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Brain MRI features in Lhermitte-Duclos disease

Achados de RM cerebral na doença de Lhermitte-Duclos

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A 24-year-old woman presented with long-standing headache, blurred vision, and a 2-week-history of progressive ataxia and vomiting with papilledema and Parinaud syndrome, suggestive of raised intracranial pressure. Neuroimaging features were highly suggestive of dysplastic gangliocytoma of the cerebellum or Lhermitte-Duclos disease (LDD)[1,2] (Figure), which was confirmed in postsurgical histopathological evaluation. LDD represents a rare harmatomatous disorder linked to germline loss of one allele of the *PTEN* gene with subsequent loss of the remaining allele[3,4]. Cranial nerve palsies, gait ataxia and obstructive hydrocephalus secondary to a slowly progressive unilateral cortical cerebellar tumor represents the most common clinical findings[5].

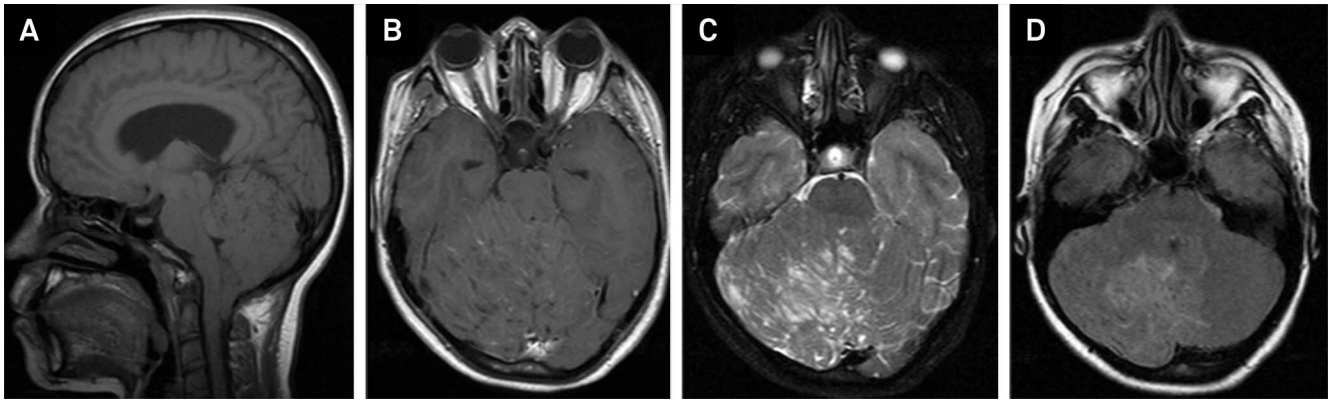

Figure. Sagittal T1-weighted MRI showing a superior vermian hypointense mass with brainstem compression and cerebellar tonsil herniation (A). Axial contrast-enhanced T1-weighted MRI unvealing non-enhancing hypointense mass in the right cerebellar hemisphere and vermis with leptomeningeal vessels enhancement in sulci between cerebellar folia (B). Axial T2-weighted (C) and FLAIR MRI sequences (D) disclosing hyperintense gyriform pattern with enlargement of cerebellar folia and alternate high- and normal-signal bands.

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