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Case Report

Characteristic Cerebellar Striated Architecture on MRI Compatible with Lhermitte-Duclos Disease in an Adult Patient

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Received: March 31, 2026; **Published:** April 10, 2026

Abstract

Lhermitte-Duclos disease (LDD), or dysplastic gangliocytoma of the cerebellum, is a rare benign lesion characterized by thickened cerebellar folia and a laminated striated appearance on MRI. Although historically confirmed by histopathology, modern practice often relies on the typical imaging pattern when symptoms are mild and conservative management is chosen. We present the case of an adult patient with progressive headaches whose MRI revealed unilateral cerebellar hemispheric enlargement, thickened folia, and a T2-hyperintense laminated pattern without enhancement, findings highly compatible with LDD. No surgical intervention was indicated, and therefore no histopathologic confirmation was obtained.

This report reviews the radiologic features of LDD, highlights key differential diagnoses, and discusses its association with PTEN-related disorders such as Cowden syndrome. Recognizing the characteristic cerebellar striated architecture is essential, as MRI frequently provides sufficient diagnostic confidence even in the absence of CT or tissue sampling.

Keywords: *Lhermitte-Duclos Disease; MRI; Cerebellum; Dysplastic Gangliocytoma; Posterior Fossa*

Introduction

Lhermitte-Duclos disease (LDD) is an uncommon cerebellar hamartomatous lesion first described in 1920 and characterized by thickened cerebellar folia and abnormal cortical lamination. It most often affects young to middle-aged adults and is strongly associated with PTEN mutations, particularly in patients with Cowden syndrome [1]. MRI is the imaging modality of choice because it reliably demonstrates the laminated "tigroid" pattern corresponding to disorganized cerebellar architecture [2]. Although histopathology historically served as the diagnostic gold standard, MRI features are sufficiently characteristic to establish a compatible diagnosis in many cases, especially when patients are managed conservatively. This report presents an adult patient with classic MRI findings of LDD and emphasizes the value of imaging-based diagnosis in the absence of CT or histologic confirmation.

Case Report

A female adult presented with a several-month history of progressive occipital headaches without nausea, vomiting, or gait disturbance. Neurologic examination and laboratory findings were unremarkable. MRI was performed to evaluate a structural cause.

Imaging demonstrated enlargement of the left cerebellar hemisphere with markedly thickened folia showing a laminated, parallel striated ("tigroid") pattern. The lesion was hyperintense on T2-weighted and FLAIR sequences (Figure 1) and hypointense on T1-weighted sequences. On diffusion-weighted imaging (DWI, Figure 2), the lesion exhibited a striped pattern, with hyperintense interfolial parenchyma alternating with relatively hypointense laminated folia, without diffusion restriction. Following gadolinium administration, there was no enhancement (Figure 3). No hydrocephalus was observed.

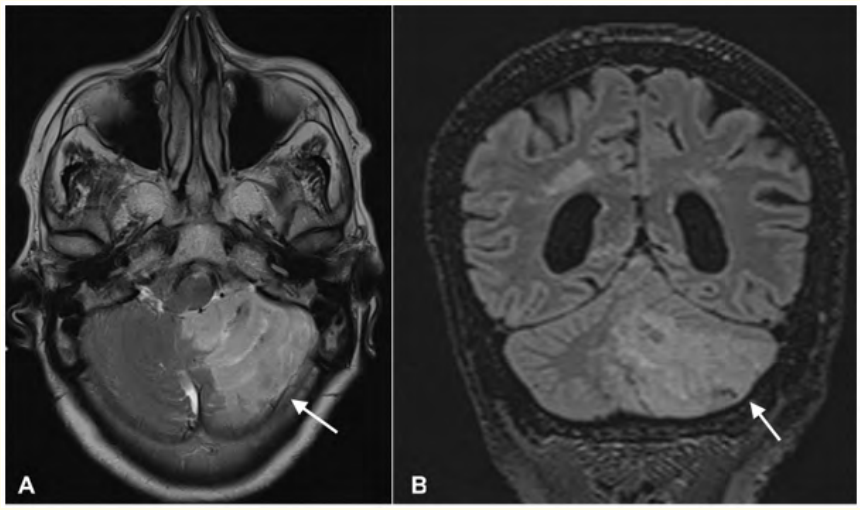

Figure 1: *(A) Axial T2-weighted image of the left cerebellar hemisphere showing markedly thickened folia with a laminated, parallel striated ("Tigroid") pattern, appearing hyperintense (Arrow). (B) Coronal FLAIR image demonstrating the lesion as hyperintense (Arrow).*

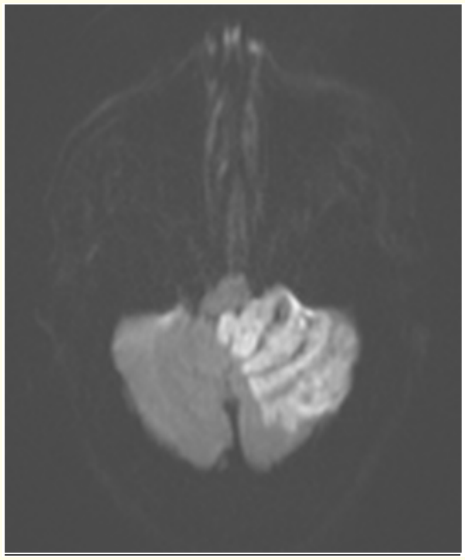

Figure 2: *Axial diffusion-weighted image of the left cerebellar hemisphere showing a striped pattern, with hyperintense interfolial parenchyma alternating with relatively hypointense laminated folia, reflecting the typical tigroid architecture.*

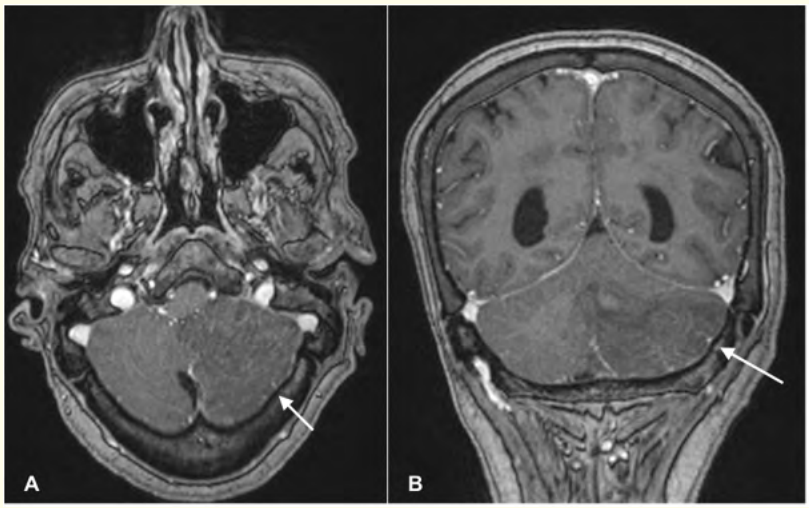

Figure 3: *Axial (A) and coronal (B) post-contrast T1-weighted images showing the lesion as hypointense with no enhancement (Arrow).*

Given the typical MRI features and the absence of severe symptoms or significant mass effect, conservative management was recommended. Surgical biopsy was not performed due to the classic imaging appearance. The patient remained clinically stable and was referred for genetic counseling to evaluate for potential PTEN-related disorders.

Discussion

Lhermitte-Duclos disease (LDD) is a rare, benign cerebellar disorder, with an estimated incidence of less than 1 per million individuals [3]. It is characterized by dysplastic overgrowth of cerebellar granule and ganglion cells, leading to thickened folia and unilateral or sometimes bilateral cerebellar enlargement. Histopathologically, LDD shows hypertrophic folia with replacement of the normal granule cell layer by dysplastic ganglion cells, preservation of cortical architecture, and absence of significant mitotic activity, reflecting its benign behavior [4]. Molecular studies have frequently identified germline mutations in the PTEN tumor suppressor gene, particularly in patients with Cowden syndrome, although sporadic cases without systemic involvement also occur [5,6].

Clinically, LDD presents with symptoms related to posterior fossa mass effect, including headache, cerebellar dysfunction, or signs of increased intracranial pressure. Many patients, however, present with mild or nonspecific symptoms, prolonging the time to diagnosis [1]. The slow, indolent growth pattern of the lesion contributes to the absence of acute neurological deterioration in many adult presentations, similar to the case described here.

MRI is the modality of choice for diagnosing LDD. On T2-weighted (and often FLAIR) sequences, the lesion typically shows a striated or “tiger striped” appearance [7,8], due to alternating bands of high and low signal intensity. The hyperintense bands correspond to interfolial white matter, while the hypointense bands represent thickened, dysplastic folia composed of abnormal ganglion and granular cells. Diffusion-weighted imaging (DWI) usually shows no restricted diffusion, consistent with the benign, dysplastic nature of the lesion. On T1-weighted images, the mass is hypointense or isointense relative to normal cerebellar tissue, and contrast enhancement is minimal or absent, which helps distinguish LDD from other cerebellar neoplasms [9].

Additional MRI techniques, when performed, such as spectroscopy may reveal decreased neuronal markers (N-acetylaspartate, NAA), low choline, and occasionally a lactate peak, reflecting metabolic alterations rather than aggressive tumor behavior. Perfusion studies may show mild vascular prominence but typically lack the high blood volume or neoangiogenesis seen in high-grade tumors [10,11].

Given these distinctive features, many authors consider the striated, nonenhancing MRI pattern sufficiently characteristic to allow a preoperative diagnosis of LDD without biopsy in asymptomatic or minimally symptomatic patients, though rare mimics on imaging must be considered.

The differential diagnosis of LDD includes disorders that may mimic cerebellar folial thickening or cause unilateral cerebellar expansion. Low-grade astrocytomas may produce cerebellar enlargement but usually exhibit some degree of enhancement, cystic components, or surrounding edema [12]. Medulloblastoma, though occurring in the posterior fossa, typically shows contrast enhancement and restricted diffusion, features absent in LDD [13]. Hypertrophic cerebellitis may lead to folial thickening, typically in a bilateral and diffuse pattern, but it lacks the systematic laminated appearance and architectural preservation characteristic of LDD [14]. Other malformative or hamartomatous cerebellar lesions may also resemble LDD but generally fail to reproduce its highly organized striated morphology.

Given these distinctions, the imaging features in this case strongly favored LDD over neoplastic or inflammatory processes.

Management of Lhermitte-Duclos disease (LDD) depends on symptom severity and the degree of mass effect. Asymptomatic or mildly symptomatic patients are generally managed conservatively, with surgery reserved for those experiencing progressive neurological decline or obstructive hydrocephalus. In our patient, a biopsy was not pursued, as MRI findings were compatible with LDD. Several modern case series and reviews report that, when the radiologic pattern is characteristic and intervention is not indicated, imaging-based diagnosis alone is often sufficient. This approach minimizes exposure to the risks of posterior fossa surgery while providing a reasonable level of diagnostic confidence [15,16].

Conclusion

Lhermitte-Duclos disease is a rare cerebellar lesion whose MRI features - including hemispheric enlargement, thickened folia, and a laminated striated pattern - are highly suggestive of the diagnosis. In cases where histopathology is unavailable, MRI can provide sufficient evidence for a diagnosis compatible with LDD, though it is not definitive. Conservative management with regular clinical and imaging follow-up is appropriate for patients with mild or absent symptoms. Recognition of this characteristic cerebellar architecture is essential for guiding management and ensuring appropriate surveillance.

Conflicts of Interest

The authors declare that they have no conflicts of interest.

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Volume 18 Issue 4 April 2026