1987). Figure 8 displays a 3-year postsurgical computerized tomography scan, which was interpreted as showing no evidence of tumor regrowth on the left and no suspicion of a tumor on the right.

To compensate for the total hearing loss, this patient's "dead" ear was fitted with a high gain in-the-ear hearing aid 90 days postsurgery. The transcranial CROS hearing aid arrangement was fitted following the procedures outlined by Sullivan (1988). While the exact benefit of a transcranial CROS hearing aid is difficult to quantify beyond the hearing test booth, Sullivan's protocol predicted that this hearing aid arrangement would provide sufficient transcranial sound conduction to restore some aspects of bilateral hearing. After 3 years, this patient continues to wear his hearing aid at least 8 hours a day and recently replaced the aid at his own expense when it was lost.

COMMENT

This case illustrates the fact that acoustic neuromas do appear in adolescents, and do appear in adolescents without NF, although the combination of an acoustic neuroma without associated NF is rare. Rare, however, does not mean nonexistent. Thus, audiologists must be
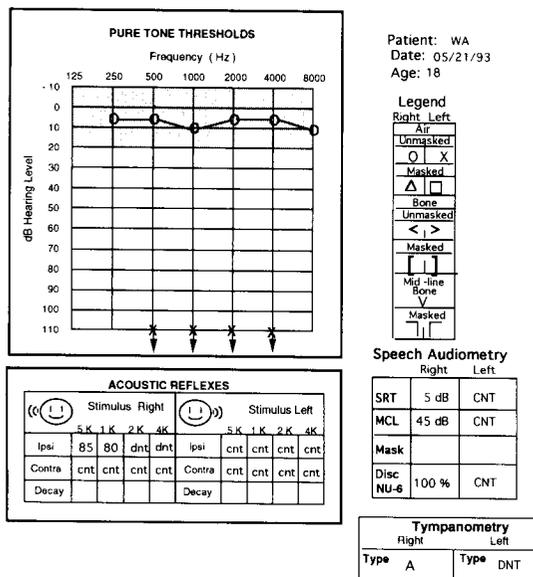

Figure 6 Audiologic test results 3 years postsurgery showing normal hearing and normal ipsilateral acoustic reflexes in the right ear and no hearing in the left ear.

sensitive to the fact that cases do present that fall outside the expected parameters in age and in symptoms.

In this particular case, the two test results that mandated further assessment, regardless of age and/or the absence of associated symp-
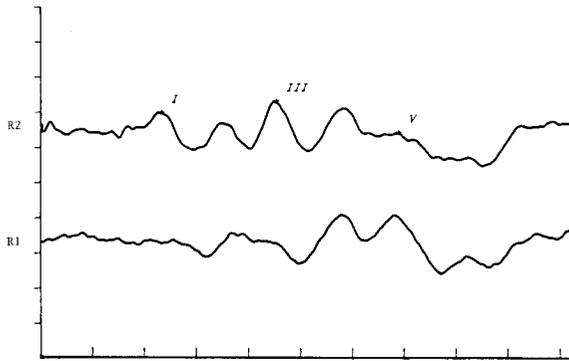
Figure 7 Auditory brainstem response 3 years postsurgery. Tracings R1 and R2 are the contralateral and ipsilateral recordings for the right ear, respectively. Time is 1 msec/division and amplitude is 0.31μV/division. Insert tube phones were used for this recording; thus, the absolute latency values were adjusted by 0.9 msec.

Figure 8 Computerized tomography study 3 years postsurgery.

toms, were a unilateral SNHL and absent ARs. While the contour of the unilateral SNHL was flat, a shape that occurs in less than 15 percent of cases with acoustic neuromas, the appearance of a SNHL in a previously normal hearing ear in itself should raise suspicions. Further, even though conventional AR threshold measures have a low "hit" rate (65%) and a high "false alarm" rate (17%) for the identification of acoustic neuromas (Fowler et al, 1993), absent ARs cannot be ignored. Clearly, an unexpected unilateral SNHL with absent ARs must be followed by an ABR.

Speculatively, this case may be an example of accelerated tumor growth in adolescents, as suggested by Kasantikul et al (1980) and Wiet

et al (1989). Even though the acoustic neuroma was large enough to compress the brain stem and affect the contralateral ABR and the crossed ARs, the patient was neurologically asymptomatic. Perhaps this lack of associated symptoms is a reflection of the patient's age and the resilience of the brain stem in younger age groups, although any given patient with an acoustic neuroma may be neurologically asymptomatic (Selesnick et al, 1993).

Contralateral ABR effects are not an unusual finding in large acoustic neuromas, > 2.5 cm, possibly due to brainstem compression (Nodar and Kinney, 1980; Moller and Moller, 1983; Musiek and Kibbe, 1986; Johnson and Selters, 1987; Moffat et al, 1989a). While the most frequent contralateral ABR abnormality is a prolongation of the III–V interpeak interval that occurred in this case, the contralateral ABR effect can also be reflected in a prolongation of the I–III and I–V interval. In this case, both the III–V and I–V interpeak intervals were prolonged. Unfortunately, we were not able to find data to determine if the continued prolongation of the III–V and I–V interpeak interval is a typical postoperative finding in large acoustic neuromas.

An interesting feature of this case is the avenue of referral. A failed school hearing screening started the diagnostic sequence—clearly, not the normal path that an acoustic neuroma case follows to diagnosis and treatment. This feature of the case reinforces the need for audiologists to be vigilant, as we are often the entry point into the health care system for a variety of auditory disorders.

Lastly, this case demonstrates that the audiologist's responsibility does not end with the identification of the acoustic neuroma. Audiologists should be willing to address the rehabilitation needs of the patient subsequent to the sudden loss of normal auditory function produced by a total unilateral loss of hearing. While the transcranial CROS hearing aid is not an aural rehabilitation panacea for total unilateral hearing loss, our experience with this case had convinced us that it deserves consideration and a trial period to determine if an individual patient can benefit from this type of hearing aid arrangement.

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