Brain-stem somatosensory dysfunction in a case of long-standing left hemispherectomy with removal of the left thalamus: a nasopharyngeal and scalp SEP study

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Abstract

We have studied median nerve somatosensory evoked potentials (SEPs) in a patient who had undergone early surgical removal of the left cerebral hemisphere and left thalamus. Stimulation of the right side evoked normal latency P9, P11 and P13 potentials at scalp as well as at nasopharyngeal (NP) leads, while P14 and N18 potentials were absent. These SEP abnormalities, that have been described previously in cervico-medullary lesions and in comatose patients with upper brain-stem involvement, suggest that in our patient the removal of the left thalamus has caused retrograde degeneration of the cuneate-thalamic projections. Moreover, this study confirms that P13 and P14 potentials have different generators.

Keywords: Somatosensory evoked potentials; Hemispherectomy; Far-field potentials

1. Introduction

Far-field median nerve somatosensory evoked potentials (SEPs) can undergo selective alterations in focal lesions; for example, scalp P14 far-field potential, which is thought to be generated in the brain-stem tract of the lemniscal pathways (Desmedt and Cheron, 1981b), is frequently abnormal in lesions of the cervico-medullary junction (Mauguière and Ibáñez, 1985). The widespread scalp N18 response is also probably generated in the brain-stem, although its source and possible clinical utility are still a matter of debate (Convers et al., 1989; Mauguière and Desmedt, 1989; Urasaki et al., 1990; Tomberg et al., 1991).

The P14 potential is preceded by a P13 response of very similar latency; sometimes both potentials can be fused in a “P13-P14 complex” (Desmedt and Cheron, 1981a; Yamada et al., 1986). Further studies with an electrode array implanted near the thalamus and brain-stem structures have suggested a common origin in the medial lemniscus for these two waves (Hashimoto, 1984; Katayama and Tsubokawa, 1987). In contrast, some authors have reported the loss of P14 with preservation of a P13 response, suggesting an independent generator for the scalp P13 (Delestre et al., 1986; Kaji and Sumner, 1987). Recent studies using also nasopharyngeal recordings have observed this SEP abnormality in cervico-medullary lesions (Mavroudakis et al., 1993; Restuccia et al., 1995) and in comatose patients with clinical evidence of upper brain-stem involvement (Wagner, 1991), thus suggesting that the P13 generator may be located more caudally than the medial lemniscus.

Adult onset thalamic lesions do not affect brain-stem components of SEPs (Mauguière et al., 1983; Mauguière and Desmedt, 1988; Convers et al., 1989). In order to determine whether early thalamic lesions can involve brain-stem pathways, median nerve SEPs were studied in a patient who underwent left hemispherectomy with removal of the left thalamus at the age of 3. To address this issue,
we used a recording technique specific for brain-stem function assessment.

2. Materials and methods

2.1. Case report

The patient, an 18-year-old woman affected by Sturge-Weber syndrome, underwent surgical removal of the left cerebral hemisphere at the age of 3 because of right seizures that could not be controlled by antiepileptic drugs. She now takes 25 mg/day of phenobarbital and has not suffered seizures for 3 years. Clinical examination showed: facial nevus in the territory of the ophthalmic division of the right trigeminal nerve, right spastic hemiparesis, right hemisensory defect and right homonymous hemianopia. CT scan, performed when she was 6 years old, showed the absence of the whole left hemisphere as well as of the left thalamus (see Fig. 2).

2.2. SEP recording technique and control data

For SEP recording, the patient lay on a couch in a warm, semidark room. Stimuli (0.3 msec square-wave pulses, 5 Hz frequency) were delivered at the wrist using skin electrodes (cathode proximal) at motor threshold intensity. The amplifier filter bandpass was 30–3000 Hz (−3 dB at cut-off point, 6 dB per octave), analysis time was 32 msec (with a bin width of 125 μsec) including a pre-analysis period of 1 msec. Two averages of 2048 each were obtained and printed by the computer on an HP-Laserjet printer. The recording electrodes (impedance below 5 kΩ) were placed in the supraclavicular fossa (Erb’s point), referred to Fz, over the spinous process of the sixth cervical vertebra (Cv6), referred to an electrode located immediately above the thyroid cartilage, and at scalp locations Fz, P3, P4, and Oz, referred to the shoulder contralateral to the stimulated side. Lastly, we placed a homemade nasopharyngeal electrode above the upper wall of the pharynx, referred to the shoulder contralateral to the stimulated side. This technique, already described in previous papers (Tomberg et al., 1991; Wagner, 1991; Restuccia et al., 1995), permits the selective recording of brain-stem evoked responses. Our control data (see Table 1) were collected from 10 healthy subjects (6 men and 4 women, mean age 27.9 ± 3 years, range 25–35), using the same recording protocol.

3. Results (see Fig. 1)

Latencies of N9 and P9 responses, as well as of the cervical N13 potential, were bilaterally normal. After stimulation of the left median nerve, we could identify in all scalp traces 4 consecutive positive waves, which we labeled according to the literature as P9, P11, P13 and P14. The latency of the P13 potential was the same as that of the cervical N13 response. Latencies of all these potentials were within normal limits. Traces recorded at the contralateral parietal location showed a normal N20 response, and traces obtained from the ipsilateral parietal electrode showed a prolonged negativity starting at a latency of 13.7 msec, with 10.2 msec duration and a maximal amplitude of 2.5 μV, which we interpreted as an N18 response. Conversely, no scalp traces after stimulation of the right median nerve showed any clear response after the subcortical P13 potential, which had the same latency as the cervical N13 response. Nasopharyngeal traces have been subtracted “off-line” from the frontal recordings in order to better analyze the brain-stem P14 response. The P14 potential is obtained with its maximal amplitude in frontal leads and it is frequently absent in nasopharyngeal traces, whereas the P13 far-field response is recorded with about the same amplitude at both recording sites (Restuccia et al., 1995). Thus, this method enables cancellation of the P13 response, which is present in both traces, and enhancement of the P14 potential, that is seen as an “N14” negative response in the resulting waveform (Tomberg et al., 1991). Nasopharyngeal-to-forehead traces showed a clearly recognizable “N14” response after stimulation of the left median nerve, but no “N14” response after right median nerve stimulation.

Table 1

<table>
<thead>
<tr>
<th>Latencies (msec)</th>
<th>N9</th>
<th>P9</th>
<th>P11</th>
<th>P13</th>
<th>P14</th>
<th>N13</th>
<th>N20</th>
</tr>
</thead>
<tbody>
<tr>
<td>n Mean ± S.D.</td>
<td>----</td>
<td>----</td>
<td>-----</td>
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</tr>
<tr>
<td>Fz</td>
<td>10</td>
<td>9.13 ± 0.43</td>
<td>9</td>
<td>11.34 ± 0.55</td>
<td>8</td>
<td>12.91 ± 0.64</td>
<td>10</td>
</tr>
<tr>
<td>P3</td>
<td>10</td>
<td>9.12 ± 0.44</td>
<td>9</td>
<td>11.33 ± 0.53</td>
<td>8</td>
<td>12.9 ± 0.62</td>
<td>10</td>
</tr>
<tr>
<td>P4</td>
<td>10</td>
<td>9.12 ± 0.44</td>
<td>9</td>
<td>11.36 ± 0.53</td>
<td>8</td>
<td>12.89 ± 0.6</td>
<td>10</td>
</tr>
<tr>
<td>Oz</td>
<td>10</td>
<td>9.11 ± 0.41</td>
<td>9</td>
<td>11.34 ± 0.55</td>
<td>8</td>
<td>12.87 ± 0.6</td>
<td>10</td>
</tr>
<tr>
<td>C6-AC</td>
<td>10</td>
<td>8.96 ± 0.52</td>
<td>-----</td>
<td>-----</td>
<td>-----</td>
<td>-----</td>
<td>-----</td>
</tr>
<tr>
<td>Erb-Fz</td>
<td>10</td>
<td>10.2 ± 0.43</td>
<td>-----</td>
<td>-----</td>
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Fig. 1. A: median nerve SEPs from our patient. Thick traces: left median nerve SEPs. Thin traces: right median nerve SEPs. Erb-Fz: Erb’s point electrode referred to the forehead. C6-AC: sixth cervical vertebra electrode referred to an anterior cervical lead (AC). P4-Sh, P3-Sh, Fz-Sh, Oz-Sh, NP-Sh: right parietal, central parietal, left parietal, central occipital, nasopharyngeal electrodes, all referred to the shoulder contralateral to the stimulated side. Erb’s point recording shows bilaterally normal N9 potentials (8.4 msec latency). Cervical recording shows bilaterally normal N13 responses (11.3 msec latency). Scalp recordings after stimulation of the left median nerve show normal P9 (8 msec), normal P11 response (asterisk; 9.6 msec), normal P13 (11.3 msec), normal P14 (12.7 msec); contralateral parietal recording shows a normal N20 potential (17 msec). Scalp recordings after stimulation of the right median nerve show only normal P9, P11, and P13 potentials with the same latency as the contralateral side. Neither P14 nor N18 are identifiable in these traces. B: traces obtained by “off-line” subtraction of nasopharyngeal from frontal recordings, grand average of 4096 stimulations. The P14 response, which is obtained with maximal amplitude in frontal leads, is injected as a negativity in grid 2 of the preamplifier and is seen as “N14” in the resulting waveform. This “N14” response is evident after left median nerve stimulation, but is not apparent after right median nerve stimulation.

4. Discussion

Many reports have already described SEP abnormalities in adult onset thalamic lesions (Chiappa et al., 1979; Mauguïère and Courjon, 1981; Mauguïère et al., 1983; Mauguïère and Desmedt, 1988; Convers et al., 1989). When the ventro-postero-lateral nucleus is involved, upper limb SEPs typically show normal subcortical far-field potentials, with absent or delayed parietal N20 response. To our knowledge, there are no SEP studies in patients with early-onset thalamic lesions. In our patient with early hemispherectomy including the thalamus, several argu-
ments suggest a dysfunction of the brain-stem tract of the somatosensory pathways. First, neither the P14 nor the N18 scalp potentials were recorded after stimulation of the right median nerve. The loss of these two responses has been already described for focal lesions of the brain-stem tract of the lemniscal pathways (Delestre et al., 1986; Kaji and Sumner, 1987; Convers et al., 1989; Mavroudakis et al., 1993; Restuccia et al., 1995). Wagner (1991) also reported the loss of the scalp P14 in comatose patients and correlated this finding with rostro-caudal degeneration involving the upper brain-stem. Second, a normal P13 response with the same latency as the cervical N13/P13 was recorded from scalp and nasopharyngeal electrodes in our patient. Although earlier reports suggested a brain-stem origin for the P13 scalp far-field response (Hashimoto, 1984; Katayama and Tsukokawa, 1987), recent reports have shown that the P13 response can be preserved, in spite of P14 abnormalities, in focal lesions of the cervico-medullary junction (Mavroudakis et al., 1993; Restuccia et al., 1995) and that its generator is possibly located in upper cervical segments (Restuccia et al., 1995). Also, in comatose patients with rostro-caudal degeneration both scalp and nasopharyngeal recordings demonstrated the preservation of an early positive response that Wagner labeled as "caudal P14" (Wagner, 1991) and that possibly corresponds to our P13 potential.

The brain-stem dysfunction in our patient is probably due to retrograde Wallerian degeneration. Transsynaptic degeneration has been claimed by Mauguière and Desmedt (1989) to explain minor abnormalities of the P14 response in some patients with long-standing hemispherectomy, but none of their patients had undergone a complete removal of the thalamus as had ours.

So far, no pathologic study of retrograde degeneration following early thalamic lesions in humans has been reported. In cat, it has been ascertained that midbrain transection causes retrograde changes of the lateral cervical nucleus (LCN), projecting to sensory thalamic relays (Morin and Catalano, 1955; Seki, 1962; Grant and Westman, 1969). Thus, it can be hypothesized that the ablation of the axonal endings of the cuneate-thalamic projections in our patient has caused a severe Wallerian degeneration of the lemniscal pathways. We cannot determine the anatomical extension of this degeneration, but this mechanism is sufficient to cause a complete loss of the P14 and N18 potentials, as already observed in cervico-medullary lesions and in comatose patients with upper brain-stem involvement.

Recording of the P13 far-field has been necessary to localize in our patient the somatosensory dysfunction in the brain-stem. The clinical utility of this wave has been thus far limited, as scalp non-cephalic recordings often fail to clearly distinguish it from the following P14 response (Yamada et al., 1986; Restuccia et al., 1995). On the other hand, the finding of dissociated loss of P14 with still preserved far-field P13 clearly demonstrates that these two
waves have different generators (Delestre et al., 1986; Kaji and Sumner, 1987; Convers et al., 1989; Mavroudakis et al., 1993; Restuccia et al., 1995). Previous reports (Wagner, 1991; Restuccia et al., 1995) have suggested that nasopharyngeal leads can preferentially record the P13 potential. This potential has been recorded in our patient with the same latency from NP as well as scalp leads. As it was clearly recorded in our patient over all scalp locations, including occipital leads, the scalp P13 is unlikely to represent the anterior cervical P13 picked up by frontal electrodes, as suggested by Sonoo et al. (1990).

The recording of P13 and P14 far-field potentials using scalp and NP recordings has proved a useful tool in determining somatosensory dysfunction in the brain-stem. In our patient, the finding of isolated loss of P14 with still preserved P13 strongly suggests that early thalamic lesions can cause retrograde lemniscal degeneration.

Acknowledgments

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References


