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

Bir Kez Görseniz Unutmasınız: Lhermitte-Duclos Hastalığı

ÖZET

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

ABSTRACT
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Elif Aktaş, Kemal Arda, Nazan Ciledağ, Bilgin Kadri Arıbas, Başak Gülpınar

Clinic of Radiology, Dr. Abdurrahman Yurtarslan Ankara Oncology Training and Research Hospital, Ankara, Turkey

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).

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.
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


Yazıma Adresi/Address for Correspondence

Uzm. Dr. Elif Aktaş
Dr. Abdurrahman Yurtarslan
Ankara Onkoloji Eğitim ve Araştırma Hastanesi
Radyoloji Kliniği
Demetevler 06200 Ankara/Türkiye
E-posta: elifaktasmd@gmail.com

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