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Dysplastic cerebellar gangliocytoma or Lhermitte-Duclos disease: a pediatric case report and literature review

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A dysplastic gangliocytoma of the cerebellum (DGC) or Lhermitte-Duclos disease is a very rare lesion of unknowns' pathogenesis. There are a little over 200 cases described in the literature, most cases in adults between 30 and 40 years old. The disease manifests itself through a cerebellar syndrome or intracranial hypertension secondary hydrocephalus. The authors describe a case of a 12-year-old girl with a 2 months history of headache and vomiting. A CCT scan showed a supratentorial hydrocephalus and a large left cerebellar non-enhancing lesion. In urgency, a shunt was placed. A few days later, through a subocciptal craniotomy was held the partial withdrawal of the tumor. After the surgery, the patient has been showing only a mild ataxia. Histopathological and immunohistochemical analysis diagnosed a characteristic cerebellar ganglionic proliferative lesion, grade 1 OMS. The authors intend to discuss this rare disease, diagnosis, treatment and prognosis, through a review of the few published cases.