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 hamartomatous disorder linked to germline loss of one allele of the \textit{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\(^5\).

References