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MR Imaging of Lhermitte-Duclos Disease: A Case Report

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Since 1920, when Lhermitte and Duclos first described a tumorlike abnormality of the cerebellum, which has come to bear their name, there has been an increasing number of reports of this rare entity [1–10]. Most cases have not had radiographic correlation, but there are several reported cases in which CT was part of the pretreatment evaluation [2–7], and one case was demonstrated on low-field MR imaging [6, 7]. We report a biopsy proved case of Lhermitte-Duclos disease imaged by CT and high-field MR imaging.

Case Report

A 43-year-old woman was well until 1 year prior to admission. At that time she developed severe headaches with no other associated symptoms. Six months before admission she noticed a sense of disequilibrium with a tendency to fall to the left. A CT scan of the neck was negative. One month before admission, despite conservative therapy, she developed gradual and progressive decline in visual acuity in her left eye.

Examination disclosed a visual acuity on the left of hand motion only while the right eye had a corrected acuity of 20/30. A left afferent pupillary defect was noted as well as slight pallor of the left disk. The right eye was unremarkable. No convergent eye movements were present but the remainder of the extraocular movements were full bilaterally and the remainder of the cranial nerves were within normal limits. Motor and sensory examinations were normal. The patient was noted to have past-pointing to the left as well as a tendency to fall to the left. Reflexes were hyperreflexic on the left with a left extensor plantar response.

A CT examination of the head demonstrated a low-density non-enhancing lesion of the left cerebral hemisphere (Fig. 1). Significant mass effect upon and displacement of the fourth ventricle was seen as well as a suggestion of slight left posterolateral impingement on the brainstem. Enlargement of the left cerebellar fossa and associated thinning of the adjacent bone were noted. Enlargement of the lateral and third ventricles was evident, consistent with obstructive hydrocephalus. An MR (GE Signa 1.5 T) examination of the brain was performed. A sagittal image, 600/20 (TR/TE), showed an inhomogeneous mass in the left cerebellum (Fig. 2A). Serpiginous low-signal areas, suggesting folia, were identified in the mass. Axial images, 2500/20,80 (Figs. 2B–2D), demonstrated focal enlargement of the left cerebellar hemisphere. Relatively homogeneous high-signal abnormality in the involved area was noted with relatively well-defined

margins. Mass effect on the brainstem was clearly shown, as were the effects of the posterior fossa. Hydrocephalus was again noted. On the basis of the imaging studies a diagnosis of Lhermitte-Duclos disease was postulated, although an atypical appearing glioma could not be entirely excluded.

The patient underwent surgery for decompression and partial resection. The lesion at surgery appeared as a swollen mass with associated flattened gyri and compressed sulci. The mass was difficult to distinguish from adjacent and deep normal cerebellum. This relatively hypovascular lesion had a poor cleavage plane, and it was resected in fragments up to a total of 3 × 3 cm.

Pathology demonstrated a thickened molecular layer with hypermyelination, an abnormal granular layer, and an absence of Purkinje cells consistent with Lhermitte-Duclos disease.

The patient did well postoperatively with resolution of the headaches and disequilibrium; however, the left visual findings remain relatively unchanged and are probably secondary to a postcompressive optic neuropathy.

Discussion

In 1920, Lhermitte and Duclos described a cerebellar abnormality consisting of focally enlarged hemispheric folia containing abnormal ganglion cells in the granular layer, with a thickened and hypermyelinated molecular layer and associated loss of Purkinje cells [1]. Since that time the entity has been called by a variety of names, including Lhermitte-Duclos disease, granular cell hypertrophy, dysplastic gangliocytoma of the cerebellum, benign hypertrophy of the cerebellar cortex, and Purkinjeoma. This confusion is, for the most part, due to the uncertainty of whether to classify this disease as a dysplasia or a neoplasm. Roessmann and Wongmongkolrit [8] described a case in a newborn, while Ambler et al. [9] noted a familial case involving a mother and son. Associated abnormalities have also been described, including megalencephaly, hydromyelia, heterotopia, polydactyly, microgyria, and multiple hemangiomas [2, 9]. These findings tend to support a dysplastic origin. The basic lesion may be secondary to a disturbance affecting the differentiation and migration of granule cell precursors [3]. The abnormal inner granular layer contains large neurons with rounded nuclei and abundant

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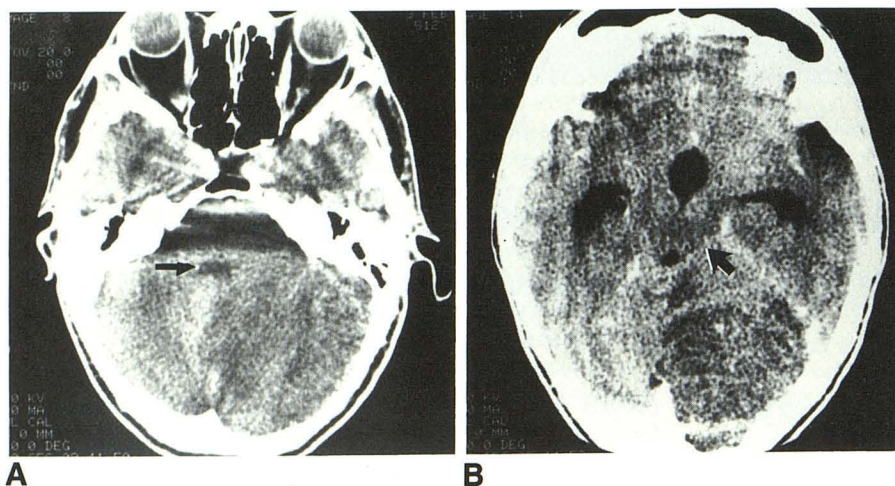


Fig. 1.—A, Unenhanced axial CT scan demonstrates a low-density mass in left cerebral hemisphere with mass effect on fourth ventricle (arrow). Enlargement of involved hemisphere is noted.

B, Enhanced axial CT scan shows a low-density, nonenhancing mass in left cerebellum with mass effect on brainstem (arrow) and associated ventricular enlargement.

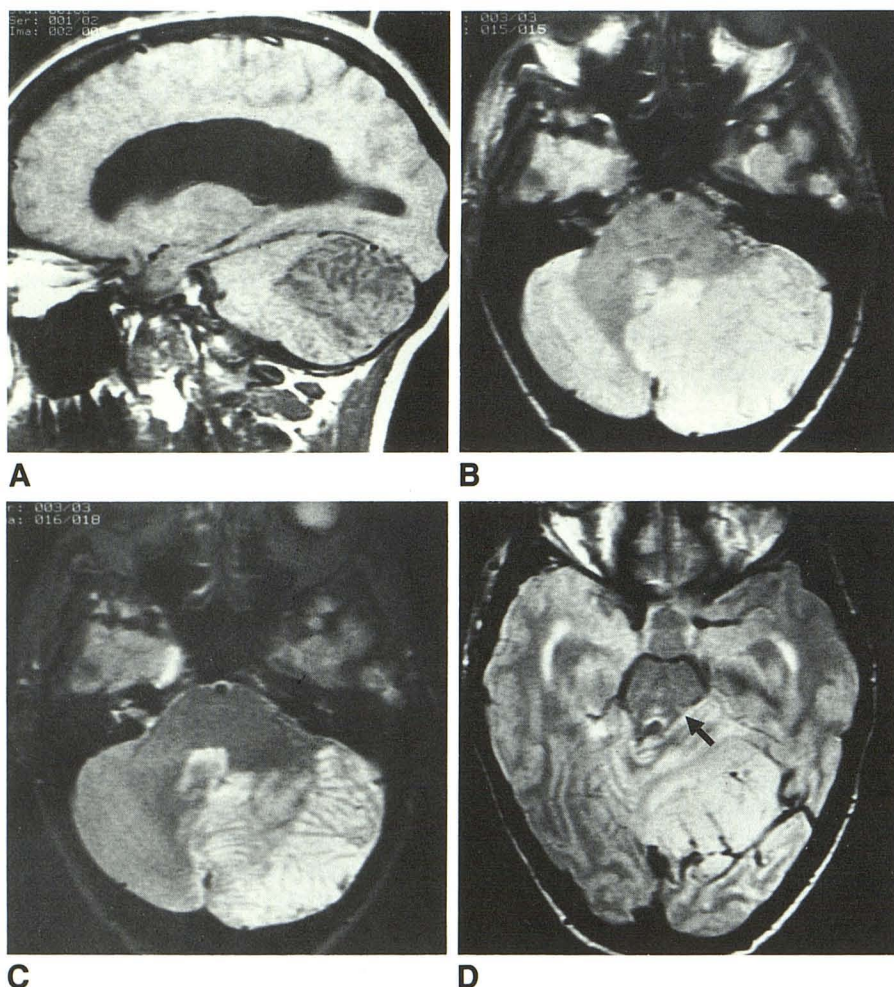


Fig. 2.—A, Sagittal MR image, 600/20 (TR/TE), shows a relatively well-defined inhomogeneous but mostly low-signal lesion in left cerebellar hemisphere. Some low-signal areas are curvilinear, possibly suggesting prominent folia.

B, Axial MR image, 2500/20, shows increased signal, relatively homogeneous, in an enlarged left cerebellar hemisphere. The margins appear well-defined.

C, Axial MR image, 2500/80, same level. There are some linear isointense bands within the high signal.

D, Axial MR image, 2500/20, shows the mass extending anteriorly to impinge upon posterolateral aspect of midbrain (arrow). Enlargement of temporal horns and third ventricle is seen.

mitochondria [4, 11]. The outer molecular layer contains thickened hypermyelinated axons with no Purkinje dendrites [11]. There is also a loss of Purkinje cell bodies and the loss of the central core of white matter in the axis of the cerebellar folia. The involved areas "hypertrophy" in size leading to mass effect. No significant invasion or edema is noted.

Clinically, the lesion has a slight female preponderance and generally presents in the third or fourth decade [5]. The lesion may be asymptomatic, with many cases discovered at autopsy. The abnormality is hemispheric and unilateral. Those cases that become symptomatic appear to do so secondary to their mass effect, causing intracranial hypertension and

cerebellovestibular signs. Very rare instances of acute respiratory arrest are associated with the disease. In one report, a patient demonstrated visual disturbances that included a scintillating scotoma [5].

In the five reported cases with CT correlation, all demonstrated a hypodense nonenhancing mass in the posterior fossa [2-7]. Three had obstructive hydrocephalus on CT, and three had focal calcification associated with the lesion. There has been enlargement of the affected posterior fossae with associated thinning of the adjacent occipital squamosa in four cases and in our own.

Our case demonstrated an inhomogeneous signal on short TR/short TE MR. A suggestion of prominent cerebellar folia was noted. On long TR/long TE there was abnormal increased signal intensity in the cerebellar hemisphere with a good margin identified. The signal characteristics may be secondary to increased intracellular water in the hypertrophied granular cell layer, since the absence of extracellular edema and the presence of hypermyelination, as noted in the neuropathologic references, does not explain the high signal intensity [11, 12]. Extension into the left middle cerebellar peduncle was better seen on MR than on CT, as was the brainstem compression. The previously reported case with MR demonstrated low signal on short TR/short TE and high signal on long TR/long TE images on a 0.3-T system [6, 7].

Because of the lack of invasiveness and slow growth associated with this entity, surgery appears to be the treatment of choice. So far there have been no reports of death associated with recurrence [4]. At surgery, the reported cases and our own demonstrated broad and flattened folia with no apparent change in color, consistency, or structure. As a result, the margins are difficult to ascertain.

In summary, the combination of a hypodense, nonenhancing mass in the posterior fossa with adjacent squamosal thinning and unilateral hemispheric expansion on CT in a

middle-aged adult should raise the possibility of Lhermitte-Duclos disease. The MR appearance consisting of hemispheric enlargement, abnormal signal intensity on short and long TR images, and prominent folia may be more specific. The margins of this process are very difficult to identify at surgery and are better delineated by MR, which can thus define the extent of lesion resection.

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