

# Clinical and Radiological Aspects of Dysplastic Gangliocytoma (Lhermitte-Duclos Disease): A Report of Two Cases with Review of the Literature

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Two cases of Lhermitte-Duclos disease confirmed by biopsy are reported. Review of the 58 published cases shows that the disease can manifest itself only by signs of increased intracranial pressure. Cerebellar symptoms are not constant. Computed tomographic (CT) scans suggest the diagnosis by showing a posterior fossa lesion, iso- and hypodense, partially calcified, and not enhanced by contrast medium. Magnetic resonance imaging (MRI) seems to define limits of the lesion better than CT scanning and could improve the surgical approach. Surgical excision of the lesion is the only satisfactory treatment. The postoperative prognosis is usually favorable. (*Neurosurgery* 22:124-128, 1988)

Key words: Cerebellum, Computed tomographic scanner, Dysplastic gangliocytoma, Lhermitte-Duclos disease, Magnetic resonance imaging

## INTRODUCTION

Dysplastic gangliocytoma of the cerebellum is an uncommon lesion. Since the first report by Lhermitte and Duclos in

1920 (19), we found 58 cases in the literature (1-29). Clinically, the disease appears most often in adults as an expanding lesion of the posterior fossa. The diagnosis has never been suspected preoperatively (2, 4). The purpose of this report is

to discuss clinical and radiological aspects of Lhermitte-Duclos disease that could support a preoperative diagnosis.

### CASE REPORTS

#### Case 1

This 25-year-old man was admitted in December 1978 with a 3-year history of intermittent headaches with blurring vision. The history of this case was previously published (25). Neurological examination showed a moderate left hypotonia. There was bilateral papilledema, and tests of labyrinthine function pointed to a left cerebellar lesion. The general examination was normal. Plain x-ray films of the skull were normal. A brain computed tomographic (CT) scan revealed a roughly limited, expanding lesion of the left posterior fossa. This area was of mixed density (iso- and hypodense), was partially calcified, and did not enhance on the injection of contrast medium. The 4th ventricle was not demonstrated, and supratentorial hydrocephalus was observed (Fig. 1). Vertebral angiography confirmed the presence of a nonvascularized mass in the left cerebellar hemisphere.

A suboccipital craniectomy performed with the patient sitting allowed the excision of a large, poorly limited lesion. The folia of the left cerebellar hemisphere were irregularly thickened and pearl gray. The removal seemed to be total.

On gross examination, the excised mass was characterized by a mixture of normal and enlarged convoluted folia (Fig. 2). Histological examination showed that the enlarged folia were constituted by two layers: an internal area of large neurons and an outer band of white matter (Fig. 3). Modifications between normal and pathological structures were gradual, without a clear-cut separation. In transitional territories, progressive changes could be observed: (a) widening of the molecular layer occupied by large myelinated fibers and small nucleated cells; (b) rarefaction and disappearance of Purkinje cells; (c) widening of the granular layer containing more and larger neurons that were more widely spaced than normal (no multinucleated cells or mitoses could be found); (d) progressive reduction and disappearance of the central white matter.

The postoperative course was uneventful, and the patient was discharged after 12 days. Two years later, he was reexamined because of nocturnal seizures. His neurological status

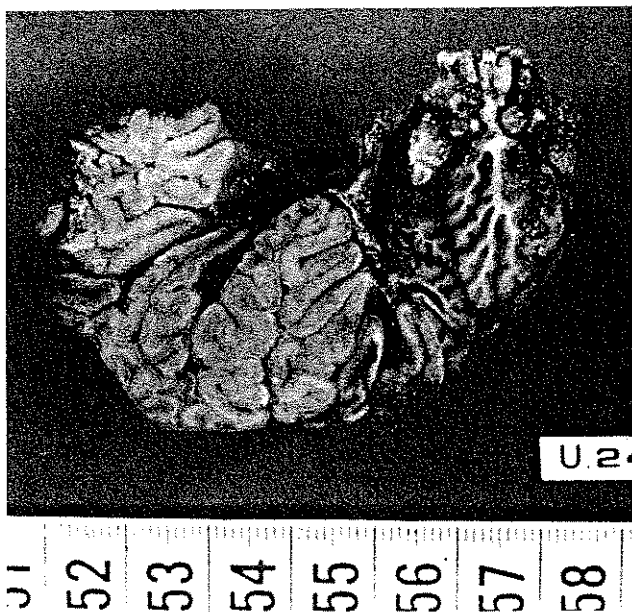


FIG. 2. Case 1. Macroscopic view of the resected tissue showing some normal cerebellar folia and abnormal, enlarged convoluted folia.

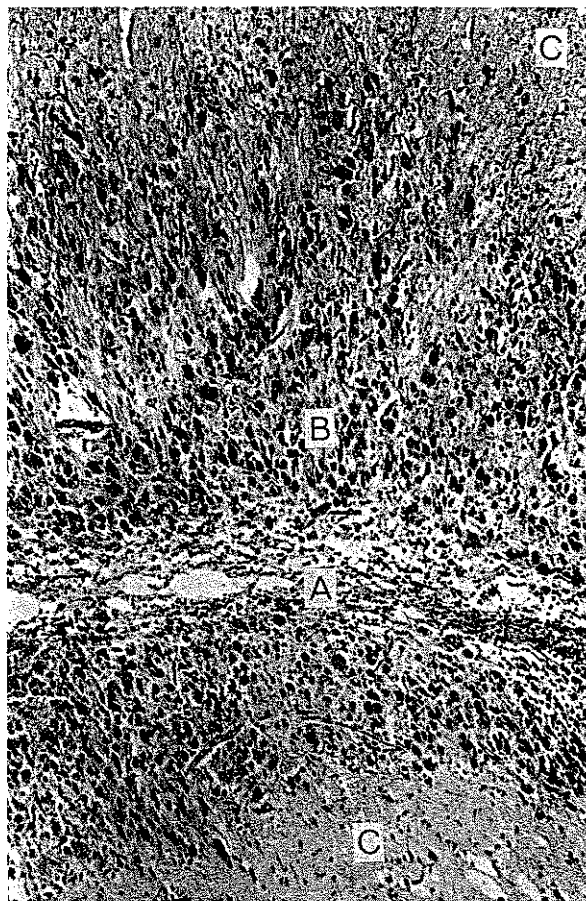


FIG. 3. Histological aspect of the lesion: A, laminated central white matter; B, layer of hypertrophied neurons; C, molecular layer (hematoxylin and eosin,  $\times 150$ ).

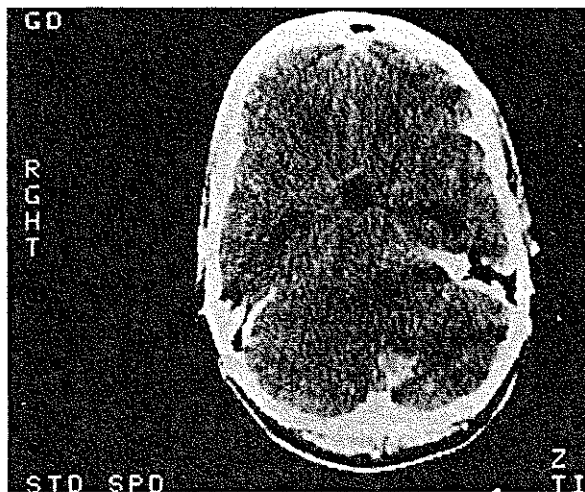


FIG. 1. Case 1. Brain CT scan showing a left posterior fossa lesion of mixed density, partially calcified. The 4th ventricle is not detectable.

was normal. A control CT scan did not show residual lesion, and he was treated with phenobarbital.

Eight years after operation, the patient (who had continued his business successfully) came back with rhinorrhea. Neurological findings remained normal. Brain CT scanning revealed no posterior fossa lesion, but demonstrated a large mucocele of the frontal sinus eroding the posterior wall.

### Case 2

This 59-year-old woman was admitted with a 1-month history of progressive headaches without nausea or vomiting. A recent disturbance of memory had been observed. She had had a reeling gait for more than 10 years.

Neurological examination showed a markedly ataxic gait and bilateral dysmetria, especially on the left side. No nystagmus was noted. The deep tendon reflexes were increased in both upper and lower extremities, with a bilateral Babinski sign. No other abnormalities were found on clinical examination.

A brain CT scan showed an enlarged left side of the posterior fossa. A large, poorly limited, left cerebellar lesion extending to the midline was observed. This area of mixed density, hypo- and isodense, was partially calcified and did not enhance after the injection of contrast medium. The 4th ventricle was displaced but quite visible (Fig. 4), and the supratentorial ventricular system was enlarged. Vertebral angiography confirmed the presence of a nonvascularized mass in the left cerebellar hemisphere with signs of tonsillar herniation.

The operation was performed in a sitting position. The left cerebellar cortex was obviously abnormal: the folia were thickened and pearl-gray. This appearance prevailed throughout most of the left cerebellar hemisphere and extended into the vermis. A frozen section revealed the diagnosis, and a subtotal resection of this poorly demarcated "benign" lesion was carried out. Histologically, the cerebellar alterations were similar to those in the previous case.

Postoperatively, the left ataxia and gait disturbance were increased, but on discharge at the 12th day the patient could walk alone. On follow-up examination 6 months later, she was still ataxic but continued to improve; there were no headaches. The Babinski sign had disappeared. A control CT scan (Fig. 5) showed the sequelae of a large resection at the site of the calcified area. The residual tissue of the left hemisphere was isodense, but it was not possible to be sure that the lesion had been totally removed. Magnetic resonance imaging (MRI) revealed a large area of increased signal on a

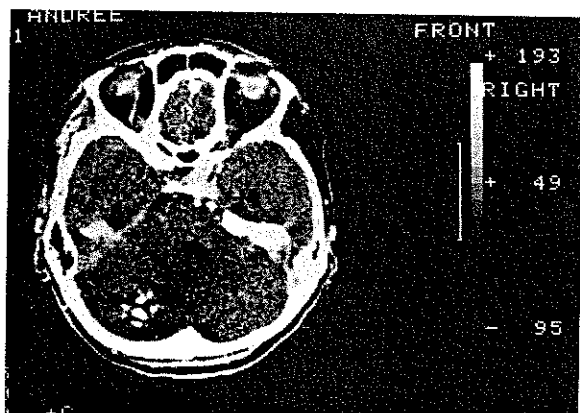


FIG. 4. Case 2. Brain CT scan showing the enlarged left posterior fossa and a partially calcified cerebellar lesion of mixed density displacing the 4th ventricle.

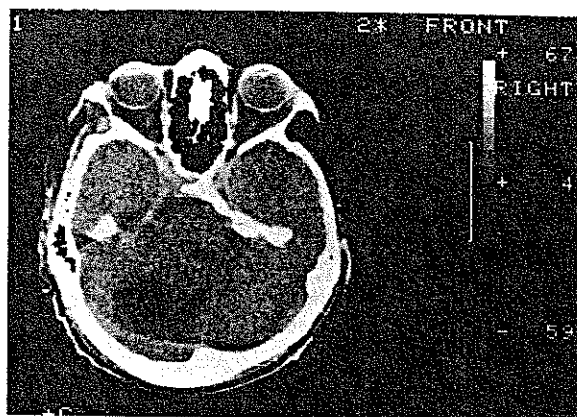


FIG. 5. Case 2. Control CT scan. Large excision at the site of the previous calcified lesion.

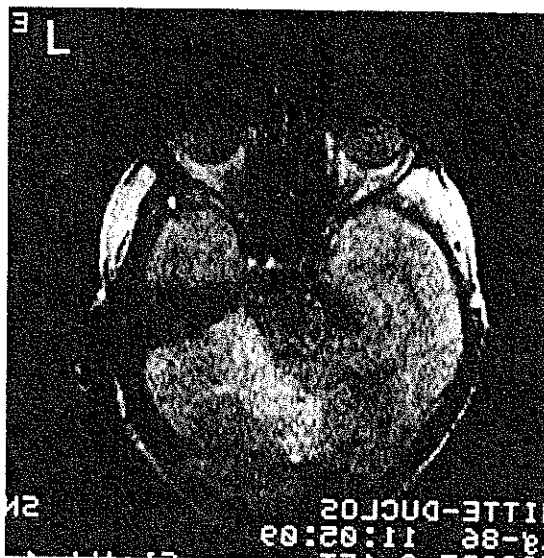


FIG. 6. MRI scan, axial view. The lesion appears as a large area of increased signal on a T2-weighted image. The pathological tissue occupies all of the left residual cerebellar hemisphere and extends beyond the midline, but without invading the brain stem.

T2-weighted image. Pathological tissue was seen in the entire residual left hemisphere, extending beyond the midline but not invading the brain stem (Fig. 6).

## DISCUSSION

### Clinical data

Lhermitte-Duclos disease occurs most often in young adults. At the time of diagnosis, the mean age ranges from 0 to 74 years, with an average of  $34 \pm 14$  years (Fig. 7) (16, 27). The sex ratio is approximately equal.

Clinically, the lesion manifests itself as a slowly growing mass in a cerebellar hemisphere. According to the literature, the duration of symptoms ranges from a few months to more than 10 years (1, 11, 18); increased intracranial pressure is the most constant feature. Cerebellar signs can be minimal and were absent in half of the cases (1, 11) (Table 1). In 20% of the patients, the cerebellar symptoms were prominent and intracranial hypertension was slight or even absent (Case 2). Association with other abnormalities (polydactyilia, hydro-myelia, dysplastic body, and megalencephaly) is frequent (1, 4, 5, 7, 11, 23-27, 29). Some authors have emphasized that,

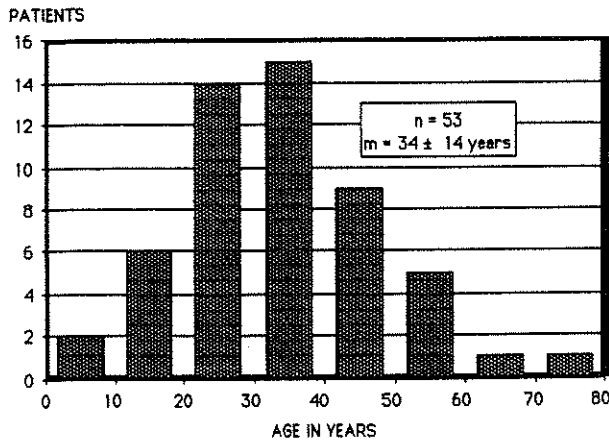


FIG. 7. Age distribution among the 53 patients with cerebellar gangliocytoma at the time of diagnosis.

TABLE 1  
Clinical Presentation (32 Cases Compiled from the Literature)

Presentation	
Headaches	75%
Instability while walking	20%
Symptoms	
Papilledema	70%
Cerebellar syndrome	50%
Megalencephaly	40%
Cranial nerve deficit	30%
Pyramidal syndrome	20%

after a long evolution, sudden decompensation and death may occur (1, 18).

#### Radiological data

Skull x-ray films can reveal an enlarged head, signs of chronically increased intracranial pressure, and local deformity of the skull (thin and ballooning occipital bone) (1, 7, 9, 17, 18, 23).

Brain CT scans have been performed preoperatively in our two patients and in four previous cases (3, 12, 24, 28). In all of these cases, the lesion was described as a large, poorly limited hemispheric mass, of mixed density (hypo- and iso-dense). Calcifications were seen in three cases. The lesion never enhanced after the injection of contrast medium. Local deformity of the skull could be seen (Case 2) (12).

Vertebral angiography confirmed the presence of a hemispheric avascular mass (4, 17, 24, 28, 29).

We are reporting the first study of such a lesion by MRI, although it was performed 3 months after partial removal of the abnormal tissue. The residual lesion appeared as a zone of increased signal on a T2-weighted image. The comparison of a postoperative CT scan and MRI revealed that MRI is a better technique for delineating the lesion.

#### Pathological aspects

The pathogenesis of this lesion is still unclear, but Lhermitte-Duclos disease seems to represent a congenital abnormality in granule cell migration and development rather than a true neoplasm (11, 25, 26, 28). Gross examination reveals segmental enlargement of the cerebellar folia, sometimes extending into the vermis (9, 23). In this abnormal region, there is an unusual gyral pattern made by greatly enlarged convoluted folia usually paler than normal folia.

Microscopic examination shows a severe distortion of the normal architecture (1, 7, 8, 11, 18, 26, 29). The molecular layer is broadened by abundant, enlarged, and irregularly myelinated axons running parallel to the subpial region. In deeper parts, these fibers are perpendicular to the surface (2, 23). These axons belong to hypertrophied neurons composing the inner layer. Most authors agree that light and electron microscopy show that these abnormal neurons are hypertrophied granule cells (1, 9, 11, 12, 14, 24, 25, 28). The transition from normal to abnormal cortex is gradual and accompanied by the disappearance of Purkinje cells and a severe reduction of the central white matter. Interstitial calcifications are often observed in the boundary zone along the smaller blood vessels (2, 5, 17, 23, 24, 28). Mitoses have not been reported (5, 8, 11, 17, 23, 26).

#### Treatment

Surgical excision is the only appropriate treatment (5, 12, 18, 26, 28). The absence of limits in the depth of the cerebellar hemisphere constitutes the only major technical problem (1, 9, 17, 23, 24). This macroscopic aspect is confirmed by histological examination demonstrating a "transitional" zone between normal and pathological cortex (1, 7, 9, 12, 23, 26). A better resolution of the lesion's extension by preoperative MRI will probably be helpful in planning the surgical treatment.

#### Postoperative prognosis

It is still difficult to establish the postoperative prognosis. Until 1955, only three of the eight patients had survived surgical excision (1, 7). Probably due to progress in anesthesiology and surgical techniques, the postoperative prognosis now seems much better, and all recent cases exhibited a good short term evolution after surgical treatment (4, 18).

According to some authors, the slow growth rate of this lesion could allow long term survival despite partial excision (1, 4, 18, 26). Unfortunately, the majority of reported cases had a short follow-up at the time of their publication.

The long term natural history of this disorder is not yet known, but recurrence may develop (2, 20). Therefore, the surgical procedure should be as radical as possible especially in young patients. Like Marano (20), we think that a large excision can lead to a better life expectancy.

#### CONCLUSION

Dysplastic gangliocytoma is an uncommon lesion of the posterior fossa involving the cerebellum. This disease should be suspected when CT scans show a large, poorly limited, and partially calcified lesion of a cerebellar hemisphere without contrast enhancement in a young adult complaining of slowly progressive instability while walking and headaches. A surgical approach is the only treatment. MRI, by better demarcating the boundaries of the lesion, could allow a larger and safer surgical excision.

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## COMMENT

There are still relatively few well-documented examples of cerebellar dysplastic gangliocytomas. The authors provide a good description and analysis of two cases in a 25-year-old man and a 59-year-old woman, respectively. In addition to CT scans, they also provide, for the first time, MRI findings and suggest that this method delineates the lesion more precisely and also provides some clues as to its nature. Precise preoperative delineation of these lesions and a reasonable prediction of their histological character by MRI will facilitate successful neurosurgical treatment of this entity.

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