

A case of Lhermitte-Duclos disease: the classical “tiger stripes” in magnetic resonance imaging

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ABSTRACT

Dysplastic gangliocytoma or Lhermitte-Duclos disease (LDD), is a rare benign cerebellar tumor often presenting with nonspecific symptoms such as intracranial hypertension, cerebellar dysfunction, or cranial nerve deficits, and associated with phacomatosis. The characteristic “tiger stripe” sign on magnetic resonance imaging (MRI) is highly suggestive of LDD, though nodular medulloblastoma remains an important differential diagnosis. We report a 51-year-old woman with arm and leg phacomatosis who presented with a right cerebellar hypodense lesion on computed tomography extending frontally. MRI demonstrated the hallmark “tiger stripe” appearance, and histopathology following surgery confirmed LDD. On follow-up, despite evidence of a recurrent mass on MRI, her cranial nerve paresis improved postoperatively. Imaging revealed hypointense striations on T1 and alternating linear hyperintensities on T2 sequences, obscuring the cerebellar folia, features that distinguish LDD from other cerebellar tumors. This case underscores the diagnostic value of MRI, emphasizing early recognition to prevent misdiagnosis and guide surgical planning.

KEYWORDS cerebellar neoplasms, Lhermitte-Duclos disease, magnetic resonance imaging

Lhermitte-Duclos disease (LDD) is a rare benign tumor characterized by aberrant cerebellar development, which can lead to increased intracranial pressure.¹ It typically affects middle-aged adults (30–50 years), although cases have been reported across all age groups, with no significant differences related to sex or race.^{2,3} Histologically, LDD is marked by overgrowth of dysplastic cerebellar gangliocytes and Purkinje cells.⁴ The primary treatment is surgical resection, often involving posterior fossa decompression and either

complete or subtotal tumor excision.^{5,6} Because of its rarity, unusual appearance, and cerebellar location, LDD is primarily diagnosed through radiological imaging. On magnetic resonance imaging (MRI), it typically appears hypointense on T1-weighted imaging (T1WI) and hyperintense on T2-weighted imaging (T2WI).⁷ A distinguishing feature is the “tiger stripe” sign on T2WI, which aids in preoperative diagnosis.⁸ This pattern is more consistently observed in LDD associated with tumors, whereas non-tumorous LDD may exhibit

atypical cerebellar imaging findings, including vascular abnormalities, intratumoral calcifications, hemorrhage, heterogeneous enhancement, and peritumoral edema.^{7,9}

LDD treatment aims to relieve the symptoms of mass effects via shunt placement or surgical resection. Surgical decisions are guided by the patient's symptoms, clarity of imaging findings, and presence of comorbidities.¹⁰⁻¹² LDD is classified as a World Health Organization class I mixed neuronal-glioma according to the 2016 version of the central nervous system classification owing to its slow progression and fair prognosis.^{13,14} The clinical presentation is frequently associated with increased intracranial pressure, cerebellar dysfunction, and cranial nerve impairments resulting from deformation of the cerebellar laminar architecture, which resembles tiger stripes and can be identified on MRI.^{15,16}

Although LDD is rare, reporting individual cases remains important because its clinical and radiological features can closely resemble other posterior fossa lesions, particularly malignant tumors

such as medulloblastoma or low-grade gliomas. Misclassification can lead to unnecessary aggressive treatment or delayed surgical planning. Given the variability of its presentation and the occasional absence of classic imaging features, each additional case contributes to a better understanding of its diagnostic spectrum and helps refine criteria for differentiating LDD from more common cerebellar pathologies. However, reports describing LDD accompanied by additional neurocutaneous findings or complex postoperative courses remain limited in the literature. To address this gap, this case highlights an uncommon presentation of LDD characterized by concurrent limb phacomatosis, recurrence of a cerebellar mass after initial surgery, and simultaneous involvement of cranial nerves VII, IX, and X. This study aimed to emphasize the diagnostic value of the characteristic "tiger stripe" MRI pattern and demonstrate how these unusual accompanying features expand current knowledge on the clinical variability of LDD.

CASE REPORT

A 51-year-old woman with decreased consciousness 3 days prior to hospitalization was referred from another hospital. She had a history of uncontrolled hypertension and reported transient dizziness over the past month, accompanied by frequent balance disturbances. She also complained of bilateral tinnitus and mild hearing impairment accompanied by repeated episodes of vomiting (approximately 10 times). In addition, she had a history of type 2 diabetes mellitus, although details regarding her medication regimen were unavailable. Meniere's disease was suspected in the previous



Figure 1. Phacomatosis or hyperpigmented skin in the patient's legs (a) and arms (b)

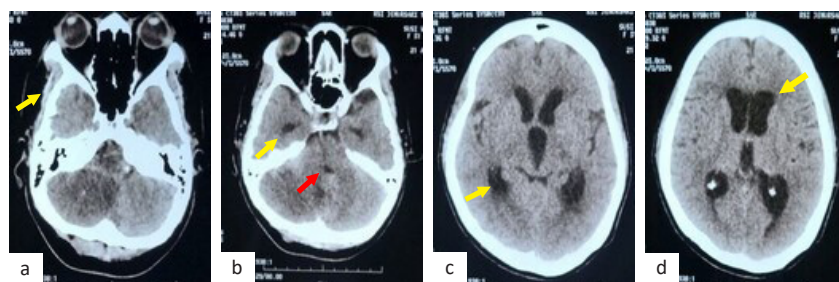


Figure 2. The unenhanced computed tomography (CT) scan. (a) Nonspecific hypoattenuating mass of the right cerebellar hemisphere extending to the vermis (yellow arrow); (b) mass pushed fourth ventricle to the left side (red arrow), temporal horns are dilated (yellow arrow); (c) dilatation of posterior horn of lateral ventricle (yellow arrow) confirming obstructive hydrocephalus; (d) periventricular edema also present (yellow arrow)

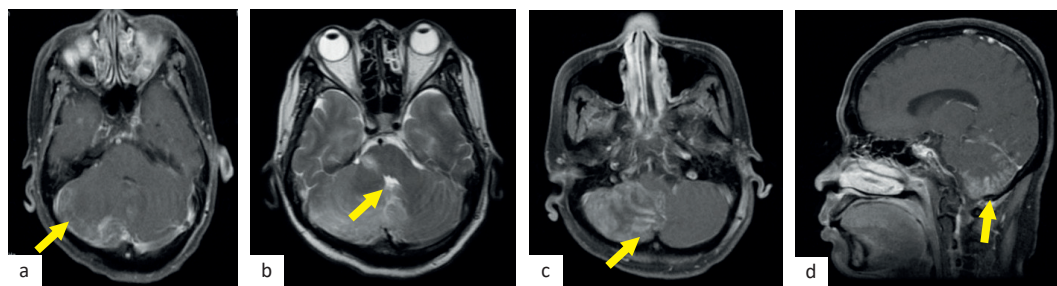


Figure 3. The MRI of the cerebellum showed a space-occupying lesion in the patient’s right cerebellar hemisphere. Overall, the lesion demonstrated a hypointense signal on T1WI with alternating slightly hyperintense linear areas (yellow arrow) (a). On T2WI, the lesion appeared predominantly hyperintense with interspersed hypointense stripes, producing the typical “tiger stripe” appearance. The lesion also caused compression and displacement of the fourth ventricle (yellow arrow) (b). Contrast-enhanced axial (c) and sagittal T1WI (d) showed non-homogeneous enhancement throughout the lesion, with scattered vascular-like areas of enhancement (yellow arrows). The imaging findings were suggestive of a typical diagnosis of LDD. LDD=Lhermitte-Duclos disease; MRI=magnetic resonance imaging; T1WI=T1-weighted imaging; T2WI=T2-weighted imaging

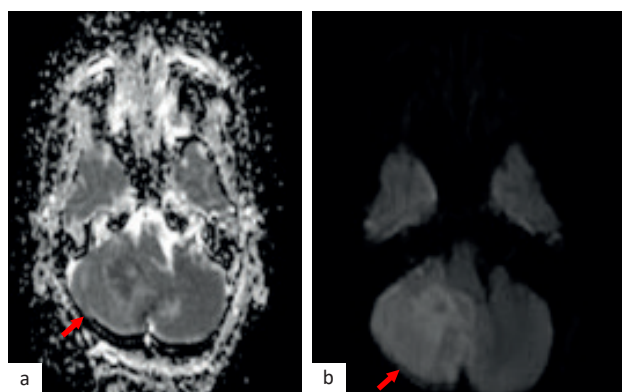


Figure 4. ADC and DWI of patients. (a) A signal on ADC (red arrow); (b) DWI showed slightly restricted diffusion (red arrow). ADC=apparent diffusion coefficient; DWI=diffusion-weighted imaging

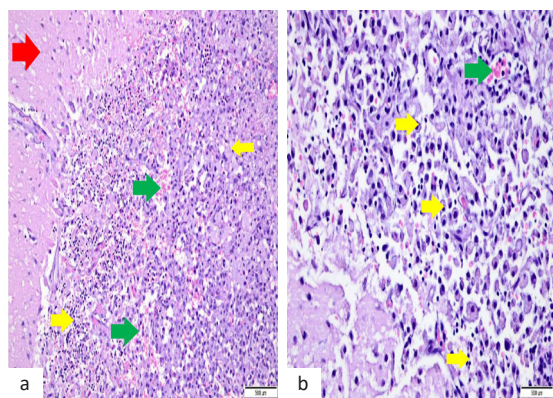


Figure 5. Internal granular layers in ganglion cells. (a) Photomicrograph of the surgical specimen revealed numerous hypertrophic ganglion cells in the internal granular layers (H&E, 200× magnification); (b) the hypertrophic ganglion cells blended with scant residual granular cells (H&E, 400× magnification). Brain parenchyma tissue consists of glial cells (red arrow) and a dense pile of inflammatory cells, lymphocytes, histiocytes (yellow arrows), and plasma cells, as well as proliferating blood vessels (green arrows) with lumens filled with RBC. H&E=hematoxylin and eosin; RBC=red blood cell

visit. No limb weakness, gait disturbances, visual impairment, or hearing loss was reported. Her family history was unremarkable, and no similar clinical signs were observed among her relatives. On examination, her blood pressure was 155/96 mmHg. Neurological examination revealed right cranial nerve VII palsy, bilateral cranial nerve IX–X palsy, and phacomatosis of the limbs and legs, accompanied by pathological Babinski reflexes and a positive Chaddock sign. She also exhibited areas of hyperpigmentation on both arms and legs, as illustrated in Figure 1.

A non-contrast head computed tomography (CT) scan conducted in the previous hospital 3 days before admission, revealed a hypodense lesion in the right cerebellar hemisphere; the lesion was anteriorly located with indistinct margins. Based on these findings, an intracranial tumor, most likely a glioma, was suspected. In addition to the acute onset of neurological symptoms, imaging findings suggested a mass effect compressing the fourth ventricle, resulting in noncommunicating hydrocephalus (Figure 2a–d). Consequently, a high-pressure ventriculoperitoneal shunt was urgently inserted to relieve intracranial pressure.

Brain MRI was performed for further evaluation, revealing a right cerebellar lesion with an ill-defined margin, measuring approximately 4.0 × 6.0 × 4.0 cm. The lesion appeared hypointense on T1WI (Figure 3a) and predominantly hyperintense on T2WI, with alternating high and normal signal intensity, giving it a tigroid appearance (Figure 3b). After contrast administration, axial and sagittal (Figure 3, c and d) showed heterogeneous enhancement, further supporting the tigroid pattern. On diffusion-weighted

imaging, the lesion demonstrated slight hyperintensity without restricted diffusion on the apparent diffusion coefficient (ADC) map (Figure 4).

Clinical pathway for LDD diagnosis: patients typically present with nonspecific symptoms such as headache, nausea, vomiting, ataxic gait, or other signs of increased intracranial pressure. Because these symptoms overlap with those of many posterior fossa conditions, LDD is rarely suspected at this stage. Initial CT imaging: non-contrast CT may revealed a posterior fossa mass or hemispheric enlargement with variable hypo- or isodensity. Findings are nonspecific and can mimic neoplastic or vascular lesions, prompting further evaluation.

In this case, the diagnosis of LDD was established based on the characteristic radiological features observed on MRI. The lesion demonstrated the classic T2-weighted “tiger stripe” pattern, a hallmark appearance that strongly suggests LDD. Additionally, the absence of contrast enhancement and lack of diffusion restriction helped exclude other differential diagnoses such as nodular medulloblastoma, low-grade glioma, or hemangioblastoma, which typically show either heterogeneous enhancement, restricted diffusion, or associated edema. Taken together, these imaging characteristics allowed a confident preoperative diagnosis of LDD in this patient.

She underwent surgical resection. Intraoperative findings included swelling of the left cerebellum and compression of the contralateral side. Histopathological examination revealed cerebellar tissue with an expansion of the cerebellar cortex. The internal granular layers were loaded with hypertrophic ganglion cells with rounded and pleomorphic nuclei and copious cytoplasm (Figure 5), confirming the pathological diagnosis of dysplastic cerebellar gangliocytoma (LDD).

Postoperative MRI showed a recurrent mass lesion in the right cerebellum measuring 2.5 × 1.7 × 2.5 cm. However, symptoms related to cranial nerve IX and X paresis resolved, whereas the peripheral paresis of cranial nerve VII improved from House-Brackmann grade III to II.

Two weeks postoperatively, she attended a follow-up visit (discharged at September 16, 2022) with an enlarged lump at the back of the head, which was identified as cerebrospinal fluid leakage from a postoperative defect. She underwent debridement with defect closure and shunt replacement. Thereafter,

her condition improved with no current postoperative neurological deficits (6 months).

Informed consent was obtained from the patient for publication of this case report and accompanying images. This study was also approved by the Research Ethics Committee of Universitas Airlangga Hospital (No: 055/KEP/2023), with protocol number UA-01-23084.

DISCUSSION

In this patient, the initial CT findings were nonspecific, revealing a hypodense posterior fossa lesion with an associated mass effect that could mimic various cerebellar pathologies, including glioma or medulloblastoma. Because CT alone could not distinguish between these possibilities, further imaging was essential. MRI provided the defining diagnostic clues: the characteristic T2-weighted “tiger stripe” pattern, together with the absence of contrast enhancement and lack of diffusion restriction, strongly supported a diagnosis of LDD. These combined features indicated a benign dysplastic process rather than a malignant neoplasm, allowing clinicians to modify the management plan accordingly and avoid overtreatment. This highlights the critical role of MRI in differentiating LDD from other posterior fossa lesions with similar initial presentations.

Our case, when compared with the study reported by Zhang et al,¹⁰ a notable difference is observed. Our case demonstrated a relatively strong enhancement of the right cerebellar lesion, whereas the cases presented in that study typically showed no enhancement or only minimal enhancement following contrast administration. Compared with Alanazi et al,¹⁷ our case shows typical LDD features such as the T2 “tiger stripe” pattern and posterior fossa mass effect, but with more prominent enhancement than the mild enhancement reported in only 14.9% of cases. Recurrence in our patient also aligns with their 8.6% rate, while the absence of genetic testing is a limitation given their one-third association with Cowden syndrome. Similarly, in comparison with Han et al,⁹ our case matches the expected “tiger stripe” sign and lack of diffusion restriction but differs in showing more marked heterogeneous enhancement and an early postoperative recurrence. Overall, these comparisons highlight that LDD can present with both classic and atypical imaging characteristics.

When MRI demonstrates the classical “tiger stripe” pattern, differential diagnoses such as nodular medulloblastoma become considerably less likely, even though these entities may share overlapping T1- and T2-weighted characteristics. In contrast to medulloblastoma—which is a malignant neuroectodermal tumor typically presenting with heterogeneous enhancement, restricted diffusion, and surrounding edema—LDD usually shows non-enhancing, non-restrictive, and more organized cerebellar folial striations. These radiological differences are clinically important because medulloblastoma requires aggressive management with surgical excision followed by adjuvant chemoradiation, whereas LDD is a benign dysplastic lesion often managed conservatively or with limited decompression when symptomatic. Recognizing these distinctions prevents unnecessary radical intervention and guides appropriate individualized treatment.

Distinguishing LDD from other posterior fossa tumors can be challenging, particularly when contrast enhancement or diffusion characteristics overlap. In this case, the lack of significant contrast enhancement and preservation of diffusion on ADC mapping were consistent with LDD, correlating with the dysplastic folial architecture and intact blood-brain barrier. The MRI “tiger stripe” appearance was the decisive feature guiding diagnosis and surgical planning. Recognizing this sign is critical for avoiding misclassification and unnecessary aggressive treatment.

The clinical importance of this case lies in the role of MRI in preoperative diagnosis, especially the “tiger stripe” sign, a hallmark for LDD. Early recognition of this imaging feature can support timely surgical decision-making and help avoid unnecessary aggressive interventions or misdiagnoses, particularly in cases with overlapping presentations such as medulloblastoma. This finding supports a more individualized and safer therapeutic strategy.

Histological studies have revealed the disruption of normal cerebellar cortex cell architecture, with dysplastic-hypertrophied ganglion cells enlarging the granule layer and broadening the molecular layer. Reduced white matter and loss of Purkinje cells were also observed. Typically, features characteristic of neoplastic processes, such as mitotic activity, necrosis, or endothelial development, were absent, and malignant transformation has not been reported to date. Observation and symptomatic therapy are

recommended unless the mass impact causes severe symptoms, in which case, surgical excision may be warranted. Complete resection is associated with low recurrence rates.

Resection was chosen in this patient because the lesion produced significant mass effect, obstructed the fourth ventricle, and resulted in noncommunicating hydrocephalus—conditions that required urgent decompression. She also presented with progressive neurological deficits, including cranial nerve VII, IX, and X paresis, indicating brainstem compression. Although LDD is a benign lesion, surgery is recommended when symptoms arise from mass effect or when hydrocephalus is present, as conservative management would not alleviate structural compression. In this case, resection provided both therapeutic decompression and definitive histopathological confirmation, guiding further management and preventing worsening neurological compromise.

This case highlights the importance of early recognition and appropriate management of LDD, which can lead to favorable short-term outcomes. Although the patient showed improvement during the initial postoperative period, the relatively short follow-up duration of 6 months remains a key limitation, as LDD is known for its slow progression and potential for delayed recurrence. Therefore, long-term monitoring is essential to fully assess treatment success and identify any late complications. The take-home message from this case is that clinicians should ensure extended surveillance in LDD patients to optimize long-term outcome.

Conflict of Interest

The authors affirm no conflict of interest in this study.

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