

LHERMITTE-DUCLOS DISEASE

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Key-word: Lhermitte-Duclos disease

Background: A 60-year-old patient was admitted to the neurology department because of two attacks of partial seizures in 2 weeks' time. Medical history revealed bilateral breast cancer, diagnosed in 1986 and 1999, respectively, hyperthyroidism and endometrial cancer. CT scan of the brain was performed, and in view of the findings, followed by MRI.

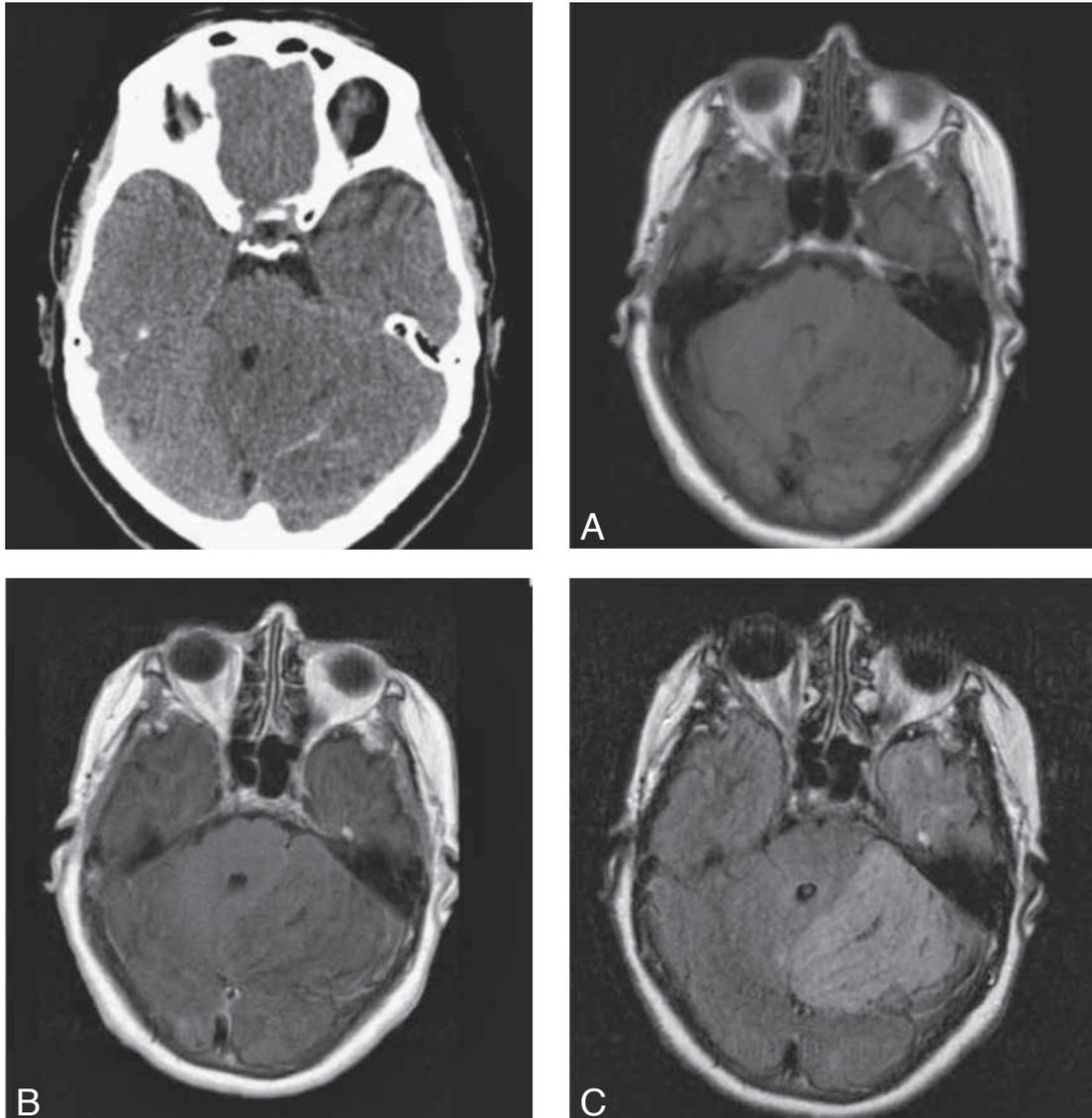


Fig.

1	2A
2B	2C

Work-up

On unenhanced CT scan of the cerebrum (Fig. 1) a mass with mixed density and containing calcifications in the left cerebellar hemisphere is seen. There is mass effect with compression on the brainstem and the fourth ventricle.

On MRI of the brain (Fig. 2), axial T1-weighted image (A), a hypo-intense lesion in the posterior fossa, with enlargement of the cerebellar cortical folia, compression on the brainstem and an asymmetrical fourth ventricle are observed. Axial Gd-enhanced T1-weighted images (B) show moderate enhancement of the left cerebellar lesion is seen. Axial T2-weighted images (C) demonstrate a well-circumscribed hyperintense lesion, with a striated pattern (tiger-striped folial pattern) in the left posterior fossa.

Radiological diagnosis

Based on the imaging findings the diagnosis *Lhermitte-Duclos disease* was suggested and the patient was sent to the dermatologist and clinical genetics department. Examination of the skin showed multiple hamartomous lesions. Genetic testing revealed a mutation in the PTEN gene. Hence, Cowden's syndrome was proven. The tumor was not resected because of the disappearance of clinical symptoms after the patient had started on levetiracetam, an antiepileptic drug.

Discussion

Lhermitte-Duclos is a hamartomous overgrowth of cerebellar ganglion cells. Patients usually present with signs of cerebellar dysfunction or signs of increased intracranial pressure. Lhermitte-Duclos is associated with Cowden's syndrome. It has been suggested that Lhermitte-Duclos is the neurological manifestation of Cowden's syndrome. Cowden's syndrome is an autosomal dominant disorder char-

acterized by hamartomous neoplasms of the skin, mucosa, gastro-intestinal tract, bones, central nervous system, eyes and genitourinary tract. Cowden's syndrome is caused by a mutation in the PTEN tumor suppressor gene on chromosome 10 (10q23). This mutation causes loss of its protein function and results in overproliferation of cells, resulting in hamartomous growths. There is a high risk of thyroid, endometrial and breast cancer.

The typical striated or tiger-striped folial pattern is a characteristic finding of Lhermitte-Duclos disease (LDD) also named dysplastic gangliocytoma.

MRI has proven to be the best imaging modality. It reveals a cerebellar mass with a typical striated folial pattern that consists of alternating bands on both T1- and T2-weighted images. The bands are iso- and hypo-intense on T1-weighted images and hyper- and iso-intense on T2-weighted images. Calcifications are an uncommon finding, but it has been reported. MRI enables physicians to make the diagnosis LDD without histopathological confirmation.

The treatment for LDD consists of surgical decompression or excision.

Bibliography

1. Boonpipattanapong T., Phuenpathom N., Mitarnun W.: Cowden's syndrome with Lhermitte-Duclos disease. *Br J Neurosurg*, 2005, 19: 361-365.
2. Moeninghoff C., Kraff O., Schlamann M., et al.: Assessing a dysplastic cerebellar gangliocytoma (Lhermitte-Duclos Disease) with 7T MR imaging. *Korean J Radiol*, 2010, 11: 244-248.
3. Nowak D.A., Trost H.A.: Lhermitte-Duclos disease (dysplastic cerebellar gangliocytoma): a malformation, hamartoma or neoplasm? *Acta Neurol Scand*, 2002, 105: 137-145.
4. Shinagare A.B., Patil N., Sorte S.Z.: Case 144: Dysplastic cerebellar gangliocytoma (Lhermitte-Duclos Disease). *Radiology*, 2009, 251: 298-303.