To the Editor: Rasmussen’s encephalitis (RE) is a rare but devastating unihemispheric brain disorder, usually affecting children and characterized by intractable seizures and progressive neurological deficits. Characteristic magnetic resonance imaging (MRI) finding of RE is progressive unihemispheric focal cortical atrophy. Bilateral RE is very uncommon. Here, we report an RE patient with faciobrachial dystonic attacks and bilateral brain atrophy but without seizures.

A 15-year-old boy presented with progressive left hemiparesis at Department of Neurology, Peking Union Medical College Hospital in November 2013, which appeared at the age of 13. One and a half years ago, the patient suffered from left arm weakness. Half a year later, he had weakness of left leg and slurred speech. The symptoms progressed gradually. Cerebrospinal fluid (CSF) tests were normal. Fifteen months after onset, a 24-h electroencephalography (EEG) was unremarkable, and brain MRI showed atrophy in the right insular cortex, caudate, putamen, and cerebral crus with high signals in the corona radiata on T2-weighted and fluid-attenuated inversion recovery images. Physical examination found dysphasia. Muscle tone in the left limbs was high, and strength was 4/5. Ankle clonus and Babinski sign were positive in the left side. Blood tests showed normal leukocyte and platelet counts. Liver and renal function and erythrocyte sedimentation rate were normal. Anti-neuronal antibodies and antibodies for herpes simplex virus, rubella virus, Cytomegalovirus, toxoplasma, and Epstein–Barr virus were negative. Repeated CSF analysis was normal. Voltage-gated potassium channel complex (VGKC) testing with CD3-T cells showed a positivity of 12% in the serum and CSF. Anti-LGI1 antibodies were positive in the serum and CSF. MRI was performed in December 2013, which showed atrophy of the head of left caudate nucleus.

During the follow-up, the patient’s neurological deficits progressed, and he developed faciobrachial dystonic attacks 2 years later, which demonstrated paroxysmal unilateral involuntary movements of the left arm and face, lasting about 5 min and occurring several times a day. He did not lose consciousness or drop during the attacks. In December 2014, repeated MRI demonstrated progressive right hemispheric atrophy, enlargement of bilateral frontal horn, and atrophy in the left insular, perisylvian cortex, and caudate. His neurological function declined during the initial 34 months. He had aphasia and spastic quadriplegia. Frequency of faciobrachial dystonic attacks increased to dozens of time. After that period, the patient passed into a stage with a stable neurological deficit. Since both imaging features and focal deficits implicated bilateral hemispheric involvement, he was diagnosed bilateral RE.

There is debate about whether faciobrachial dystonic attacks were movement disorder or seizures. Faciobrachial dystonic attacks were often seen in limbic encephalitis associated with positive VGKC/LGI1 antibodies. In this case, the attacks were more likely movement disorder rather than seizures. No loss of consciousness or drop occurred during the attacks, and ictal EEG showed no epileptiform changes. The duration of the attacks lasted about 5 min, which was much more suggestive of hemidystonia, while faciobrachial dystonic seizures are very brief, usually lasting < 3 s.

Another feature of this case was that the neuroimaging showed predominant basal ganglia involvement with putaminal and caudate atrophy, which could explain the extrapyramidal manifestations such as dystonia and dysphasia. The faciobrachial dystonic attacks of the right arm and face might be associated with the prominent atrophy of the head of left caudate nucleus.

Address for correspondence: Dr. Jian-Hua Chen, Department of Neurology, Peking Union Medical College Hospital, Chinese Academy of Medical Sciences, Beijing 100730, China. E-Mail: deweiyy@163.com.

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This case demonstrated that the manifestation of epilepsy can be timely dissociated from the inflammatory and degenerative features of RE,[5] confirming that seizures are not an obligatory presenting symptom of RE. Epilepsy might be relatively rare in RE presented with dystonia.

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Conflicts of interest
There are no conflicts of interest.

References
1. Bien CG, Granata T, Antozzi C, Cross JH, Dulac O, Kurthen M, et al. Pathogenesis, diagnosis and treatment of Rasmussen encephalitis: A European consensus statement. Brain 2005;128(Pt 3):454‑71. doi: 10.1093/brain/awh415.
2. Striano P. Faciobrachial dystonic attacks: Seizures or movement disorder? Ann Neurol 2011;70:179‑80. doi: 10.1002/ana.22470.
3. Irani SR, Michell AW, Lang B, Pettingill P, Waters P, Johnson MR, et al. Faciobrachial dystonic seizures precede Lgi1 antibody limbic encephalitis. Ann Neurol 2011;69:892‑900. doi: 10.1002/ANA.22307.
4. Maramattom BV, Jeevanagi SR, George C. Facio‑brachio‑crural dystonic episodes and drop attacks due to leucine rich glioma inactivated 1 encephalitis in two elderly Indian women. Ann Indian Acad Neurol 2013;16:590‑2. doi: 10.4103/0972‑2327.120480.
5. Ferrari TP, Hamad AP, Caboclo LO, Centeno RS, Zaninotto AL, Scattolin M, et al. Atypical presentation in Rasmussen encephalitis: Delayed late‑onset periodic epileptic spasms. Epileptic Disord 2011;13:321‑5. doi: 10.1684/epd.2011.0455.