



RESEARCH ARTICLE

DYSPLASTIC GANGLIOCYTOMA OF THE CEREBELLUM (LHERMITTE-DUCLOS SYNDROME) : A CASE REPORT

B.Taibi, O. Ayouche, Fz. Lamrani and L. Jroundi

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Abstract

Lhermitte-Duclos disease or dysplastic cerebellar gangliocytoma is a rare entity characterized by a hamartomatous lesion in the posterior fossa. Cowden's syndrome or hamartoma-neoplasia syndrome is a rare underdiagnosed autosomal dominant genodermatosis with a high incidence of malignant tumors. We report the case of a 42-year-old man presented with a cerebellar syndrome, Magnetic resonance (MR) imaging of the brain was performed and revealed a cerebellar left lesion suggestive of a Lhermitte-Duclos disease. At operation, a demarcated lesion was excised from the apparently normal surrounding cerebellar tissue and proved histologically to be Lhermitte-Duclos disease.

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Introduction:-

Lhermitte-Duclos disease, also known as dysplastic cerebellar gangliocytoma, is a rare tumor of the cerebellum. It is probably hamartomatous, although the exact pathogenesis remains unknown [2,6]. Even though it may not be neoplastic, it is considered a WHO grade I tumor in the current WHO classification of CNS tumors [1].

Case Report:-

A 42-year-old men, initially noted a vague distortion of vision in a diagonal strip across the vision of the right eye that resolved after a few days, one month after that, he presented with progressive headaches and ataxia.

On Examination a left afferent pupillary defect was noted as well as slight pallor of the left disk. The right eye was unremarkable, Reflexes were hyperreflexic on the left with a left extensor plantar response.

Computerized tomography (CT) of the head revealed a large indistinct mass in the posterior fossa with areas of calcification for better characterization a magnetic resonance imaging was performed in the axial, coronal, and sagittal planes. The axial images demonstrated a process diffusely enlarging the right cerebellar hemisphere. A coronal image indicated that the process involved primarily the cerebellar cortex, impinging on the cerebellar white matter. A gyriform pattern was seen corresponding to the enlarged but preserved folia of the cerebellar cortex. In addition, there was a little mass effect on the fourth ventricle and yet no hydrocephalus and on the brainstem. Both cerebellar tonsils were displaced below the foramen magnum.

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.

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 a resolution of the headaches and disequilibrium; however, the left visual findings remain relatively unchanged and are probably secondary to a most compressive optic neuropathy.

Figures:-

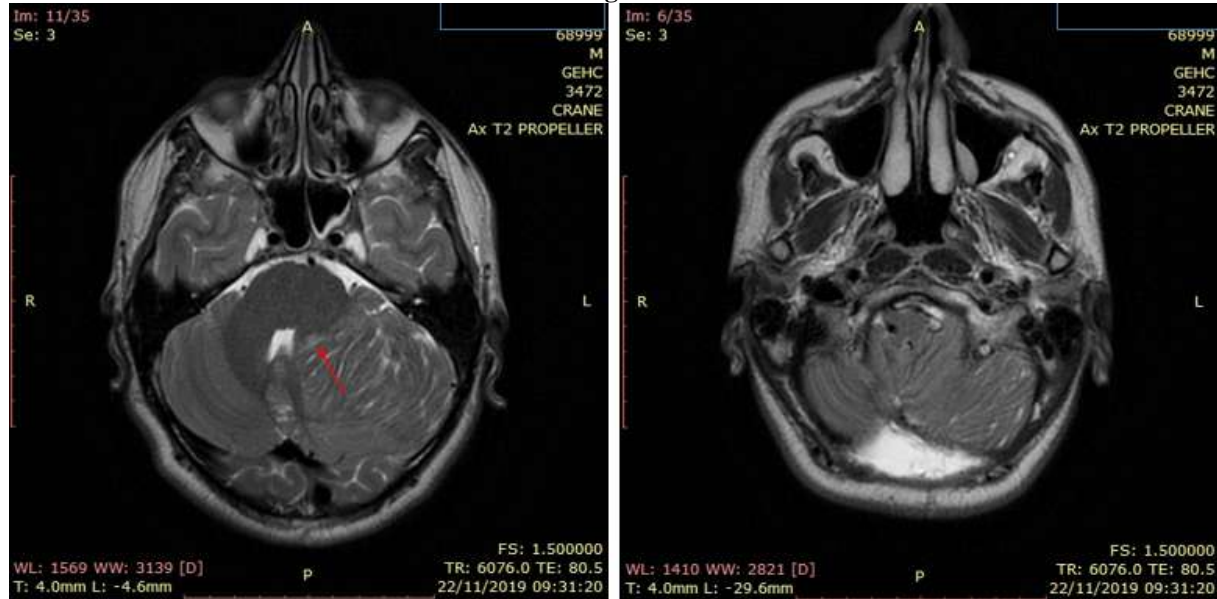


Figure 1:- T2-weighted axial magnetic resonance (MR) images of the cerebellum. A mass lesion is seen occupying the left cerebellar hemisphere. Within the abnormal area, there is a distinct gyral pattern corresponding to the enlarged cerebellar folia. These images also show the mass effect on the brainstem and the fourth ventricle (arrow).



Figure 2:- T1 fatsat after gadolinium injection on axial and coronal images there is no enhancement of the mass because of the absence of brain-blood barrier changes, nor extracellular oedema. We can also distinguish the mass effect on brainstem and the tonsillar engagement (arrow)

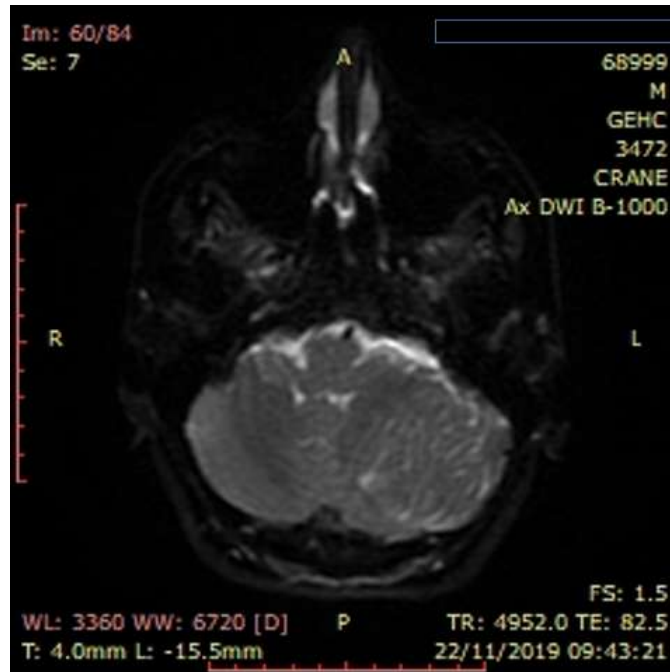


Figure 3:- In the DWI sequences, the lesion appears isointense because the level of free water does not change.

Discussion:-

Gangliocytomas are rare and benign neuronal tumors. They are known on the upper level because of their peripheral cortical and temporal location, reflecting their epileptogenic character (2).

In FCP, their characteristics are less known due to their high clinical latency: in fact, unlike supratentorial lesions, these tumors are only expressed by their mass effect, which is often late due to their weak progression potential.

Lhermitte-Duclos disease is a special entity also called dysplastic ganglioneuroma of the cerebellum: as the name suggests, the origin of this lesion is uncertain. It should be noted that Lhermitte-Duclos disease can be part of Cowden syndrome or multiple hamartoma syndrome (2).

However, its appearance in imaging is often characteristic, allowing a diagnosis of near certainty.

In both cases, these are tumors of children and young adults for gangliocytomas and young adults for Lhermitte-Duclos disease (between the second and fourth decade).

Discovered in young adults with cerebellar symptoms and sometimes intracranial hypertension linked to hydrocephalus.

Certain dysplastic gangliocytomas of the cerebellum fall within the framework of Cowden's disease, which is an autosomal dominant phacomatosis of transmission and which associates cutaneous and mucous hamartomas and various neoplastic lesions (3).

In imaging, the difference is obvious, since Gangliocytoma is rather hemispherical, cerebellar, but may also be bulbar(4).

Cerebellum dysplastic gangliocytoma results in enlargement and disruption of cerebellum folia. In CT the enlarged folia is associated with a small increase in densities and is responsible for a mass effect.

on the fourth ventricle. In MRI, the appearance is variable: they can be cystic and sometimes calcified. Their signal is not specific. After injection, the enhancement is not constant (5).

None of these signs are specific. However, it should be noted that the mass effect and the edema caused by the tumor are always very moderate, whatever its size.

In MRI, Lhermitte-Duclos disease appearance is more characteristic the lesion is hypointense in T1 and slightly hyperintense in T2, with the persistence of the morphology of the folia, thus giving a "tabby" appearance; contrast enhancement is only rarely noted, which appears to be linked to dilated veins located between the folia, as seems to be demonstrated by magnetic susceptibility sequences. In diffusion, the signal is slightly hyperintense due to a T2 effect. Generally, no enhancement after contrast injection. (4,5,6)

MRI sometimes demonstrates obstructive hydrocephalus, exceptionally syringomyelia linked to chronic obstruction of the foramen magnum, linked to the engagement of the cerebellar tonsils (7).

Areas with hypermetabolism and increased blood cerebral volume are sometimes identified within the lesion; spectroscopy notes a decrease in NAA and choline, with the possibility of a lactate peak (8).

It has been suggested that neurologic imaging is sufficient for diagnosis of this condition. Although a case of medulloblastoma mimicking dysplastic gangliocytoma at neurologic imaging has been reported (22). Imaging remains reliable in the diagnosis of this condition, obviating biopsy in asymptomatic patients. MR imaging is also invaluable for preoperative planning and as an aid to determine the extent of resection. In asymptomatic patients, MR imaging helps in the assessment of the lesion extent, mass effect, presence of hydrocephalus, and need to perform surgery. Regular follow-up is required.

Conclusion:-

In summary, the combination of a hypodense, non enhancing mass in the posterior fossa with 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, and prominent folia may be more specific. The margins of this process are very difficult to identify at the surgery and are better delineated by MR, which can thus define the extent of lesion resection.

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