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Lhermitte-Duclos Disease: MRI Characteristics and Literature Review Based on a Clinical Case

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Abstract **Case Report**

Lhermitte-Duclos disease, also known as dysplastic cerebellar gangliocytoma, is a rare and benign tumor of the central nervous system. It is characterized by a hypertrophic ganglionic proliferation, which preferentially affects the cerebellar cortex, with an abnormal laminar arrangement [1]. Magnetic resonance imaging plays a major role in identifying this pathology. We report a case of a patient monitored for Cowden syndrome who presented with left hemiparesis and balance disorders. Brain MRI demonstrated a left hemispheric area, poorly defined, with isointense signal on T1 and hyperintense signal on T2 and FLAIR, and diffusion without ADC restriction, not enhanced after Gadolinium injection. This area is crossed by hypointense striations creating a striated and lamellar appearance. We shed light through this clinical case of a known Cowden syndrome carrier on the radiological aspects of this rare entity.

Keywords : Lhermitte-Duclos disease, Diagnosis, Radiological characteristic, Case repport.

INTRODUCTION

Lhermitte-Duclos disease is a very rare benign cerebellar tumor, called dysplastic cerebellar gangliocytoma, characterized by abnormal development and enlargement of the cerebellum [1]. This benign tumor is often observed in Cowden syndrome, which is characterized by a predisposition to the occurrence of multiple tumors [2]. Brain MRI plays a crucial role in the diagnosis of this tumor, thanks to its characteristic appearance "striated inverted cortex pattern" [3].

This work aims to show the contribution of brain MRI in the diagnosis and follow-up of Lhermitte-Duclos disease.

CASE REPORT

This is a 45-year-old patient being monitored for Cowden syndrome, diagnosed due to the combination of digestive polyposis, desmoid tumor of the mesentery, and multinodular goiter. He presented with left hemiparesis and balance disorders. Clinical examination revealed a static cerebellar syndrome. Brain magnetic resonance imaging showed a left hemispheric area, with poorly defined borders, iso-intense on T1 and hyperintense on T2 and FLAIR, and diffusion without ADC restriction, not enhanced after Gadolinium injection. This area is crossed by strands of hypo-intensity, creating a striated and lamellar appearance. The patient underwent partial resection. Early outcome was marked by transient persistence of the cerebellar syndrome.

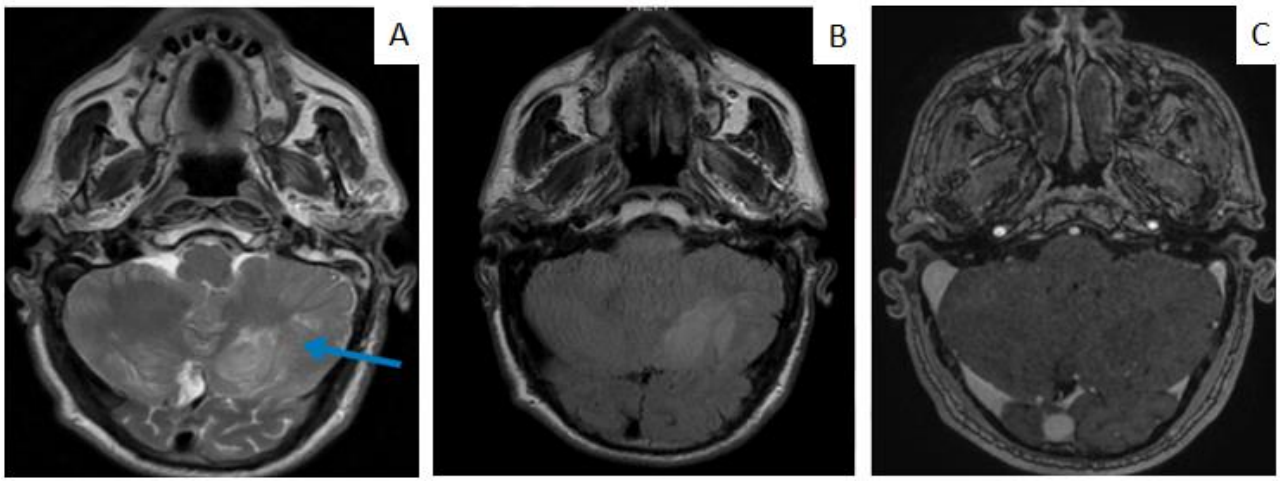

Figure 1: Axial MRI of the brain showing lamellar lesions (blue arrow), A: in T2 sequence, B: in FLAIR sequence, C: after contrast agent injection

DISCUSSION

Lhermitte-Duclos disease, also called dysplastic cerebellar gangliocytoma, is a rare tumor of the cerebellum characterized by hypertrophy of the cerebellar folia [1], frequently associated with Cowden syndrome, an autosomal dominant genomic anomaly caused by mutations in the tumor suppressor gene PTEN [2-4].

The diagnosis of Lhermitte-Duclos disease is clearly guided by the radiological appearance observed on brain MRI due to its pathognomonic "inverted cortex" aspect with an iso-hyperintense signal on T2-weighted sequence [2,3]. This radiological lamellar striation appearance is related to the replacement of Purkinje cells by dysplastic ganglion neurons [5].

In Lhermitte-Duclos disease, brain MRI shows a distortion of the laminar architecture of the cerebellum forming a mass with a gyriform appearance, iso/hypointense on T1 and hyperintense with iso/hypointense streaks on T2, hyperintense on diffusion without ADC restriction and without enhancement after Gadolinium injection [6.7]. The analysis of FLAIR sequences completes the morphological assessment to distinguish this entity from other posterior fossa tumors presenting as hyperintense [8].

Several retrospective series confirm that MRI could dispense with biopsy to confirm the diagnosis of this disease [9]. Although it is very sensitive, cerebral MRI has certain limitations, notably atypical forms such as pure hemispheric Lhermitte-Duclos disease without vermian involvement and pseudocystic or atrophic forms [10].

CONCLUSION

Lhermitte-Duclos is a very rare tumoral disease that mainly affects the cerebellum. Its radiological appearance on T2 hypersignal with a striped pattern, absence of contrast uptake, and at the level of the cerebellar cortex remains very suggestive, which allows for easy diagnostic guidance.

Conflicts of Interest: The authors declare no conflictsof interest.

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