Lhermitte-Duclos Disease

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Summary. A new case of Lhermitte-Duclos disease (dysplastic gangliocytoma of the cerebellum) was recognized in an adult man after neurosurgery. Conventional, Golgi, and electron-microscopic investigations support the view that this rare lesion results from a progressive hypertrophy of granular cell neurons with excess of myelination of their axons in the molecular layer while inducing the local disappearance of Purkinje cells and the central core of the cerebellar folia. After a follow-up of 4 years, the patient now leads a normal life; no recurrence has occurred and no other neuropathologic alteration have been detected.

Key words: Cerebellum - Dysplastic gangliocytoma - Lhermitte-Duclos disease

Introduction

After the publication of Lhermitte and Duclos (1920), attention has been drawn to a rare disease of the cerebellum about which less than 40 cases have been published (see Ambler et al. 1969; Ferrer et al. 1979 for the previous literature). Generally discovered in adults, this lesion manifests itself as a slowly growing mass formed by abnormal neurons that appear more malformative than tumoral. The morphology and the origin of these neurons arranged in folia with a cortex-like structure have been differently interpreted, thus leading to various denominations: Lhermitte-Duclos disease, granular cell hypertrophy, benign hypertrophy of cerebellar cortex, Purkinjeoma, ganglioneuroma, dysplastic gangliocytoma of the cerebellum.

With only one familial example (Ambler et al. 1969), this lesion has been eventually associated with other anomalies of the brain or viscera. At present, only ten biopsied cases have been reported in the literature (Christensen 1937; Oppenheimer 1955; Carbone et al. 1955; Brucher and Vandenbergh 1959; Christian 1961; Cook et al. 1962; Bellamy et al. 1963; Harff and Vincken 1966; Daum et al. 1967; Ferrer et al. 1979) with a survival duration varying from a few days up to 3 years at the time of publication.

In this paper, we report a new surgical case with a perfect clinical evolution and a morphological study confirming that the main pathological process appears as a graded hypertrophy of granular cell neurons in the mature cerebellar cortex.

Case Report

Clinical History

Born in 1953, this white male had a normal development. In 1975, because of transient headaches, ENT and ophthalmologic examinations were performed and considered as normal. In September 1978, after recurrence of headache with blurring of vision and a neurologic status showing left-side hypotonia, a brain CT scan was made that revealed a partially calcified mass in the left posterior fossa and moderate signs of hydrocephaly (Fig. 1). Among other investigations, a vertebral angiography confirmed the presence of a non-vascularized mass in the left cerebellar hemisphere with anterior displacement of the brain stem and asymmetric herniation of both cerebellar amygdalae. A posterior craniotomy permitted the excision of a limited mass (5 x 3 x 2 cm) from the left cerebellar hemisphere. The neurosurgeon had indicated that it was hazardous to differentiate this mass from the normal cerebellar structures. Clinical evolution was satisfactory, and the patient was discharged from the hospital 15 days after surgery.

In March 1979, a control brain CT scan confirmed the excision of the lesion. In October 1980, the patient was re-examined in the hospital because he suffered a few nocturnal seizures. His neurologic status was considered normal. EEG was unspecific. To investigate the possibility of associated lesions, a new brain CT scan was performed but no other modification could be detected. The patient was treated by administration of 5 cg Gardental twice daily.

In March 1982, he continued to present a few nocturnal seizures without incontinence. Clinically, there was moderate left-side hypotonia. No sign of phacomatosis could be detected. A control CT scan did not reveal encephalic change. At that time, the patient had become an official customs agent and he continued his lifework with success.
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Fig. 1. Brain CT scan showing a partially calcified mass in the left posterior fossa.

Fig. 2. Cut surface of the excised mass exhibiting some normal cerebellar folia (upper 1/3) and large convoluted lamellae (lower 2/3). Bar: 1 cm

Fig. 3. Slightly modified cerebellar cortex with area of normal granular cell neurons (single arrow) and layer of hypertrophied neurons (double arrow). Nissl, × 50

Fig. 4. Detail of the altered cortex. From top to bottom: A molecular layer; B area of hypertrophied neurons; C laminated spongious central core; D hypertrophied neurons in the contiguous lamella. Nissl, × 150

Neuropathology

Gross examination of the excised mass revealed two different areas (Fig. 2). On one side, normal cerebellar folia were easily recognized with the cortex approximately 1 mm thick. On the other side which constituted the larger part of the lesion, there was a gyral pattern made by greatly enlarged convoluted folia 2–3 mm wide. In this abnormal region, a continuous laminar myelinated band was observed outside the broadened gray substance, while no subjacent white matter seemed to exist in the center of the folia.

Gradual modifications of the structures were discovered between these two sides, without a clear-cut separation.