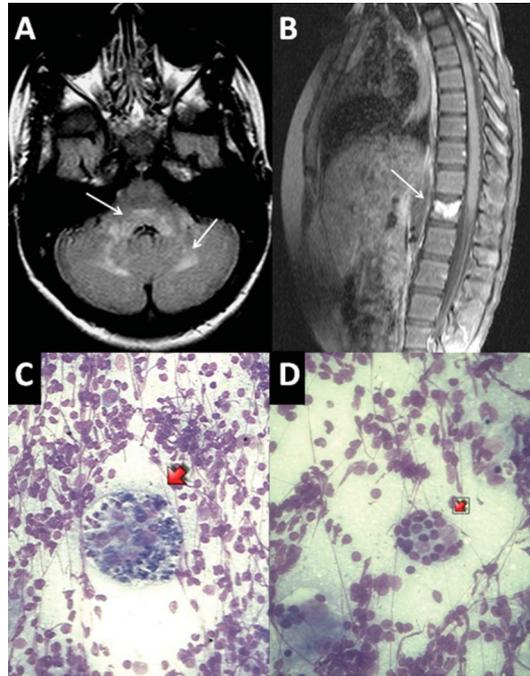


Teaching NeuroImages: Rosai-Dorfman disease presenting with progressive early-onset cerebellar ataxia

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Figure Imaging and histopathology



(A) Axial fluid-attenuated inversion recovery-weighted brain MRI shows cerebellar white matter (dentate nucleus), middle cerebellar peduncles, and dorsal pons involvement. (B) Spine MRI discloses a heterogeneous lesion with gadolinium enhancement on T12 vertebral body. (C, D) Cervical lymph node biopsy shows emperipolesis: histiocytes with abundant cytoplasm and engulfed viable lymphocytes, classically seen in Rosai-Dorfman disease.

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 1969.¹ The CNS

is involved in 5% of cases and generally mimics meningiomas. Bone erosion can be detected in the spine.² 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, case description execution, neuroimaging 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:

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