LHERMITTE-DUCLOS DISEASE IN A YOUNG ADULT CASE REPORT

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SUMMARY :

Dysplastic gangliocytoma of the cerebellum (Lhermitte-Duclos diease) is a rare disorder characterized by a slowly enlarging mass lesion in the cerebellum. In this paper, our experience with a case of Lhermitte-Duclos disease with an unusual vermian localization is reported and our findings are discussed in the light of the recent literature.

KEY WORDS:

Cerebellar neoplasm, Cerebellar dysplastic gangliocytoma, Lhermitte-Duclos disease.

INTRODUCTION:

Dysplastic gangliocytoma of the cerebellum is a very rare lesion. The other names assigned to this pathology are Lhermitte-Duclos disease, ganglioneuroma, hamartoma of the cerebellum, purkengioma, granule cell hypertrophy or granulomolecular hypertrophy of the cerebellum. Only few cases have been reported in the literature (1-20), since the first report by Lhermitte and Duclos (11). Clinically, the disease appears most often in young adults as a slowly expanding lesion of the posterior fossa, specifically originating from the cerebeller hemispheres. The pathophysiology of the disease is poorly understood, and this accounts for the multiple names assigned to the entity (1,7). The most important point is that the diagnosis has never been suspected preoperatively in all published cases.

In this paper, we present a case of Lhermitte-Duclos disease with an unusual vermian localization. Our purpose is to discuss the clinical and radiological aspects of the disease that could support a preoperative diagnosis.

CASE REPORT:

A 17-year-old female patient was admitted to our clinic with the complaints of headache and progressively increasing gait-difficulty for one year. Her symptoms had worsened over the last week and especially vomiting had occurred for the last three days. Neurological examination revealed bilateral papilloedema, bilateral horizontal nystagmus and cerebellar disturbances. Computerized tomography (CT) demonstrated an isodense, poorly limited vermian mass, that extended to the level of the tentorial notch. The IVth ventricle was not visible, and there was mild dilatation of the supratentorial ventricular system with bilateral tonsillar herniation. The mass did not show contrast enhancement and contained multiple small areas of calcification (Figs: la and 1b).

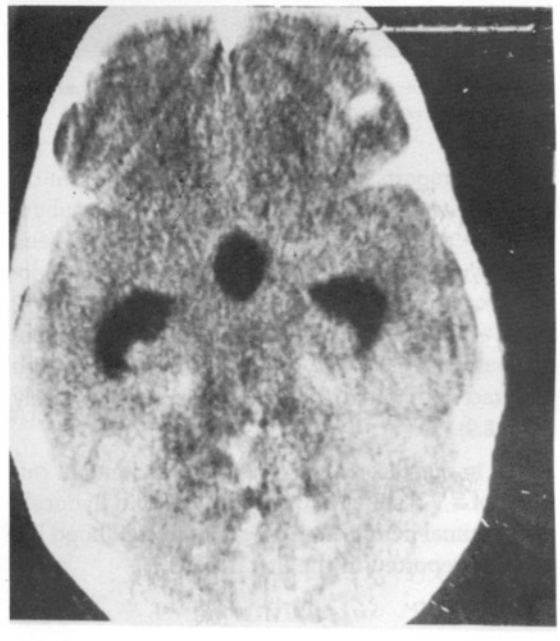

Figure 1a : Preoperative CT scans revealed a vermian mass without demarcation. The lesion contains areas of calcification. The IVth ventricle is not visible.

Operation: On the day after admission, with a diagnosis of posterior fossa tumour, a suboccipital craniectomy was performed with the patient in the prone position. After the dural opening, the irregu

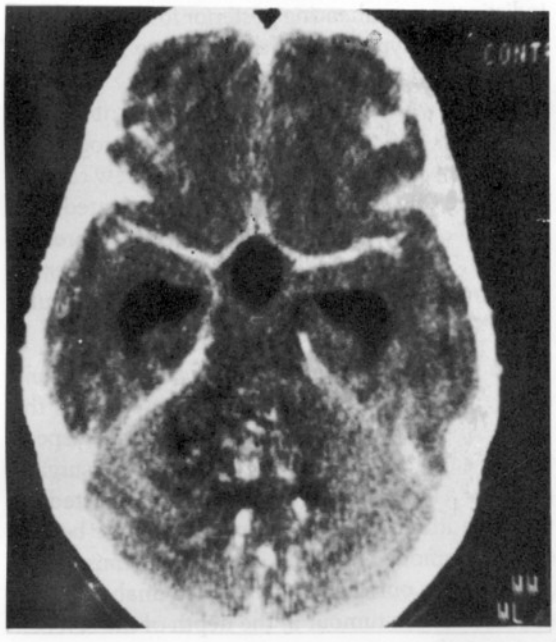

Figure 1b : The mass did not show contrast material enhancement.

larly thickened and peraly-gray folia of the vermis was exposed. A vertical incision of the vermis was performed and a tumour which indistinctly merged with the both lobes of the cerebellum was observed. Subtotal resection of this poorly demarcated lesion was carried out. In addition, cervical laminectomies of C1 and C2, and a large duraplasty was performed for decompression of bilateral tonsillar herniation.

Histopathological examination revealed a very distinctive architectural derangement. The outer cell layer showed markedly hypertrophic myelinated and unmyelinated fibres. Additionally, the inner layer demonstrated abnormal neurons, some of which resembled Purkinje cells. Another feature of this particular tumour was the grandual transition of the cells from normal to abnormal cortex and accompanying disappearance of the Purkinje cells. Both PTAH and Bielchowsky stains identified intercellular neurological fibres and myelin sheats (Figs: 2a and 2b).

Follow-up: On the 2nd postoperative day a ventriculo-peritoneal shunt was inserted because of neurological deterioration and dilatation of the supratentorial ventricles. Following V-P shunting, the patient's clinical status improved rapidly and she was discharged with only mild cerebellar disturbance on the 14th postoperative day. A control CT revealed the

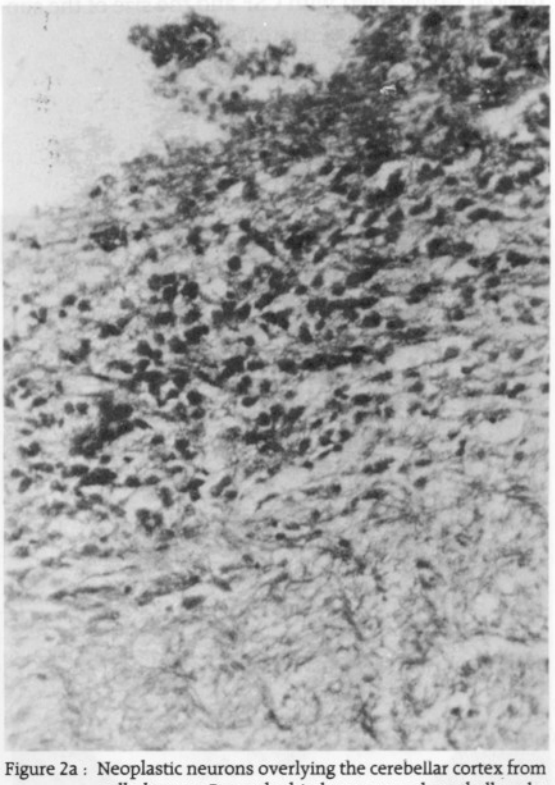

Figure 2a : Neoplastic neurons overlying the cerebellar cortex from cell clusters. Beneath this layer normal cerebellar elements may be seen (HEx100 original magnification).

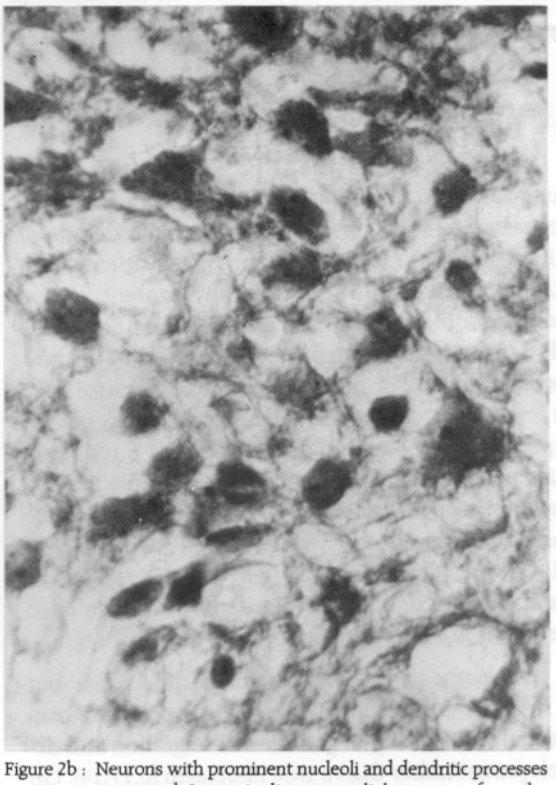

Figure 2b : Neurons with prominent nucleoli and dendritic processes are noted. Intermingling neuroglial processes from the background (neurophil) (Phospotungstic acid-Hematoxylin X400).

tumour cavity filled with CSF and the size of the supratentorial ventricles decreased (Fig: 3).

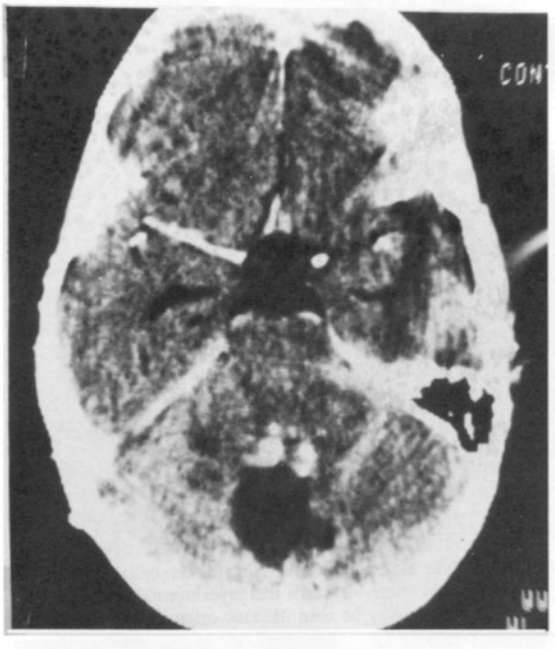

Figure 3 : Postoperative CT scan showing the tumour cavity and a small residual part of the tumour.

DISCUSSION:

Lhermitte-Duclos disease occurs most often in young adults. According to the literature, the majority of cases were diagnosed in the 3rd of 4th decade of life (9,18). There is no sex dominance. Patients with Lhermitte-Duclos disease typically present with a long-standing history of neurological abnormalities due to the insidious expansion of a mass in the posterior fossa (1). The duration of symptoms range from a few months to more than 10 years, an average of approximately one year as in our case (1,6,10). Association with other abnormalities such as polydactylia, hydromyelia, megaloencephaly, heterotopia, multiple haemangioma and dysplastic body is frequent (1,4,6,14,17,20), but none of them was detected in our case. Sudden decompensation and death has been reported in some cases (1,10).

The pathogenesis of the lesion is still unclear, but L hermitte-Duclos disease seems to represent a congenital abnormality in granule cell migration and development rather than a true neoplasm (6,17,19). On the other hand, some authors have shown that the lesion has the potential for regrowth and can recur many years after total removal of the mass (12).

The neuroradiological diagnosis of Lhermitte-Duclos disease has never been suspected peroperatively, as in our case (2,4,13). CT scans revealed an indistinct, non-enhancing posterior fossa mass of mixed density (hypo and isodense), often with focal areas of calcification (3,15,17,19). This disease should be suspected when CT scans show the described view in a young aldult complaining of slowly progressive gait disturbance and headaches. Only a few cases of Lhermitte-Duclos disease have been evaluated with MRI and only two of them were performed preoperatively (8,12,13,16). We think that MRI, by better demarcating the boundaries of the lesion in this entity, could allow a correct diagnosis in the future.

The outcome in unoperated patients was uniformly poor in early cases, presumably due to the progressive nature of the disease (1). In this respect, the only appropriate treatment seems to be surgical excision. The major technical problem reported by various authors is the absence of a cleavage between the tumour and the cerebellar hemispheres (1,2,14,15). In our case, we were also unable to find the limits of the tumour in the depth of the cerebellar hemispheres.

The natural history of this disorder is not yet known. The slow growth rate of the lesion could allow long term survival despite partial excision (1,4,10). The efficacy of radiation therapy is unknown and is not recommended as an initial treatment (12). Because of recurrences, we believe that radical surgical resection, with shunting if necessary, is the treatment of choice in this entity and can lead to a better life expectancy.

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