Letters to the Editor

Progressive cerebroretinal microangiopathy with calcifications and cysts syndrome: An unusual cause of complex partial seizure

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PROGRESSIVE CEREBRORETINAL MICROANGIOPATHY syndrome is characterized by cerebral calcification and cyst formation (CRMCC), as defined in recent years,[1] and is described in the published reports as Coats plus syndrome, Labrune syndrome and leukoencephalopathy calcification and cysts (LCC) syndrome.[2] Cerebral, ophthalmic, skeletal, intestinal involvement can be seen. CRMCC determined in late adolescence is limited. Autosomal recessive inheritance is reported.[3] In the majority of patients, neurological findings showing progression have been reported, such as spasticity, dystonia, ataxia and loss of cognitive abilities. Imaging findings with laboratory data should support the diagnosis.[4]

The case presented here is of a 20-year-old male patient whose indications began in late adolescence and were defined as CRMCC characterized by central nervous system involvement. The patient had a history of febrile convulsions at the age of 2, and until 20 years old had suffered no seizures. He started to have complex partial seizure (CPS) with an aura of loss of speech and extremity numbness followed by extremity spasms accompanied by spasm of the head and neck area, versive rotation movements and manual automatisms after 20 years of age. Familial inheritance was not shown.

The neurological and ophthalmological examination was normal. There were no psychiatric, psychological or behavioral problems besides CPS. Memory disturbance due to left temporal lobe dysfunction was determined by the Wechsler Memory Scale (WMS) psychometric tests. Under observation in the video electroencephalography monitorization unit, CPS were seen three times. Seizures lasting up to 1 min were observed, before the seizure, showing a defining aura of loss of speech and numbness on the right side of the body, while during the seizure he had a behavioral arrest, his head was versive turned to the right and there were oral-alimentary and bilateral manual automatisms. Spike, sharp and slow wave activities were observed starting from the left frontal region and emitting to the temporal region during the seizure. For this reason, the seizure from the left frontal region and spread to the left temporal region was concluded.

Computed tomography showed multiple calcified foci in the cyst wall, basal ganglia, thalami and cerebral white matter. Magnetic resonance (MR) imaging revealed signal abnormalities in the cerebral white matter, and a right thalamo-caudate cyst and a smaller pontine cyst. There was no restriction of diffusion-weighted MR images. MR spectroscopy demonstrated minimal increase in the choline peak, mild decrease in the N-acetylaspartate peak, and a lactate peak. Serology positive data were not determined.

CRMCC cases generally show indications at an early age and the number of cases determined in late adolescence and older is relatively limited. Cases may present with different clinical tables related to the system involvement. To the best of our knowledge, cases of CRMCC-related complex partial seizures have not been reported.

In the radiological imaging data of CRMCC syndrome as leukoencephalopathy (demyelinization and white matter edema), calcification and cyst formation were found. It is a syndrome which may accompany retinal microangiopathy. A differential diagnosis of CRMCC should be considered for cases presenting with CPS and radiological imaging findings. The patient gave the authors written informed consent to publish this report.

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Preferences of help regarding behavioral health problems among the Japanese general population

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THE JAPANESE HEALTH-CARE system is a free-access system: patients are allowed to see any doctor in any subspecialty. Therefore, an understanding of why and where people seek care for behavioral disorders is important for planning for the behavioral health-services system.[1,2] This study was undertaken to investigate where people would seek help when they were presented with typical behavioral problem scenarios.

Of 192 000 individuals who registered as survey participants in a Japanese online research survey website, survey questionnaires were distributed to those who agreed to participate in this study.

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