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Characteristic EEG findings in childhood Moyamoya syndrome

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Abstract

Moyamoya syndrome-specific alterations of EEG recordings are only observed in children. These consist of a gradual frequency decrease and amplitude activation after hyperventilation. This is referred to as re-build-up phenomenon. Thus, a tentative diagnosis of Moyamoya syndrome in children can be based on the EEG finding. This should be radiologically confirmed by angiography.

Keywords: EEG in childhood, Moyamoya syndrome, re-build-up phenomenon.

1 Introduction

Moyamoya syndrome has rarely been diagnosed outside Asia. Because of its characteristic clinical symptoms, however, it should not be difficult to consider this disease in differential diagnoses in non-Asian countries.

The dominant feature in children is alternating hemiplegias reflecting transitory ischaemic attacks of varying duration, often induced by hyperventilation [2]. Hemiplegia may already occur when the child blows on a dish of hot soup to cool it, takes several deep breaths with emotional upheaval, or when sucking in long noddles [6]. Other symptoms include cerebral infarction and epilepsy, often focal in character. In adults, cerebral haemorrhage – frequently subarachnoid bleeding – is the main symptom [14].

2 Case report

Only in children can the EEG tracing offer a definite indication of Moyamoya syndrome even when clinical symptoms are missing, as described in the following:

An 11-year-old boy presented with school problems. He had been born as one of a pair of dizygotic twins two weeks prematurely, footling presentation, with the umbilical cord twisted twice. Apgar score was 1/5/7. The boy was the second twin born. The neonatal period was uneventful. At the age of 10 months he suffered a convulsion during fever and purulent tonsillitis. EEG recordings were not made at this point.

Physical examination showed marked bilateral muscular proprioceptive reflexes with widened reflex zones, pyramidal signs absent. Tests of coordination showed remarkable associated movements. Overall, the finding was minimum cerebral dysfunction with perinatal stress.

EEG recording at rest showed bilateral occipital polymorphic θ-α-wave mixed activity of medium amplitude. Right temporo-parietal spike-wave complexes indicated convulsive predisposition (Figure 1). With hyperventilation there was a decrease in frequency and discrete amplitude activation right parieto-occipitally. After completing the HV test a high-amplitude slowed frequency in the form of δ-wave mixed activity built up and reached an amplitude of 120 μV. It is more pronounced on the right than on the left. The phenomenon lasted as long as 22 minutes after completion of the hyperventilation test (Figure 2). No signs of motor or sensory disturbances were observed during the EEG recording.

3 Conclusion

The MRT showed multiple extinction of signals in the region of both basal ganglia – more pronounced on the right than on the left side – as

consequent reduction of cerebral perfusion. KONISHI [4] standardized these physiological changes in the EEG with respect to children. The build-up phenomenon disappeared with increasing age [4].

4 Discussion

The slowness of the appearance and disappearance of the re-build-up phenomenon, or a high amplitude frequency decrease directly after HV in Moyamoya syndrome has not yet been explained. The response of the collateral vessels to the pCO_2 reduction might possibly be slow but extended, and thus explains the slow normalization of the EEG. This typical EEG finding after HV is not seen in adults [1, 12], independent of the disease's stage. One possible explanation might be that a HV response is hardly detectable even in healthy adults.

In children, appearence of a re-build-up phenomenon in the EEG after hyperventilation always suggests Moyamoya syndrome. Such EEG findings justify cerebral angiography for a definite diagnosis.

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