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Original Research Paper

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AN UNUSUAL CASE STUDY ON LHERMITTE DUCLOS DISEASE WITH INTRA-LESIONAL CEREBELLAR METASTASIS

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ABSTRACT Lhermitte–Duclos disease (LDD) is a rare tumor. It is often associated with Cowden syndrome (CS); however, it can occur by itself. Mainly diagnosed by MRI, when it comes to preoperative, the T2-weightened MRI demonstrates the classical "tiger-striped" pattern. The treatment for LDD consists of surgical decompression or excision. We present here a rare case of Lhermitte–Duclos disease in a patient of carcinoma lung evaluated for metastasis with incidental detection of Lhermitte–Duclos disease with intra-lesional cerebellar as well as cerebral metastasis.

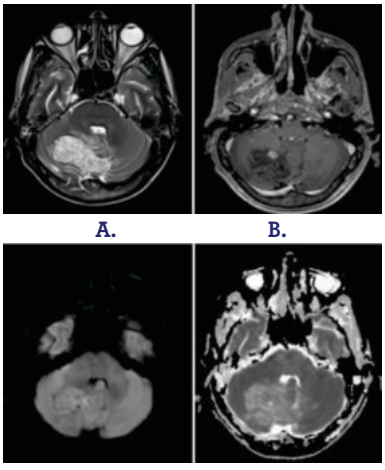
KEYWORDS : Lhermitte Duclos disease, dysplastic gangliocytoma, tiger striping sign, cerebellum.

INTRODUCTION

Lhermitte-Duclos disease is a disorder due to a rare benign lesion of uncertain pathogenesis. Lhermitte Duclos disease is a dysplastic gangliocytoma, characterized by distortion of the normal cerebellar laminar cytoarchitecture. The incidence is approximately 5 per million per year.1 It is an extremely rare benign tumor of specific cerebellar localization, with poorly understood etiopathogenesis. It was first described in 1920 by two French people, Lhermitte and Duclos. There is recognition of a higher female preponderance. Magnetic resonance imaging can identify its characteristic image due to a distortion of the laminar architecture of the cerebellum and impregnating a tiger striped and flaky appearance to the lesion area of the cerebellum. The lesion appears as a non-enhancing mass and has a striated pattern.

Lhermitte-Duclos can occur sporadically or can be associated with Cowden syndrome (CS), a genetic disorder caused by a mutation in the tumor suppressor gene PTEN. When LD disease occurs with CS, it is referred to as Lhermitte-Duclos–Cowden syndrome, a condition marked by multiple hamartomas, benign tumors, and increased cancer risk. The treatment is a simple monitoring if the patient is asymptomatic or decompression excision surgery when the patient is hyper symptomatic.

There is extremely rare to have cerebellar metastasis in this dysplastic tissue, this case presented with carcinoma lung with brain metastasis. The metastatic lesion within the dysplastic cerebellar tissue.



A. B.

C. D.

Figure 1: Right cerebellar hemisphere. (A) Axial T2-weighted image reveals a heterogeneously high-signal mass whose laminated architecture is characteristic of Lhermitte-Duclos disease. (B) A contrast-enhanced axial T1-weighted image shows limited patchy enhancement within the lesion. A heterogeneously enhancing small lesion within the dysplastic cerebellar tissue – intra-lesional cerebellar metastasis. (C) DWI shows increased signal. (D) High signal on ADC.

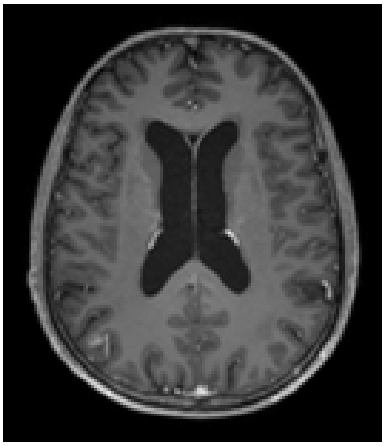


Figure 2: A heterogeneously enhancing intra-axial lesion in right parietal lobe (gray white matter junction)- cerebral metastasis.

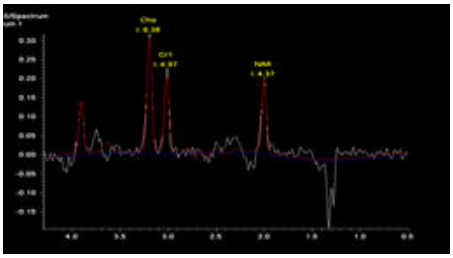


Figure 3: MRS of cerebellar metastatic lesion shows choline peak with increase choline/creatine ratio (1.6).

Case Study

A 31 year old male, known case of adenocarcinoma lung underwent metastatic evaluation. Neurological examination revealed no focal, sensory or motor deficit and no cerebellar dysfunction.

Contrast enhanced abdomen + thorax revealed malignant

lesion in right lung with metastatic meditational lymphadenopathy with pulmonary, liver and skeletal metastasis.

MRI brain done revealed few (4-5) relatively well defined heterogeneously enhancing intra-axial lesions seen in bilateral cerebral hemisphere (gray white matter junction) and right superior cerebellar hemisphere. They appeared iso-hypointense on T1 with T1 hyperintense rim, hypointense on T2/FLAIR, did not show diffusion restriction. Most of them show hemorrhagic component within. On multivoxel MRS there is choline peak with increase in choline/creatine ratio (1.6). On perfusion, there was reduction in rCBV. Few (3-4) peripherally enhancing, T2 hypointense lesions were seen in the right frontal and parietal bones. On CT correlation, the lesions appeared sclerotic.

In a case of Carcinoma Lung these features were suggestive of cerebral, cerebellar and calvarial metastasis.

A well-defined intra-axial altered signal intensity right cerebellar lesion with hypointensity on T1WI, hyperintensity on T2WI with alternate high and normal signal intensity giving it a tigroid appearance. In DWI it appeared bright with high values on ADC (T2 shine through). No post contrast enhancement was seen. No calcification/hemorrhage. Causing mass effect in the form of effacement of 4th ventricle and compression over right middle cerebellar peduncle & right hemi-pons. These features were consistent with benign lesion- dysplastic gangliocytoma of cerebellum.

Few (2) of the above mentioned metastatic lesions were seen within the cerebellar dysplastic parenchyma.

The patient subsequently underwent FDG-PET, which revealed FDG uptake in the above mentioned cerebral and cerebellar metastatic lesions. However, the dysplastic cerebellar tissue did not show FDG uptake.

CONCLUSIONS

In a patient with metastatic lung adenocarcinoma, multimodal imaging differentiated benign dysplastic gangliocytoma of the cerebellum (Lhermitte-Duclos disease, LDD) from intracranial metastases. Key distinguishing features included a cerebellar lesion with characteristic "tigroid" (striated) T2/FLAIR signal, no contrast enhancement, no diffusion restriction, low rCBV on perfusion, and absence of FDG uptake — in contrast to multiple enhancing, hemorrhagic, high FDG uptake metastases in cerebrum, cerebellum and calvarium. Recognition of these imaging hallmarks prevented misclassification of the cerebellar lesion as metastasis and underscores the importance of integrating anatomical, functional and metabolic imaging in neuro-oncologic evaluation.

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