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C0476 - LHERMITTE-DUCLOS DISEASE OR CEREBELLAR DYSPLASTIC GANGLIOCYTOMA

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Resumen

Objectives: Lhermitte-Duclos disease is a rare autosomal recessive disease. We present a 6-year-old child patient who has been diagnosed with Lhermitte-Duclos in our clinic.

Methods: 6-year-old child with congenital Erb-Fallen paralysis, applied with headache to our clinic. Right cerebellar mass was found on MRI due to headache. MRI has been reported as a low-grade glial tumor. The lesion is microscopically gross total excised with right suboccipital craniotomy.

Results: Frozen section result was reported as normal cerebellar tissue during surgery. Histopathological result were reported microscopically and immunohistochemically as Lhermitte-Duclos. Focal hyperplasia of the cerebellar cortex is seen as pathological feature. Asymmetric growth of the cerebellum and also sharp thickening and condensation are observed in affected Gyrus macroscopically.on the other hand Condensation is observed in neurons similar to Purkinje cells with broad cytoplasm in the granular layer of the cerebellar cortex as microscopically.

Conclusions: Lhermitte-Duclos is a rare and slow progressive disease, generally seen in adolescence or in 3-4 decades. Early diagnosis and surgical treatment increase survival in these patients.