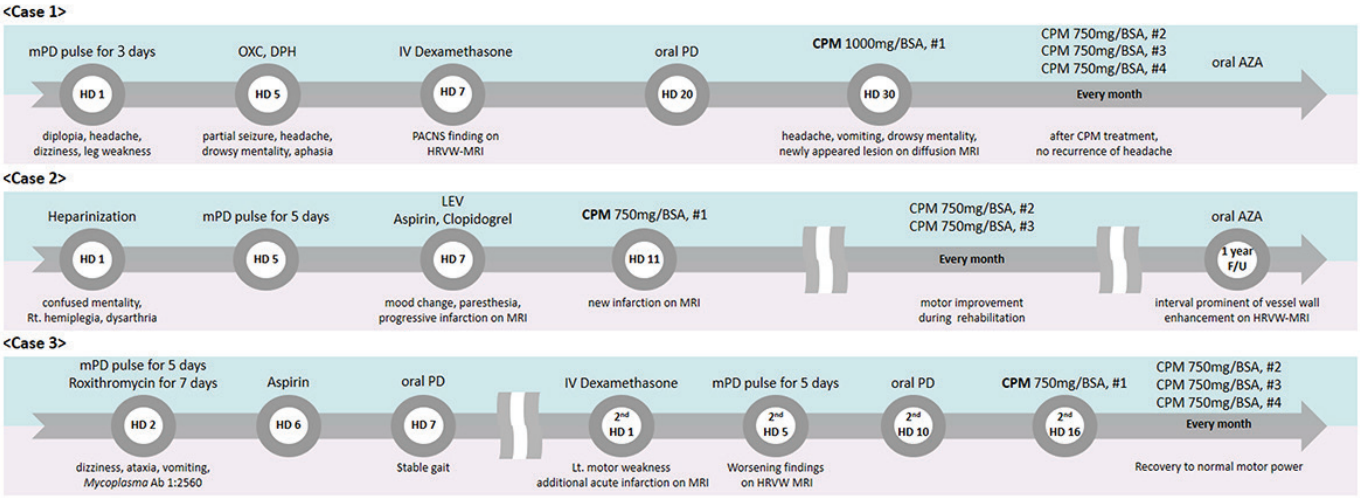

Supplementary Fig. 1. Clinical course of pediatric patients with primary angiitis of the central nervous system (PACNS). Case 1: An 8-year-old male patient experienced headache, dizziness, leg weakness, and diplopia at initial presentation. He experienced partial seizures and aphasia in 4 days. Brain magnetic resonance imaging (MRI) revealed multifocal acute infarction in the left occipital area. High-resolution intracranial vessel wall-magnetic resonance imaging (VW-MRI) revealed PACNS; therefore, he was treated with high-dose steroid therapy and cyclophosphamide monthly as induction therapy, followed by azathioprine as maintenance therapy. Case 2: An 11-year-old female patient presented with a sudden onset of confused mentality, right hemiplegia, and dysarthria. Brain MRI indicated an acute infarction in the left basal ganglia. VW-MRI revealed vessel wall enhancement in the left distal internal carotid artery and middle cerebral artery. Cyclophosphamide was administered due to progressive infarction even after high-dose steroid therapy. Her symptoms improved, but the last VW-MRI showed a relapse of vessel wall enhancement, and azathioprine was initiated. Case 3: A 6-year-old male patient presented with dizziness, vomiting, and ataxia. Brain MRI demonstrated multistage infarction, mainly involving the left thalamus and cerebellum. VW-MRI revealed vessel wall enhancement in the right anterior inferior cerebellar artery. Although the patient received high-dose steroid therapy, he experienced motor weakness with a newly developed infarction. In addition to the second high-dose steroid therapy, cyclophosphamide was initiated. mPD, methylprednisolone; OXC, oxcarbazepine; DPH, diphenylhydantoin; IV, intravenous; PD, prednisolone; CPM, cyclophosphamide; BSA, body surface area; AZA, azathioprine; LEV, levetiracetam.

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