LHERMITTE-DUCLOS DISEASE ASSOCIATED WITH MENINGIOMA – A RARE CASE

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ABSTRACT

Lhermitte-Duclos disease (LDD), also known as dysplastic gangliocytoma of the cerebellum is a benign disease, extremely rare, characterized by loss of normal cerebellar cortical architecture and focal thickening of the folia. Lhermitte-Duclos disease as a major CNS manifestation of Cowden disease is a rare condition. The association between Lhermitte-Duclos disease and meningioma is even more rare, only few cases having been documented in the literature. We present a very rare case of a 53-year-old woman who was admitted in our department for recurrent episodes of headache and epilepsy. Magnetic resonance imaging revealed the presence of a well-defined lesion with an abnormal laminated pattern of cortical architecture involving most of the right cerebellar hemisphere, compressing the fourth ventricle and also a meningioma. Complete removal of the meningioma and partial removal of the cerebellar lesion was performed and histopathological exam confirmed Lhermitte-Duclos disease.

Keywords: Lhermitte Duclos disease, meningioma

INTRODUCTION

Lhermitte-Duclos disease (LDD), also known as dysplastic gangliocytoma of the cerebellum is a benign disease, extremely rare, characterized by loss of normal cerebellar cortical architecture and focal thickening of the folia. We report a rare case of Lhermitte- Duclos disease associated with a meningioma.

CASE PRESENTATION

We present this rare association between Lhermitte-Duclos disease and cerebral meningioma in a patient operated on in our neurosurgery department. A 53-year-old right handed woman was admitted in our department for recurrent episodes of headache, focal seizures and generalized tonic-clonic seizures. She presented headaches for about 8 years and for about 2 she had seizures. Two months before admission, she presented weakness and paresthesia in the right arm and leg. Neurological examination revealed: ataxia, dysarthria, right hemiparesis, right facial nerve palsy and nystagmus at lateral gaze.

CT-scan showed a hypo-hyperdense mass in the right cerebellar hemisphere without contrast enhancement causing mild supratentorial hydrocephalus. It also revealed a large left frontal mass with intense and homogeneous enhancement and midline shift (Fig. 1,2).

MRI exam revealed on transaxial T2-weighted sequences a hyperintense, well-defined large mass on the left frontal convexity with broad implantation on the dura. In the posterior fossa, the MRI showed a lesion in the right cerebellar hemisphere with compression and torsion of the brainstem and of the fourth ventricle. The cerebellar folia can easily be recognized within the mass and appeared thickened (Fig. 3,4).

The supratentorial mass is hypointense on transaxial T1- weighted sequences and the infratentorial mass showed the thickened right cerebellar folia in the lesion. Transaxial, coronal and sagital gadolini-

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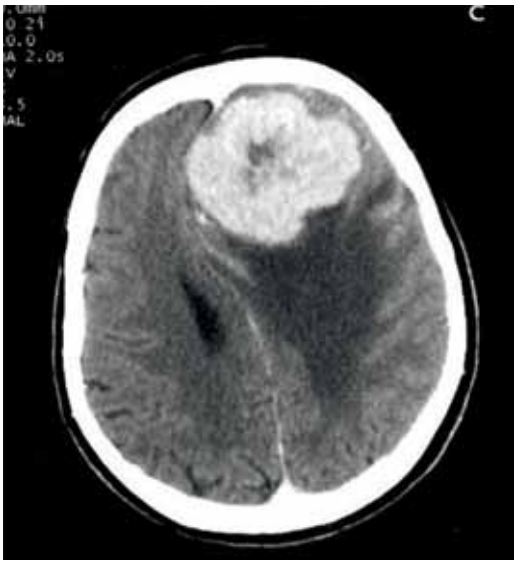
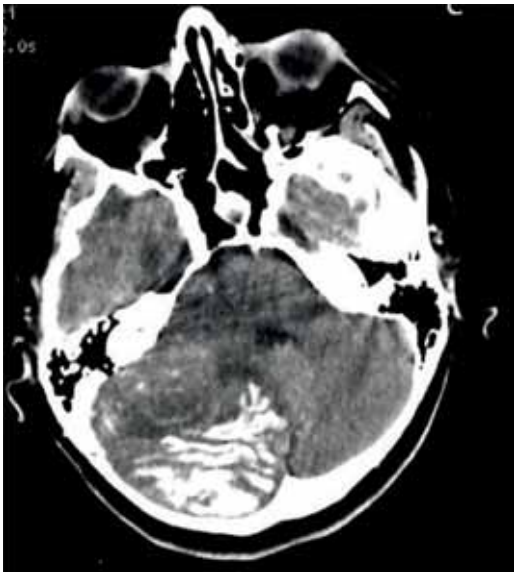


FIGURE 1. *CT scan – left frontal tumour*



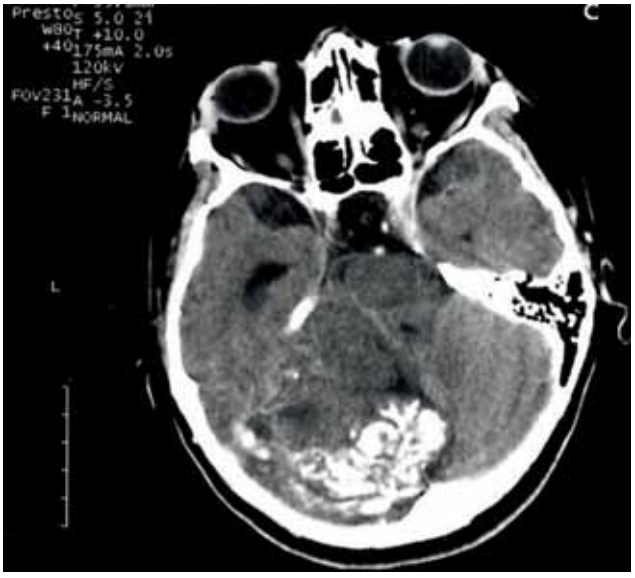


FIGURE 2. *CT scan shows a hypo-hyperdense mass in the right cerebellar hemisphere with „tiger-striped" appearance*

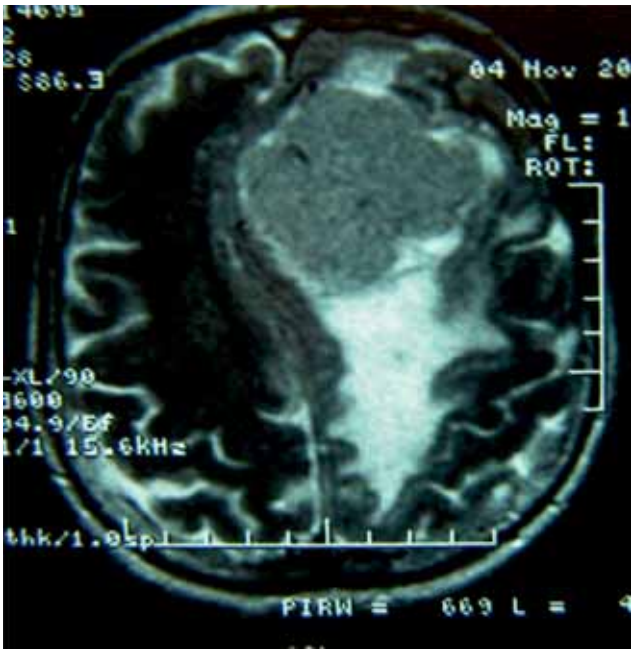


FIGURE 3. *MRI – Axial T2 weighted sequence shows a large left frontal tumour*

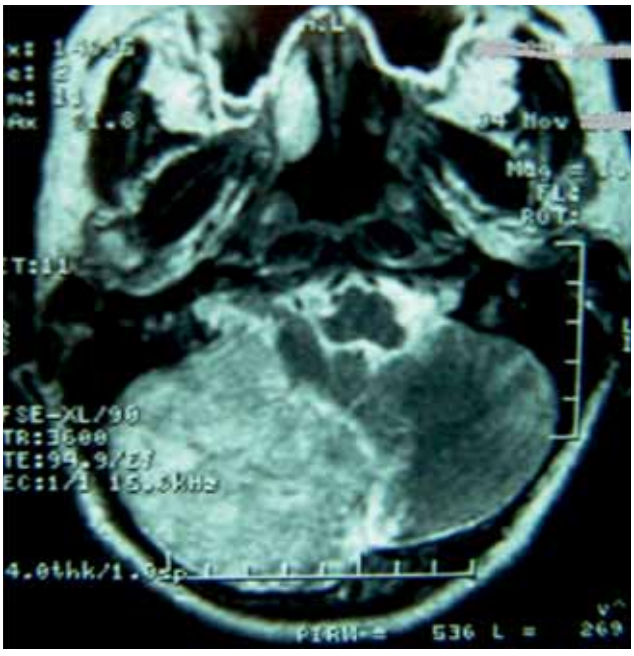


FIGURE 4. *Axial T2 weighted sequences shows a predominantly hyperintense lesion in the right cerebellar hemisphere with striated appearance*

um-enhanced sequences showed intense and homogenous enhancement of the left frontal lesion. Broad implantation on the convexity is seen as a circumferential dural tail. Absence of contrast enhancement of the right cerebellar tumor (Fig. 5, 6).

The conclusion of the MRI examination was that images are suggestive for a left frontal meningioma and for a Lhermitte-Duclos disease.

Surgical findings. The patient was operated on in 2 steps. First, a left frontal craniotomy was performed and a 5 cm diameter left frontal tumor was

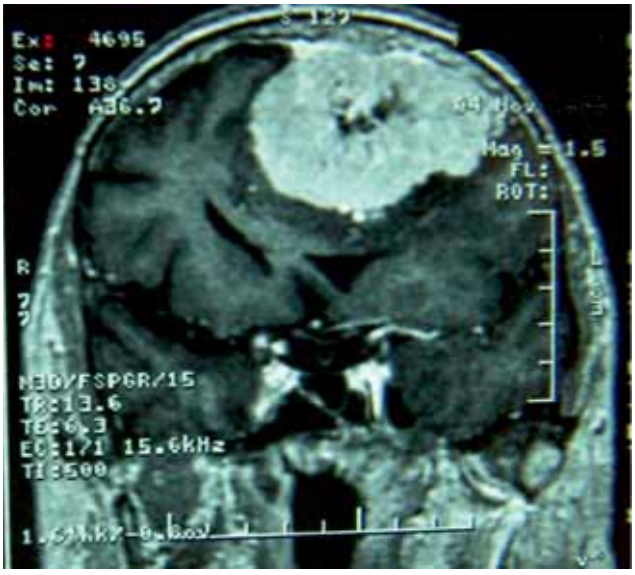


FIGURE 5. *T1-weighted post-gadolinium coronal MRI shows a large dural-based mass over the left frontal convexity with diffuse enhancement.*

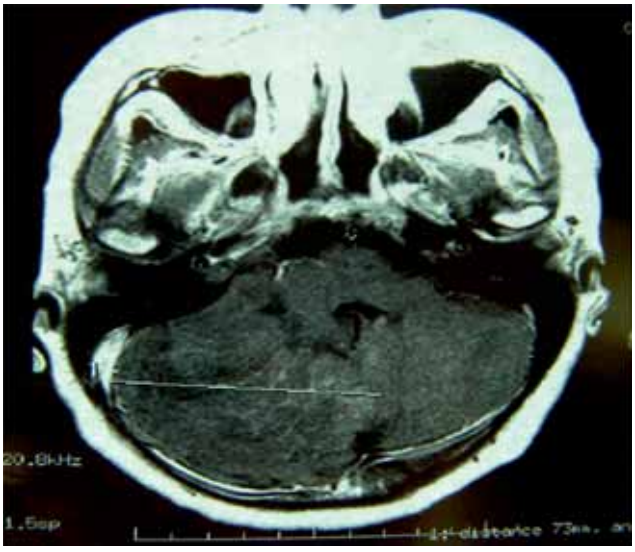


FIGURE 6. *Axial T1 weighted post-gadolinium MRI shows the a bsence of contrast enhancement of the right cerebellar tumor*

found with broad implantation on the dura mater. Anatomopathology was consistent with meningioma.

After 6 days a posterior fossa craniectomy was performed and the right cerebellar hemisphere appeard markedly enlarged, with a thickening of the cerebellar folia, but without evidence of a mass tumour. A subtotal removal of the lesion was done. The frozen section biopsy revealed the absence of tumor cells and no further resection was performed. It was decided to do a large decompression of the posterior fossa with duroplasty (Fig. 8).

The histopathologic exam confirmed the diagnosis of cerebellar dysplastic gangliocytoma or Lhermitte-Duclos disease. The patient was dis-

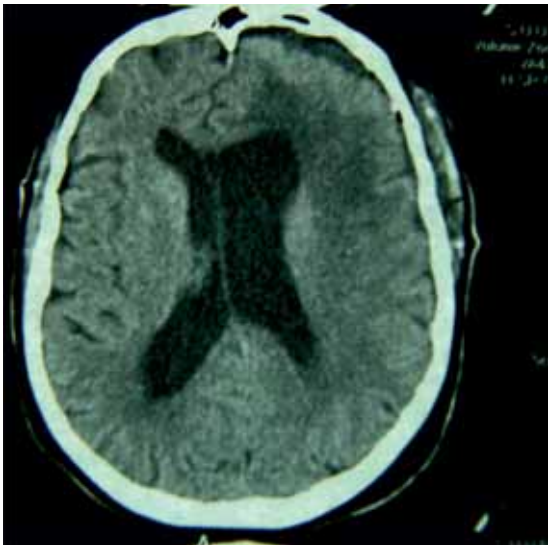


FIGURE 7. *Postop CT scan shows complete removal of the lesion*

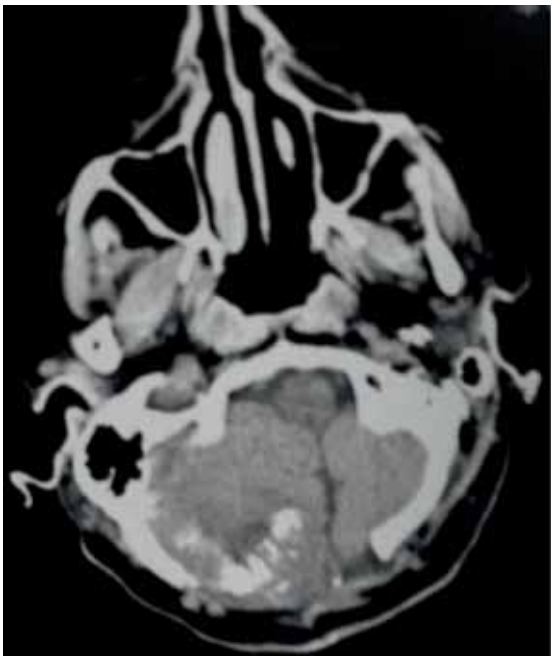


FIGURE 8. *Postop CT scan shows subtotal removal of the cerebellar lesion*

charged without right hemiparesis, no facial nerve palsy and only with a mild ataxia.

DISCUSSION

Lhermitte and Duclos first described the cerebellar dysplastic gangliocytoma in 1920. They reported on a 36-year-old man who presented occipital headaches and diminished hearing on the left side, that was progressive over 10 months. Before the presentation he suffered repeated episodes of paroxysmal vertigo with recurrent falls, memory deficits and gait ataxia. His condition deteriorated and he died. The first surgery attempted for LDD

was performed in 1930 by Bielschowsky and Simons. Approximately 240 patients with LDD have now been reported in the literature with another case added in this report (1).

Lhermitte- Duclos disease can be familial or sporadic. PTEN (phosphatase and tensin homolog)/ AKT (protein kinase B) pathway, an important regulator of cell growth, is considered to be involved. The dysplastic cerebellar gangliocytoma is a hamartomatous overgrowth disorder of the cerebellar cortex. This disorder has been linked to an autosomal dominantly inherited condition characterized by multiple hamartomas, an increased incidence of breast, uterine, and thyroid cancer and germline mutations in the PTEN (phosphatase and tensin homolog) gene at locus *10q23.2*, called Cowden syndrome (2,3). The prognosis of patients with LDD has improved markedly with advances in neuroimaging, especially MRI. Clinical manifestations are related to posterior fossa lesions with secondary obstructive hydrocephalus. On computed tomography (CT), the cerebellar tumour is well defined, circumscribed, isodense or hypodense. Calcification is an uncommon finding. MR imaging is the modality of choice, it is typically nonenhancing. A mass in the posterior fossa with unilateral hemispheric expansion, hypointense on T1-weighted images and hyperintense on T2-weighted images with a laminar pattern of alternating high and low signal, 'tiger-striped' appearance, is considered suggestive for the diagnosis. Despite the benign nature, surgical excision is the treatment of choice. In some cases a wide excision is necessary due to the risk of recurrence. In this case we decided for subtotal removal, the reason for this surgical attitude was that in this case, a clear border between tumour and surrounding healthy cerebellum tissue is missing, so there is a risk for incomplete removal of the tumour. The surgical removal of these lesions in an open MRI unit would be a good indication.

CONCLUSIONS

Lhermitte-Duclos disease is a very rare disease. The association between Lhermitte- Duclos disease and meningioma is even more rare, as far as we know, only few cases have been documented in the literature. MRI is a valuable diagnosis tool in such cases.

REFERENCES

1. **Shenandoah R., Cohen A.** Cowden Disease and Lhermitte-Duclos Disease: An Update, *Neurosurg Focus.* 2006;20(1), http://www.medscape.com/viewarticle/522324_4
2. **Norden A., Chheda M, Wen P. et al.** Uncommon brain tumors, http://www.uptodate.com./contents/uncommon-brain-tumors, 2014
3. **Abel T.W., Baker S.J., Fraser M.M. et al.** Lhermitte-Duclos disease: a report of 31 cases with immunohistochemical analysis of the PTEN/AKT/mTOR pathway, *J Neuropathol Exp Neurol.* 2005; 64(4):341
4. **Moenninghoff C., Kraff O., et al.** Assessing a Dysplastic Cerebellar Gangliocytoma (Lhermitte-Duclos Disease) with 7T MR Imaging Korean *J Radiol.* 2010 Mar-Apr; 11(2): 244-248