Short report

Auditory dysfunction in Ramsay Hunt syndrome

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SUMMARY A 48-year-old woman with a Ramsay Hunt syndrome due to herpes zoster had a hearing deficit. Brainstem auditory evoked potentials (BAEPs) localised the site of dysfunction to the ipsilateral eighth nerve. Clinical improvement was associated with improvement of the BAEP. Conventional audiological studies and BAEPs provided no evidence of involvement of the cochlea or the brainstem. In Ramsay Hunt syndrome, BAEPs may help to localise the site of involvement within the auditory pathway and follow the course of the disease.

Ramsay Hunt syndrome is characterised by a herpetic vesicular eruption in the external auditory canal or the auricle associated with facial paresis. Hunt initially attributed the syndrome to a herpes virus infection of the geniculate ganglion, but subsequent studies indicate that an interstitial neuritis of the seventh nerve is a more common cause of the facial dysfunction.¹ Vestibular and acoustic symptoms may also occur but, as is the case with other manifestations of the disease, the site of the lesions responsible for these symptoms is uncertain since pathological confirmation has seldom been obtained. Hearing loss has been attributed to involvement of the ganglion of Corti,²⁻⁷ the auditory nerve,²⁻⁷ or the brain stem.^{5,6} This is a report of a patient with herpes zoster auricularis, facial palsy, auditory and vestibular symptoms in whom brain stem auditory evoked potentials (BAEPs) demonstrated eighth nerve dysfunction that subsided as hearing impairment improved.

Case report

A 48-year-old woman with polycystic kidney disease and chronic renal failure developed nausea, vomiting, vertigo and severe pain in the left ear and mastoid region that was followed several hours later by complete paralysis of the left forehead and face. She also noticed some tinnitus and hyperacusis in the left ear. Two days after the onset of symptoms she developed a vesicular eruption in the left external audi-

tory canal. Examination demonstrated pain to pressure in the left mastoid area, a complete left peripheral facial palsy, and decreased taste in the anterior two-thirds of the tongue on the left side. Gait was unsteady and she veered to the right. CSF examination and head CT scan without contrast were normal. Rinné test was normal bilaterally. Audiological evaluation of the right ear, including pure tone audiometry, tone decay, speech discrimination and impedance audiometry, was normal. There was mild to moderate decreased hearing in the left ear for frequencies of 1000 Hz or higher. Speech discrimination was fair (72%) at 35 dBSL and poor (46%) at 45 dBSL, a performance versus intensity function for phonetically balance words (PI-PB) indicative of "rollover" effect. Tone decay at 2000 Hz was negative. Left ear impedance could not be tested because a seal could not be maintained due to severe pain in the left mastoid area. BAEPs were done using standard stimulation and recording procedures.⁸ With left ear stimulation the peak latency of wave I was within normal range but subsequent waves displayed a moderate latency prolongation. The I-III, and I-V interpeak latencies were abnormally prolonged but the III-V interpeak latency was normal (fig). BAEPs to right ear stimulation were normal.

The tinnitus and hyperacusis gradually subsided over the subsequent weeks, as did the pain behind the left ear and the deficiency in taste. However, the motor function of the face had shown no improvement on follow-up examination 10 weeks after the onset of illness. Audiological evaluation of the right ear remained normal. The left ear showed normal hearing for frequencies up to 3000 Hz with a mild decrease for higher frequencies. Speech discrimination, tone decay and tympanometry were normal. Acoustic reflexes were present probe right; reflexes were present probe left with ipsilateral stimulation but did not emerge with contralateral stimulation until an intensity of 115 dB was reached. BAEPs to stimulation of the left ear showed wave I with the same latency as in the initial examination but the latencies of waves II, III, and V were approximately 0.5 ms shorter (fig).

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Fig BAEPs to left ear stimulation at the onset of illness (top traces) and 10 weeks later (bottom traces). Note at 10 weeks the decrease in the peak latencies of all potentials except wave I, and in the I-II, I-III, and I-V IPLs. The latency of wave I and the III-V IPL, initially normal, did not change. Two consecutive averages of 2000 responses each recorded from vertex-to-ipsilateral earlobe derivations are superimposed (recordings from vertex-to-contralateral earlobe derivations, not shown in the figure, were simultaneously obtained). Upper limits of normal latencies, in ms (mean + 3 SD) are: I-2.2; II-3.3; III-4.5; V-6.5; I-III-2.7; III-V-2.4; I-V-4.7.

The I-III IPL was still slightly prolonged, but the I-V IPL was within normal limits. The III-V IPL, initially within normal limits, remained unchanged. BAEPs to right ear stimulation continued to be in the normal range. Three months later, the patient, then living overseas, reported in a letter that the facial palsy was gradually resolving.

Discussion

This patient suffered from a Ramsay Hunt syndrome with herpes zoster oticus and peripheral facial palsy accompanied by tinnitus and sensorineural hearing impairment. The initial BAEPs showed a normal wave I and approximately equal prolongation of all subsequent waves. Wave I is the eighth nerve com-

pound action potential in the segment of the nerve close to the cochlea, wave II reflects activity either in the intracranial portion of the auditory nerve close to the brain stem or in the area of the cochlear nuclei, and wave III in the superior olivary complex.^{9,10} An abnormal separation between wave I and waves II and III, therefore, suggest a retrocochlear, extra-axial auditory dysfunction, that is, a conduction defect in the eighth nerve. Conventional audiological evaluation was also consistent with an eighth nerve site of dysfunction.¹¹ Interpeak latencies of BAEP waves generated within the brain stem (II to V) were within normal range suggesting a normal conduction along the intra-axial brain stem auditory pathways.

As many as 90% of cases of Ramsay Hunt syndrome have vestibular disturbances but auditory symptoms occur less often.¹² The most frequent auditory symptom is hyperacusis, which occurs in about one-third of the cases, an incidence similar to that of patients with idiopathic facial palsy.¹³ Seventh nerve dysfunction with disruption of the innervation to the stapedius muscle rather than cochlear involvement may be responsible for this symptom. Sensorineural hearing loss, on the contrary, is observed in less than 10% of patients with Ramsay Hunt syndrome and virtually in no case of idiopathic facial palsy.¹³ The CSF in these patients is said to be usually abnormal with increased number of cells and protein content, reflecting a more widespread neurological involvement than in cases without auditory deficit.¹⁴ Our patient, however, had normal CSF and rather subtle cochlear symptoms without subjective hearing loss. Auditory involvement in Ramsay Hunt syndrome may occur more often than is commonly appreciated and may be overlooked when symptoms are subtle or overshadowed by other more prominent manifestations such as pain, facial palsy or vestibular symptoms.

The scarce number of pathological studies of patients with auditory impairment associated with Ramsay Hunt syndrome, usually done many months after the herpetic eruption has subsided, have shown lymphocytic and round cell infiltration of the auditory nerve as well as inflammatory changes in the organ of Corti and spiral ganglia.¹² Conventional audiological studies have suggested either cochlear or retrocochlear involvement.^{3,7} In our patient, BAEPs complemented conventional audiological studies and provided objective evidence for dysfunction in the left auditory nerve. There was, however, no evidence of involvement of the cochlea or the brain stem. Subsequent BAEPs obtained when the patient's hearing deficit had improved, documented an increase in conduction velocity along the auditory nerve that approached the range of normal. In patients with Ramsay Hunt syndrome BAEPs may be of assistance

in detecting involvement of the auditory system, localising the site of dysfunction and following the course of the disease.

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