The relationship between MRI findings and clinical features in patients with hypothalamic hamartoma

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Background and Objective: Hypothalamic hamartoma (HH) is a non-neoplastic lesion characterized by peculiar symptoms, including precocious puberty, mental retardation, behavioral problems, and epilepsy. However, little is known about the relationship between MRI findings and epilepsy. The objective of this study was to clarify the relationship between MRI findings and clinical manifestations in patients with HH.

Methods: We retrospectively reviewed MRI, EEG, and clinical data (mental retardation, precocious puberty, behavioral problems, and epilepsy) in 10 patients (male: 5, age:5 to 29 years) with HH. Based on the MRI classification by Arita *et al*[1], we categorized HH into two groups: a parahypothalamic (P) type, and intrahypothalamic (I) type. We also calculated the tumor volume of HH using DICOM images.

Results: Only one patient was classified as having P-type HH, with 9 as I-type HH. The HH volume ranged from 202 to 11,250 mm^3. One patient with P-type HH showed only behavioral problems. On the other hand, patients with I-type HH showed a variety of symptoms. All children developed epilepsy (gelastic seizure and complex partial seizure). Seven of 9 patients had behavioral problems. Eight of 9 patients developed precocious puberty. The volume of the tumor was inversely correlated with the age at epilepsy onset and the degree of mental retardation (DQ). Inter-ictal EEG also tended to change from focal spikes to diffuse spike and wave complex, depending on HH volume.

Discussion: There is limited information about the relationship between MRI findings and epilepsy in patients with HH. Arita *et al* proposed a new classification of HH into the P-type and I-type, based on MRI findings, and reviewed 61 reported cases of HH.[1] In their study, the P-type was generally associated with precocious puberty alone, whereas the I-type was generally accompanied by a variety of symptoms, including epilepsy, behavioral disorder, and developmental delay. They found no significant correlation between tumor size and seizures. On the contrary, Mullatti *et al*[2] studied 19 patients with epilepsy due to HH. They noted that a bigger size of HH was associated with an earlier onset of epilepsy and greater number of seizure types. In the present study, one patient was classified as P-type and nine as I-type. The P-type patient had solely behavioral problems. On the other hand, the I-type patients developed epilepsy (9/9), mental retardation (8/9), precocious puberty (8/9), and behavioral problems (7/9). In our I-type group, some clinical features of epilepsy depended on the tumor volume. The inter-ictal EEG tended to vary with tumor volume. We found an inverse relationship between the volume of HH and age at the onset of epilepsy. Our findings suggested that a difference in the extent of hypothalamic involvement by HH may be responsible for the clinical variability of epilepsy associated with HH. However, further studies with a large sample size are necessary to confirm our results.

Conclusion: Our data indicated that the MRI classification by Arita, if combined with tumor volume, might be more helpful in predicting the clinical manifestations in patients with HH.

References

1. Arita K, Ikawa F, Karisu K, *et al*. The relationship between magnetic resonance imaging findings and clinical manifestations of hypothalamic hamartoma. *J Neurosurg* 1999; 91(2): 212-20.
2. Mullatti N, Selway R, Nashef L, *et al* The clinical spectrum of epilepsy in children and adults with hypothalamic hamartoma. *Epilepsia* 2003; 44: 1310-9.