Moyamoya disease clinical features

Children and adults with moyamoya disease may have different clinical presentations. The symptoms and clinical course vary widely, with the disease ranging from being asymptomatic to manifesting as transient events to causing severe neurologic deficits. Adults experience hemorrhage more commonly; cerebral ischemic events are more common in children.

Children may have hemiparesis, monoparesis, sensory impairment, involuntary movements, headaches, dizziness, or seizures. Mental retardation or persistent neurologic deficits may be present.

Adults may have symptoms and signs similar to those in children, but intraventricular, subarachnoid, or intracerebral hemorrhage of sudden onset is more common in adults.

Dysexecutive syndrome

Dysexecutive syndrome is common in patients with moyamoya disease (MMD).

Fang et al., aimed to determine which aspects of executive function are impaired in patients with MMD, in addition to the specific dysexecutive functions present among its clinical subtypes and the mechanisms underlying dysexecutive function in these patients.

The authors administered 5 typical executive function tests (the Stroop test, the Hayling Sentence Completion Test [HSCT], the verbal fluency [VF] test, the N-back test, and the Sustained Attention to Response Task [SART]) to 49 patients with MMD and 47 IQ-, age-, education-, and social status-matched healthy controls. The dysexecutive questionnaire (DEX) was also used to assess participants' subjective feelings about their executive function.

A total of 39 of the patients were evaluated by CT perfusion (CTP) before the assessments were performed, and the correlations among the performances of the patients on the above tests with the parameters of cerebral blood volume, cerebral blood flow (CBF), mean transit time (MTT), and time-to-peak (TTP) in the frontal lobes of these patients were also analyzed.

Many aspects of executive function in the patients with MMD were significantly poorer than those in the healthy controls, and the patients performed particularly poorer on the VF test, HSCT, N-back test, and SART. The patients with hemorrhagic MMD exhibited worse executive inhibition, executive processing, and semantic inhibition compared with those with ischemic MMD, but the latter group presented a worse working memory and poorer sustained attention. There were no significant differences in the DEX scores between the patients with MMD and healthy controls. The other findings were as follows: CBF was significantly positively correlated with the number correct on part B of the HSCT ($r = 0.481$, $p = 0.01$) and accuracy on the 0-back task of the N-back ($r = 0.346$, $p = 0.031$); MTT was significantly positively correlated with accuracy on the 2-back task of the N-back ($r = 0.349$, $p = 0.034$) and factor 5 of the DEX ($r = 0.359$, $p = 0.032$); and TTP was significantly positively correlated with the number correct on part B of the HSCT ($r = 0.402$, $p = 0.034$) and the 1-back reaction time of the N-back ($r = 0.356$, $p = 0.026$).

The patients with MMD exhibited impairments in semantic inhibition, executive processing, working memory, and sustained attention, but they were not aware of these deficits. Moreover, differences in dysexecutive function existed between the different subtypes of MMD. Hypoperfusion of the frontal

lobe may be related to working memory and semantic inhibition impairments in patients with MMD [1].

Movement disorders

Movement disorders are a rare manifestation of Moyamoya angiopathy (MMA). Data on prevalence and clinical presentation are warranted. Possible involuntary movements include focal motor seizures, tremor, limb-shaking transient ischemic attacks, choreiform and spastic or dystonic movement disorders.

Kraemer et al developed a questionnaire to systematically assess movement disorders in MMA. Patients' history of involuntary movements and their clinical presentation were assessed systematically by interview. Additionally, demographic data were assessed as well as localization of movements, possible trigger factors and the presence of other symptoms.

The questionnaire was administered to 63 European patients with MMA. The response rate was high with 93.6% participating patients. Twenty-eight patients (47.4%) reported involuntary movement disorders including periodic tremor, irregular jerks, involuntary movements with loopy or pranced character, stiffness and muscle cramps. From those patients, 16 (57.1%) individuals had the symptoms prior to the diagnosis of MMA. The most common involuntary movements were irregular jerks witnessed by 17 (60.7%) patients, followed by stiffness and muscle cramps in 10 (35.7%). Eight (28.6%) Patients suffered from unintended loopy and pranced character, while 4 individuals (14.3%) remembered periodic tremor. Of the 28 patients who witnessed movement disorders, 23 had undergone revascularization surgery (82.1%). From the latter subgroup, movement disorders were reversed in 7 out of 12 patients (58.3%) with irregular jerks and 4 out of 7 patients (57.1%) with unintended loopy and pranced character.

The study elucidates the high incidence of movement disorders in an unselected consecutively recruited cohort of European MMA patients [2].

1)
Fang L, Huang J, Zhang Q, Chan RC, Wang R, Wan W. Different aspects of dysexecutive syndrome in patients with moyamoya disease and its clinical subtypes. J Neurosurg. 2016 Aug;125(2):299-307. doi: 10.3171/2015.7.JNS142666. Epub 2016 Jan 1. PubMed PMID: 26722860.
2)
Kraemer M, Trakolis L, Platzen J, Schwitalla JC, Bersano A, Albrecht P, Schlamann M, Berlit P. Movement symptoms in European Moyamoya angiopathy - First systematic questionnaire study. Clin Neurol Neurosurg. 2016 Nov 23;152:52-56. doi: 10.1016/j.clineuro.2016.11.017. [Epub ahead of print] PubMed PMID: 27898361.