If You See It Once, You Do Not Forget: Lhermitte-Duclos Disease

Bir Kez Görseniz Unutmazsınız: Lhermitte-Duclos Hastalığı

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Turk Norol Derg 2011;17:211-213

ÖZET

Lhermitte-Duclos hastalığı tipik manyetik rezonans görüntüleme bulguları ile karakterize patogenezi bilinmeyen nadir bir hastalıktır. Cowden sendromu ile ilişkili olabilir. Bu nedenle diğer eşlik eden tümörleri dışlamak için preoperatif tanısı önemlidir. Bu makalede Lhermitte-Duclos hastalığı olan iki olgumuzu ve tipik manyetik rezonans görüntüleme sinyal özelliklerini sunuyoruz.

Anahtar Kelimeler: Hamartoma sendromu, multipl, manyetik rezonans görüntüleme.

ABSTRACT

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Lhermitte-Duclos disease is a rare disorder of unknown pathogenesis, characterized by typical magnetic resonance imaging findings. Lhermitte-Duclos disease can be associated with Cowden's syndrome; thus, preoperative diagnosis is important for excluding the other associated tumors. Herein, we present two cases of Lhermitte-Duclos disease and describe the typical magnetic resonance imaging signal characteristics.

Key Words: Hamartoma syndrome, multiple, magnetic resonance imaging.

INTRODUCTION

Lhermitte-Duclos disease (LDD) is a rare benign lesion of the cerebellum that is characterized by a cerebellar mass composed of enlarged cerebellar folia containing abnormal ganglion cells (1). It is unclear whether this lesion is in fact a neoplasm, hamartoma or dysplasia (2). It is characterized as a World Health Organization (WHO) grade I tumor (3). LDD usually presents with symptoms secondary to intracranial pressure and hydrocephalus. It is most common in the third and fourth decades of life, and there is no sex predilection (4). LDD can be associated with Cowden's syndrome. We present two cases of LDD and describe the typical magnetic resonance imaging (MRI) signal characteristics.

CASE

Our first case was a 14-year-old female, admitted to our hospital with a history of headache and vertigo for two months. Nausea and vomiting were added to the complaints in the last days before presentation. Physical examination was normal except for horizontal nystagmus and blurriness of optic disk margins. MRI revealed a mass in the left cerebellar hemisphere with low signal on T1-weighted images and high signal on T2-weighted images. Minimal folial contrast enhancement was observed. The mass had parallel stripes, which was characteristic of the disease, and led to compression of the pons and fourth ventricle and dilatation of the third and lateral ventricle. No edema was found around the lesion (Figure 1,2).

The second case was a 49-year-old male admitted to the hospital with a history of headache and dizziness lasting for three months. Physical examination and neurologic examination were within normal limits. MRI findings revealed a mass lesion involving the entire right cerebellar hemisphere with minimal compression of the fourth ventricle. The lesion was hyperintense on T2-weighted images and hypointense on T1-weighted images with striated pattern and isointense bands within it. No enhancement was noted after injection of contrast material (Figure 3,4).

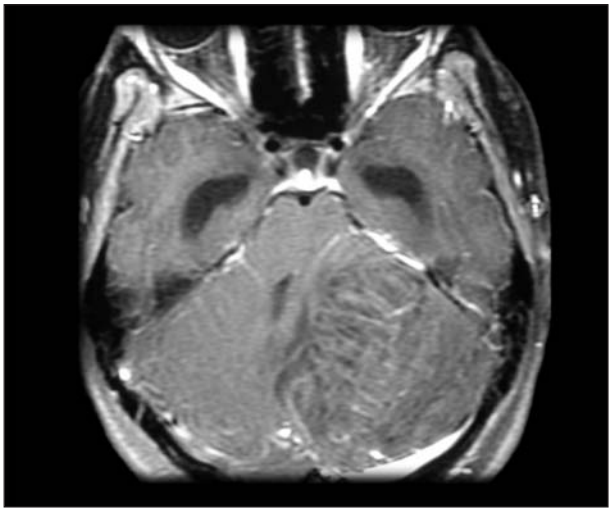

Figure 2. Enhanced MRG shows that there is minimal folial contrast enhancement.

Both cases were examined for Cowden's syndrome. Their family histories were negative for Cowden's syndrome. Their breast-thyroid ultrasonography examinations and colonoscopy were normal. No dermatologic lesions were found.

Total excision of the lesions was performed, and postoperative pathological examinations confirmed the diagnosis as dysplastic gangliocytoma of the cerebellum.

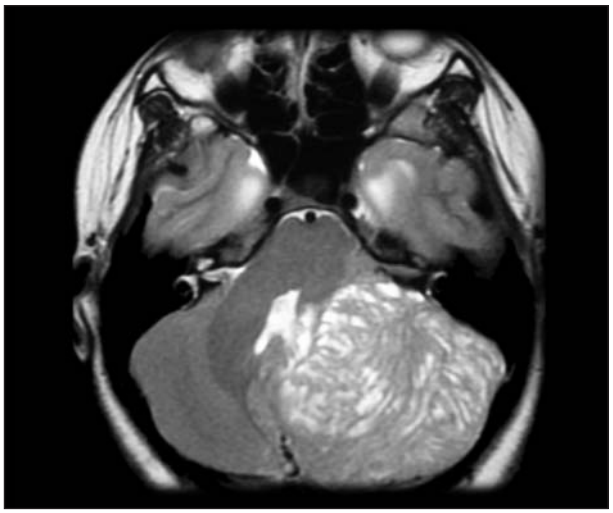

Figure 1. T2-weighted images of the lesion demonstrate the obvious mass effect and high-intensity striations within the mass. Note that the mass does not destroy the folial pattern.

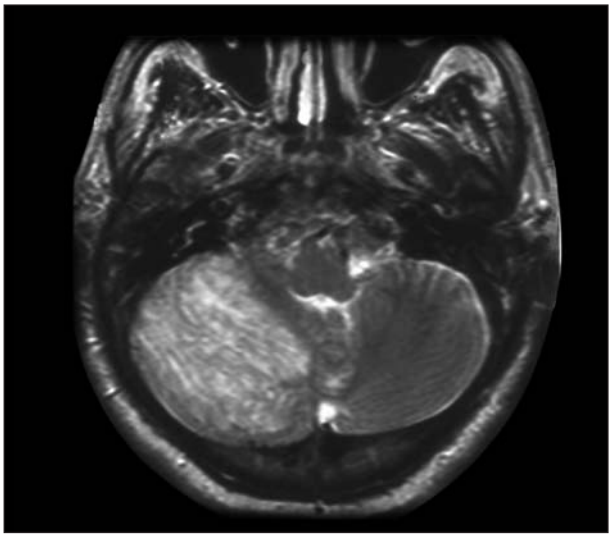

Figure 3. T2-weighted image displays a serpentine and striated high-intensity pattern with mass effect.

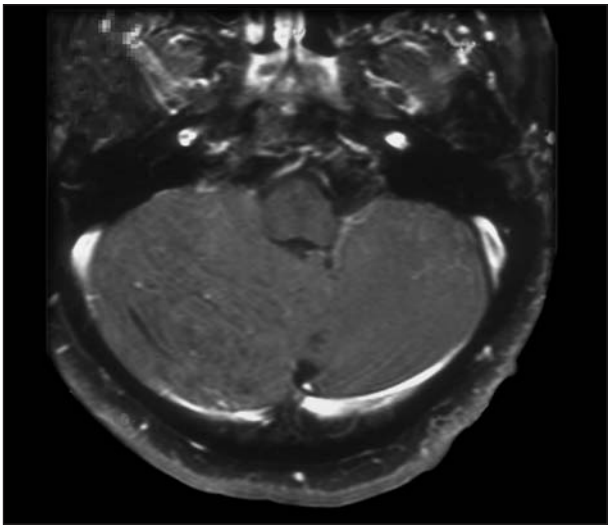

Figure 4. Enhanced MRG shows that there is no significant contrast enhancement.

DISCUSSION

MRI findings of LDD are characteristic, showing parallel linear stripes within the mass without significant contrast enhancement. No edema is found around the lesions. The MRI findings of LDD are distinctive and easily differentiated from other tumors of the cerebellum, because most other cerebellar masses destroy the folial pattern and show significant enhancement after contrast medium administration. Most of the patients can be diagnosed preoperatively with MRI (4,5). We can see a similar striated appearance in tuberous sclerosis, which is the true dysplasia of the cerebellum; however, tuberous sclerosis appears in younger patients. Cortical tubers, subependymal nodules and white matter changes are associated with tuberous sclerosis (6).

The disease is also important because LDD is associated with Cowden's syndrome. Cowden's syndrome is a rare autosomal dominant familial cancer syndrome with multiple skin lesions and an increased predisposition to breast cancer and thyroid tumors (5). In patients with LDD, systemic screening should be performed to evaluate for any other accompanying disorder.

REFERENCES

1. *Wu CH, Chai JW, Lee CH, Chen WH, Lee T, Chen CC. Assessment with magnetic resonance imaging and spectroscopy in Lhermitte-Duclos disease. J Chin Med Assoc 2006;69:338-42.*
2. *Grosmann RI, Yousem DM. Neoplasm of the Brain. Neuroradiology. 2nd ed. USA: Elsevier, 2003:162-3.*
3. *Yağci-Küpeli B, Oguz KK, Bilen MA, Yalçin B, Akalan N, Büyükpamukçu M. An unusual cause of posterior fossa mass: Lhermitte-Duclos disease. J Neurol Sci 2010;290:138-41.*
4. *Haris AA, Chandra SR, Peethambaran B. Lhermitte-Duclos disease. Neurol India 2008;56:495-6.*
5. *Xu L, Gao PY, Lin Y, Tian TD, Lei J, Ma L. Magnetic resonance imaging findings in Lhermitte-Duclos disease: reports of three cases. Chin Med J 2005;118:1933-6.*
6. *Shinagare AB, Patil NK, Sorte SZ. Case 144: dysplastic cerebellar gangliocytoma (Lhermitte-Duclos disease). Radiology 2009;251:298-303.*

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geliş tarihi/received 20/02/2011
kabul ediliş tarihi/accepted for publication 13/04/2011