Lhermitte-Duclos disease

Doença de Lhermitte-Duclos

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Lhermitte-Duclos disease (dysplastic gangliocytoma of the cerebellum) is a rare benign cerebellar mass of unknown etiology which is characterized by enlargement of the cerebellar folia^{1,2}. Despite the controversy regarding its

A B

Figure 1. Typical MRI findings of Lhermitte-Duclos disease. T1-weighted axial imaging shows a non-enhancing hypointense cerebellar mass on the left hemisphere (A, B). T2-weighted axial and coronal imaging reveals a well circumscribed hyperintense mass with lamellar areas of isointensity which occurs as a result of enlargement of cerebellar folia and compressed sulci (C, D). This striated pattern is referred to as "tiger striping" sign.

pathogenesis, imaging and histopathological findings are rather typical 1,2,3,4 .

A 17-year-old female presented with a 2-year history of progressive headaches and gait imbalance. Cranial CT revealed a cerebellar expanding mass and hydrocephalus. The patient underwent shunt insertion and was referred to our department for treatment. MRI demonstrated a non-enhancing left hemisphere cerebellar tumor (Figure 1) which was subtotally resected during surgical procedure. Figure 2 shows the histological findings. Postoperative course was uneventful.

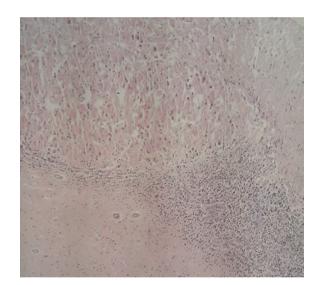


Figure 2. Typical histological features of Lhermitte-Duclos disease (H&E stain, original magnification x100). Photomicrograph reveals loss of normal cerebellar cortex architecture due to enlargement of the internal granular layer (lower right) and molecular layer (lower left), which are filled with dysplastic ganglion cells (upper left).

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Received 16 December 2013; Accepted 14 January 2014.

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