Teaching NeuroImages: Rosai-Dorfman disease presenting with progressive early-onset cerebellar ataxia

A 13-year-old girl presented with a 3-year history of progressive gait abnormality. She recently had a self-limited cervical lymphadenopathy. Neurologic examination showed brisk tendon reflexes and moderate ataxia. Brain MRI disclosed hyperintense lesions in the cerebellum and pons (figure, A). Spine MRI showed a heterogeneous lesion in the T12 vertebra (figure, B). Histopathology of the cervical lymph node confirmed Rosai-Dorfman disease (RDD) by showing emperipolesis (figure, C and D). The patient will be followed up in order to determine disease progression and therapy.

RDD is a rare autoimmune histiocytic proliferative disorder first recognized in 1969. The CNS is involved in 5% of cases and generally mimics meningiomas. Bone erosion can be detected in the spine. Herein, we describe a rare CNS manifestation of RDD resembling a neurodegenerative ataxia.

AUTHOR CONTRIBUTIONS
Dr. da Silva: case description conception, neuroimaging conception, pathology conception, writing of the first draft (nothing to disclose). Dr. Pedroso: case description conception, case description organization, pathology organization, writing of the first draft, manuscript review and critique (nothing to disclose). Dr. Moraes: case description conception, neuroimaging conception, pathology conception, writing of the first draft. Dr. Rivero: neuroimaging conception, neuroimaging organization, manuscript review and critique. Dr. Callegari: figure imaging and histopathology.
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Carolina Candeias da Silva, José Luiz Pedroso, Fabiano Moulin de Moraes, et al.
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