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Atypical MR Appearance of Lhermitte-Duclos Disease with Contrast Enhancement

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Summary: A case of surgically confirmed Lhermitte-Duclos disease demonstrated contrast enhancement on MR. Histologic examination verified corresponding increased vascularity in the molecular layer and adjacent leptomeninges.

Index terms: Cerebellum, magnetic resonance; Phakomatoses

Previous reports have described the characteristic appearance of Lhermitte-Duclos disease as that of a slowly growing, nonenhancing cerebellar mass (1–6). We report the findings in a patient with surgically confirmed Lhermitte-Duclos disease, which demonstrated contrast enhancement.

Case Report

A 27-year-old woman presented with a 1-week history of black spots in her visual fields, balance and coordination difficulties, and occasional headaches. Her medical history was otherwise unremarkable. Physical examination demonstrated papilledema.

Magnetic resonance (MR) examination (Fig 1A–C) demonstrated obstructive hydrocephalus secondary to a left cerebellar mass. The alternating bands typical of Lhermitte-Duclos disease were noted. After administration of gadopentetate dimeglumine there was marked enhancement of portions of the isointense bands on the T1-weighted sequences.

Histologic examination after surgical removal (Fig 1D) demonstrated the classic features of Lhermitte-Duclos disease. The cortex consisted of an expanded hypermyelinated molecular layer overlying a cellular layer composed of large neurons. Increased numbers of small blood vessels were identified in the molecular layer of the cerebellum and adjacent meninges. Histologic evidence of dystrophic calcification was associated with many of the vessels.

Discussion

Lhermitte-Duclos disease is a rare disorder characterized by a slowly enlarging mass in the

cerebellum. The normal cerebellar cortex consists of an inner granular layer, an outer molecular layer, and an intervening Purkinje layer. In Lhermitte-Duclos disease the folia become thickened and enlarged, with loss of secondary arborization, resulting in a layered pattern that can frequently be recognized on MR studies. There is marked reduction in the central white matter, which is often replaced by a slitlike cavity, whereas there is hypertrophy of the granular cells with an expansion of the granular layer and myelination of the molecular layer. The histologic appearance seems to combine the features of congenital malformation and neoplasia; hence, the term “dysplastic gangliocytoma of the cerebellum” has been applied to Lhermitte-Duclos disease (7–9).

The reported MR appearance is that of a nonenhancing mass with a layered appearance consisting of alternating bands of relative isointensity and hypointensity to the normal brain on T1-weighted sequences and relative isointensity and hyperintensity on T2-weighted sequences. The bands of relative isointensity on T1- and T2-weighted sequences seem to correspond to the outer portion of the enlarged folia, whereas the regions of T1 and T2 prolongation represent the central core.

We have noted an instance in which portions of the isointense bands that represent the outer portion of the enlarged folia (molecular layer) markedly enhance with contrast material. We think this is most likely secondary to the vascular proliferation noted microscopically. Such vascular proliferation has been noted involving the molecular layer and the adjacent pia (10, 11). The presence of contrast enhancement

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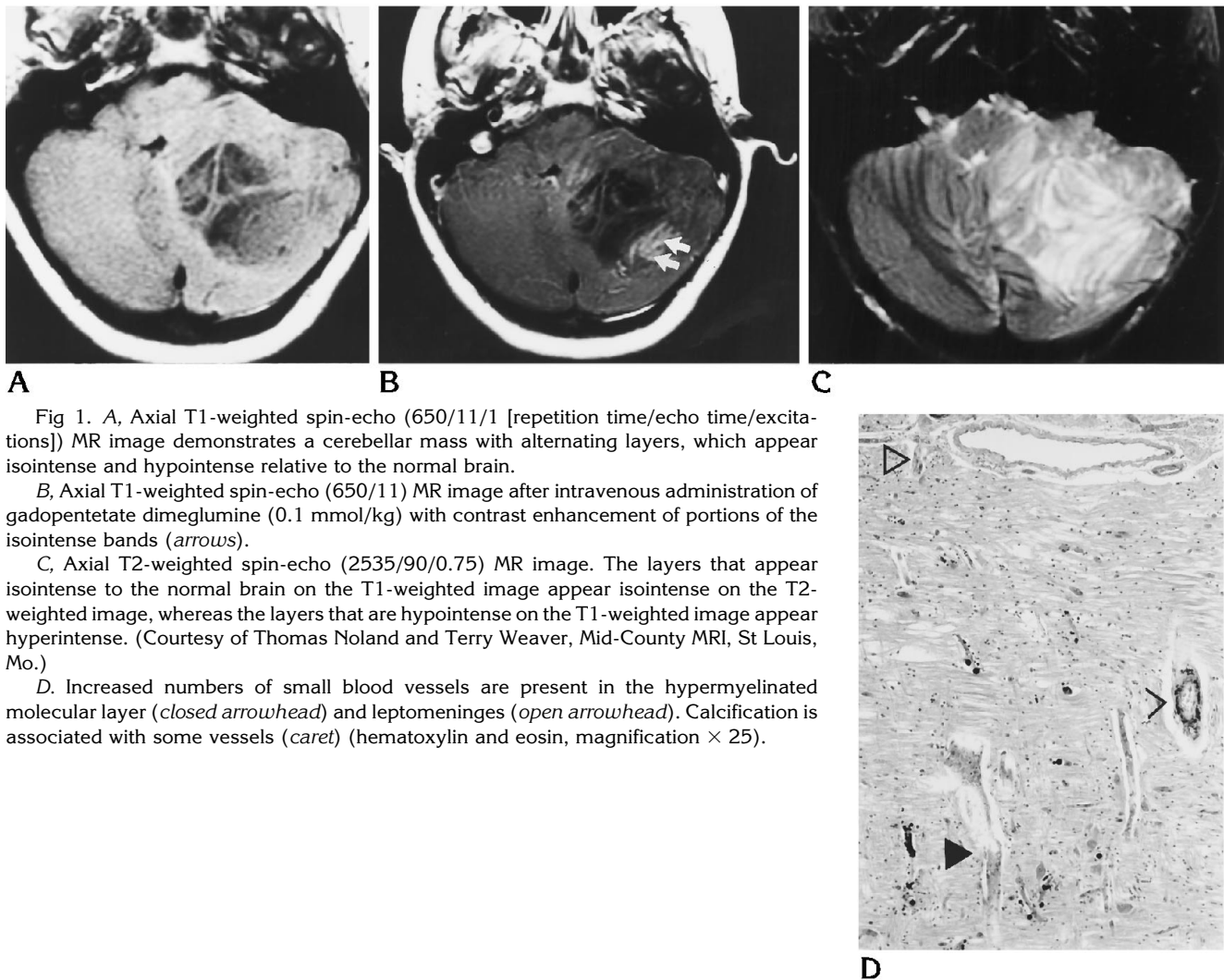


Fig 1. A, Axial T1-weighted spin-echo (650/11/1 [repetition time/echo time/excitations]) MR image demonstrates a cerebellar mass with alternating layers, which appear isointense and hypointense relative to the normal brain.

B, Axial T1-weighted spin-echo (650/11) MR image after intravenous administration of gadopentetate dimeglumine (0.1 mmol/kg) with contrast enhancement of portions of the isointense bands (arrows).

C, Axial T2-weighted spin-echo (2535/90/0.75) MR image. The layers that appear isointense to the normal brain on the T1-weighted image appear isointense on the T2-weighted image, whereas the layers that are hypointense on the T1-weighted image appear hyperintense. (Courtesy of Thomas Noland and Terry Weaver, Mid-County MRI, St Louis, Mo.)

D, Increased numbers of small blood vessels are present in the hypermyelinated molecular layer (closed arrowhead) and leptomeninges (open arrowhead). Calcification is associated with some vessels (caret) (hematoxylin and eosin, magnification $\times 25$).

should not exclude the diagnosis in the presence of characteristic MR findings.

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