Dopa-responsive acute disseminated encephalomyelitis with marked atrophy of the striate body

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Accepted 17 May 2016

DESCRIPTION

A 16-year-old boy was admitted to our hospital, with a 4-day history of hyperthermia, vomiting, confusion, nuchal rigidity and dysbasia. Cerebrospinal fluid (CSF) examination showed: white cell count,

673/ μ L (69% polymorphonuclear cells); protein, 305 mg/dL; glucose ratio of CSF to serum, 0.35; and negative results for culture, antiviral antibodies, Epstein-Barr virus DNA and oligoclonal IgG bands (OB). Antibiotics and acyclovir were given

Figure 1 Axial T2-weighted imaging (T2WI) (left), axial T1WI (middle) and coronal fluid-attenuated inversion recovery (FLAIR) (right) imaging of the brain. (A) One week after admission, bilateral caudate nuclei and putamina show higher than normal signal intensity with swelling on T2WI. T1WI shows neither abnormalities nor gadolinium enhancement. Cerebral white matter shows multiple high-intensity lesions on FLAIR. (B) One month after admission, atrophy of bilateral striate bodies and cerebrum begins to appear on T2WI. Bilateral striate bodies have become hyperintense without gadolinium enhancement on T1WI, although the multiple cerebral white matter lesions appear diminished on FLAIR. (C) Three and a half months after admission, atrophy of the striate bodies and cerebrum appears markedly worsened on T2WI, although hyperintensity of the striate bodies on T1WI and cerebral white matter lesions on FLAIR have disappeared.

To cite: Goto Y, Warabi Y, Sunami Y, et al. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2016-215859

intravenously under a tentative diagnosis of infectious meningitis. However, unconsciousness was prolonged. CSF myelin basic protein (MBP) and IgG index were abnormally elevated to 257 pg/mL (normal, <102 pg/mL) and 1.52, respectively. Cerebral white matter showed multiple high-intensity lesions on fluid-attenuated inversion recovery (FLAIR) imaging, and bilateral striate bodies showed signal hyperintensity with swelling on T2-weighted imaging (figure 1A). We finally diagnosed acute disseminated encephalomyelitis (ADEM). Two courses of high-dose intravenous methylprednisolone, intravenous immunoglobulin and oral prednisolone therapy improved CSF findings within 2 weeks. However, extrapyramidal symptoms, including mask-like facies, rigidity of the left extremities, neck and trunk, and myoclonus, developed in addition to the pre-existing prolonged bedridden and mute state at 3 weeks after admission, and lasted for another 2 months. Atrophy of the striate body and cerebrum began to appear and bilateral striate bodies appeared hyperintense without gadolinium enhancement on T1-weighted imaging (T1WI), although multiple cerebral white matter lesions improved (figure 1B). After starting levodopa therapy and gradually increasing the dose to 600 mg/day by nasogastric tube, extrapyramidal symptoms rapidly improved over 3 days, and the patient regained the ability to eat, talk and walk independently over the subsequent month. At that time, atrophy of the striate body and cerebrum showed marked deterioration, although striate body hyperintensity on T1WI and cerebral white matter lesions on FLAIR had disappeared (figure 1C).

Our case was a dopa-responsive ADEM patient with marked atrophy of the striate body. In the present report, we described the value of levodopa trial for ADEM with basal ganglia lesions and extrapyramidal movement disorder. Few reports have examined the suitability of levodopa treatment for ADEM with basal ganglia lesions and extrapyramidal movement disorder.¹ ADEM is characterised by a severe inflammatory attack with a monophasic course and frequent grey matter involvement compared with multiple sclerosis.² Basal ganglia lesions were seen in 18% of patients with ADEM and in 80% of patients with poststreptococcal ADEM.³ Poststreptococcal ADEM has been distinguished from ADEM and termed paediatric autoimmune neuropsychiatric disorders associated with streptococci (PANDAS). High-dose intravenous methylprednisolone therapy is recommended for patients with ADEM without grey matter involvement, as well as for those with basal ganglia lesions and extrapyramidal symptoms.³ However, extrapyramidal movement disorder in our patient with ADEM did not respond to high-dose intravenous methylprednisolone, intravenous immunoglobulin and oral prednisolone therapy. As a reason for the rapid effectiveness of levodopa in our patient with ADEM with extrapyramidal symptoms due to striate body involvement, we speculated that the basal ganglia pathology might have involved demyelination with some preservation of levodopa-responsive neurons in the atrophic striate body.

Although our case did not have preceding infection and vaccination, diagnosis of ADEM was made according to the

diagnostic criteria for ADEM in children by the International Pediatric Multiple Sclerosis Study Group.² Brain MRI in ADEM typically shows the following three characteristics. Diffuse, poorly demarcated, large lesions involving predominantly the cerebral white matter. T1 hypointense lesions in the white matter are rare. Deep grey matter and cortical lesions can be present. However, there are wide variations in ADEM lesions and gadolinium enhancement occurs in <30% of cases.² Moreover, differential diagnosis is important for ADEM. Inflammatory and autoimmune diseases, CNS malignancy such as lymphoma and glioma, leukodystrophy, CNS infectious diseases, vitamin deficiency, granulomatous diseases and mitochondrial diseases are needed in the differential diagnosis. Our case showed negative serum and CSF laboratory results for these differentials, the clinical course denied incurable diseases and MRI showed the three characteristics of ADEM aforementioned.

Learning points

- ▶ If extrapyramidal symptoms in acute disseminated encephalomyelitis (ADEM) with basal ganglia lesions do not respond to immune therapies, a levodopa trial might prove valuable.
- ▶ Few reports have examined the suitability of levodopa treatment for ADEM with basal ganglia lesions and extrapyramidal movement disorder.
- ▶ Compared with multiple sclerosis, ADEM is characterised by more frequent grey matter involvement. Basal ganglia lesions were seen in 18% of patients with ADEM and in 80% of patients with poststreptococcal ADEM. Poststreptococcal ADEM has been distinguished from ADEM and termed paediatric autoimmune neuropsychiatric disorders associated with streptococci (PANDAS).

Contributors The patient was managed by YG and YS. The manuscript was written by YW. YG and EI contributed to critical revision of the manuscript.

Competing interests None declared.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

REFERENCES

- 1 Alves RS, Barbosa ER, Scaff M. Postvaccinal parkinsonism. *Mov Disord* 1992;7:178–80.
- 2 Krupp LB, Tardieu M, Amato MP, et al. International Pediatric Multiple Sclerosis Study Group criteria for pediatric multiple sclerosis and immune-mediated central nervous system demyelinating disorders: revisions to the 2007 definitions. *Mult Scler* 2013;19:1261–7.
- 3 Dale RC, Church AJ, Cardoso F, et al. Poststreptococcal acute disseminated encephalomyelitis with basal ganglia involvement and auto-reactive antibasal ganglia antibodies. *Ann Neurol* 2001;50:588–95.

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