DOI:10.4274/tnd.2021.22567
Turk J Neurol 2023;29:67-70

Case Report / Olgu Sunumu

Dyke Davidoff Masson Syndrome with Crossed Cerebellar Atrophy: A Case Report and Review of the Literature

Çapraz Serebellar Atrofinin Eşlik Ettiği Dyke Davidoff Masson Sendromu: Olgu Sunumu ve Literatürün Gözden Geçirilmesi

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Abstract

Dyke Davidoff Masson Syndrome (DDMS) is characterized clinically by hemiplegia or hemiparesis, mental retardation, and epilepsy and radiologically by cerebral hemiatrophy, ipsilateral calvarial thickening, and dilation and increased aeration of the ipsilateral sinuses. Atrophy in the cerebral hemisphere may be accompanied by atrophy in the contralateral cerebellum. This condition is defined as crossed cerebellar atrophy (CCA). Although CCA's pathogenesis is unclear, it has been associated with damage to the corticopontocerebellar pathways and neuronal connections between the cerebellum and contralateral cerebral hemisphere. Diffusion tensor imaging and conventional magnetic resonance imaging may help to demonstrate axonal connection pathways and make a diagnosis. Here we present a case of a 39-year-old female patient with clinical and radiological findings of DDMS associated with CCA.

Keywords: Dyke Davidoff Masson Syndrome, crossed cerebellar atrophy, magnetic resonance imaging, diffusion tensor imaging

Öz

Dyke Davidoff Masson Sendromu (DDMS) klinik olarak hemipleji veya hemiparezi, mental retardasyon, epilepsi ve kraniyal görüntülemelerde serebral hemiatrofi, ipsilateral kalvaryal kalınlaşma, ipsilateral sinüslerde genişleme ve havalanma artışı ile karakterize olan bir tablodur. Serebral hemisferdeki atrofiye kontralateral serebellumda atrofi eşlik edebilmektedir. Bu durum çapraz serebellar atrofi (ÇSA) olarak tanımlanmaktadır. ÇSA patogenezi net olmamakla birlikte serebellum ve karşı serebral hemisferi bağlayan kortikopontoserebellar yolaklarda ve nöronal bağlantılarda oluşan hasar ile ilişkilendirilmiştir. Konvensiyonel manyetik rezonans görüntüleme yöntemleri ile birlikte diffüzyon tensor görüntüleme yöntemlerinin birlikte kullanımı; aksonal bağlantı yollarının gösterilmesinde ve tanı koymada yardımcıdır. Burada 39 yaşında bir kadın hastada DDMS'nin ÇSA ile ilişkisini gösteren klinik ve radyolojik görüntüleme bulguları sunuldu.

Anahtar Kelimeler: Dyke Davidoff Masson Sendromu, çapraz serebellar atrofi, manyetik rezonans görüntüleme, difüzyon tensör görüntüleme

Introduction

Dyke Davidoff Masson Syndrome (DDMS), first described by Dyke (1) in 1933, is characterized by cerebral hemiatrophy, thickening of the ipsilateral skull, enlargement, and aeration of the paranasal sinuses, contralateral hemiparesis and hemiplegia, facial asymmetry, epilepsy, and mental retardation. DDMS is associated with congenital and acquired causes (2). Although the etiology is unknown, intrauterine infections, congenital malformations, intracranial hemorrhage and ischemia, vascular malformations, postpartum infections, brain tumors, and trauma should be considered in the differential diagnosis of this syndrome (3). Cerebellar atrophy occurring in the prenatal, natal, and early childhood periods is usually ipsilateral; it is reported that atrophy is contralateral in lesions in later periods (4).

Positron emission tomography is used to show the hypometabolism that occurs in the brain tissue. However, it is not a routine imaging method (5). Diffusion tensor imaging (DTI) is a magnetic resonance imaging (MRI) technique that demonstrates corticopontocerebellar neuronal connection pathways based on the diffusion rate of water molecules (6). DTI findings in DDMS are very rarely reported.

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Received/Geliş Tarihi: 06.04.2021 **Accepted/Kabul Tarihi:** 22.11.2021

Here, a case of DDMS associated with crossed cerebellar atrophy (CCA) in the postpartum period is presented. Due to the contralateral course of DDMS and CCA, acquired causes are considered in this case, etiological factors are investigated, and imaging findings are discussed.

Case Report

A 39-year-old female patient was referred to the clinic by ophthalmology with left homonymous hemianopia. She had a normal birth history from spontaneous vaginal delivery at term with no asphyxia. Her motor development steps were normal until the eighth postpartum month. She had a febrile convulsion in the eighth month, after which weakness developed in the upper and lower extremities. Atrophy and spasticity occurred over time. The patient had no other seizure history after the first febrile seizure. Neurological examination revealed mild mental retardation, left spastic hemiparesis, left hemiatrophy, and increased deep tendon reflexes on the left side. Visual field examination revealed left homonymous hemianopia. Widespread slowing of the electroencephalogram was observed in the right hemisphere. Genetic thrombophilia panel and coagulation parameters were assessed to exclude thromboembolic events, which were found to be within normal limits. Cranial MRI revealed atrophy in the right cerebral hemisphere, ex vacuo dilatation in the ipsilateral lateral ventricle, and atrophy in the contralateral cerebellum (Figure 1A, B). Also, there was thickening in the right hemi-calvarial bony structures (Figure 2A) and enlargement in the ipsilateral frontal and sphenoid sinuses (Figure 2B, C). Cranial magnetic resonance angiography (MRA) significantly decreased the right middle and posterior cerebral artery calibrations (Figure 1C). In DTI, it was observed that the white matter fibers in the corticopontocerebellar pathway on the lesion side were thinning, and encephalomalacia and parenchymal loss caused the tract to begin at a lower level (Figure 3). Considering the patient's clinical and radiological imaging findings, she was diagnosed as having DDMS with CCA.

Discussion

In this case, DDMS with CCA is presented in an adult patient with bilateral homonymous hemianopia. The etiological factors and imaging findings associated with the patient's clinical presentation are discussed.

DDMS is clinically characterized by the symptoms of hemiplegia or hemiparesis, mental retardation, and epilepsy, and radiologically by cerebral hemiatrophy, ipsilateral calvarial thickening, and dilation and increased aeration of the ipsilateral sinuses (7). Brain MRI findings were similar to previously reported DDMS cases in this case. In cases where hemiatrophy occurs in early childhood, the thickening of the bone structures and hypertrophy of the sinuses can be observed as a compensatory factor (4). In the present case, the age of diagnosis was 39 and based on imaging findings, it was suggested that cerebral hemiatrophy had developed in early childhood.

Vascular malformations, thromboembolic events, and vasculitic processes have been reported in cases of DDMS (8,9). In this respect, an MRA and a routine brain MRI are recommended. In this patient's vascular imaging, while the left middle cerebral artery was normal, no flow was observed in the right middle cerebral artery. The genetic thrombophilia panel, coagulation parameters, and vasculitic markers were within normal limits.

DDMS can be associated with congenital and acquired causes (7). Congenital causes include cerebrovascular damage, vascular malformation, intrauterine infections, and congenital malformations. Acquired causes include trauma, infections, tumors, and cerebrovascular disease (4). Crossed cerebellar diaschisis (CCD) represents contralateral hypometabolism secondary to supratentorial injury and may be reversible. CCA has a similar biological process to CCD and is irreversible (4). Ipsilateral or contralateral cerebellar atrophy may accompany DDMS cases. It is known that ipsilateral cerebellar atrophy is found when supratentorial damage occurs in the prenatal and natal periods, and anatomical differences accompany contralateral cerebellar atrophy during development in later cases (3,4). In this case, the prenatal and natal history was evaluated as normal, clinical symptoms started in the eighth month, and cerebellar atrophy was contralateral. Therefore, the development of DDMS was considered to be secondary to acquired causes emerging in the postnatal period.

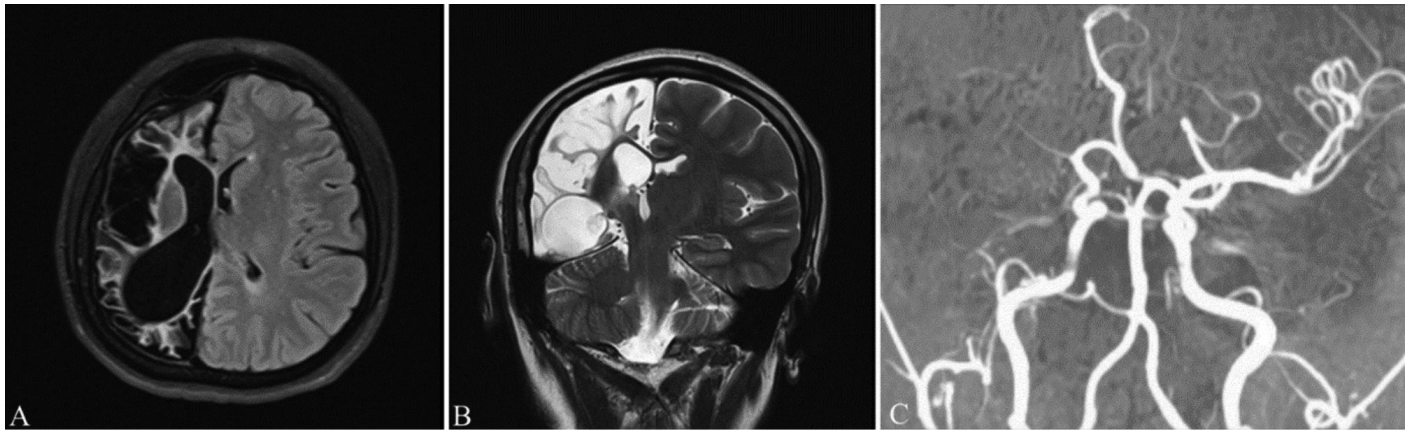

Figure 1. (A) Axial fluid-attenuated inversion recovery magnetic resonance imaging revealing cortical atrophy in the right cerebral hemisphere. (B) Atrophy in the right cerebral hemisphere and contralateral cerebellum in the coronal T2 weighted image. (C) While the left middle cerebral artery is normal, no flow is observed in the right middle cerebral artery

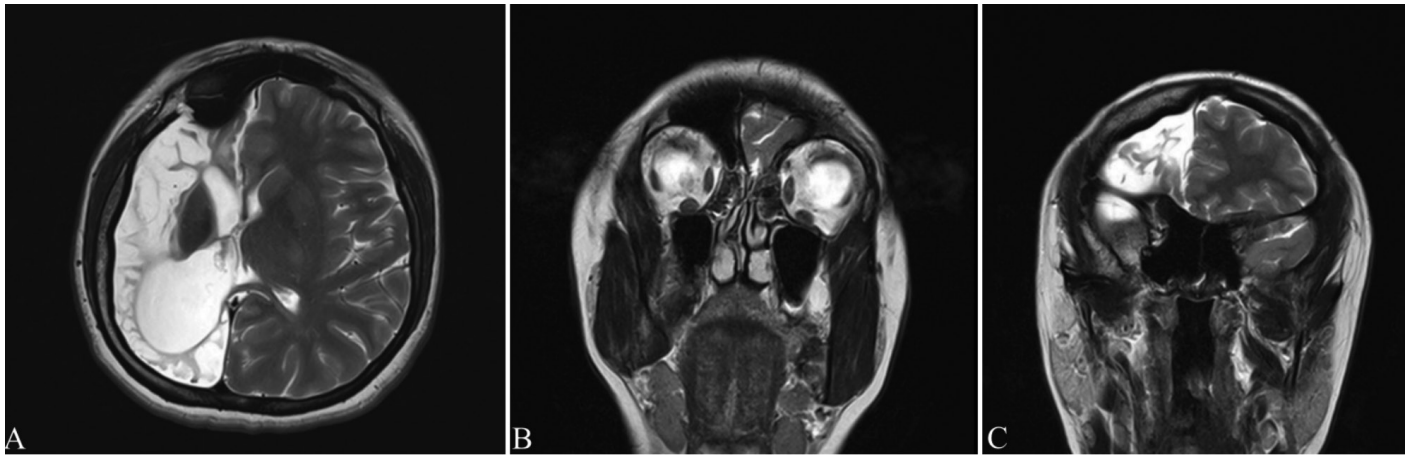

Figure 2. (A) Axial T2 weighted magnetic resonance imaging showing thickening of the right hemi-calvarial bone. (B) Coronal T2 weighted magnetic resonance imaging showing enlargement of the right frontal sinus. (C) Enlargement of the right sphenoid sinus

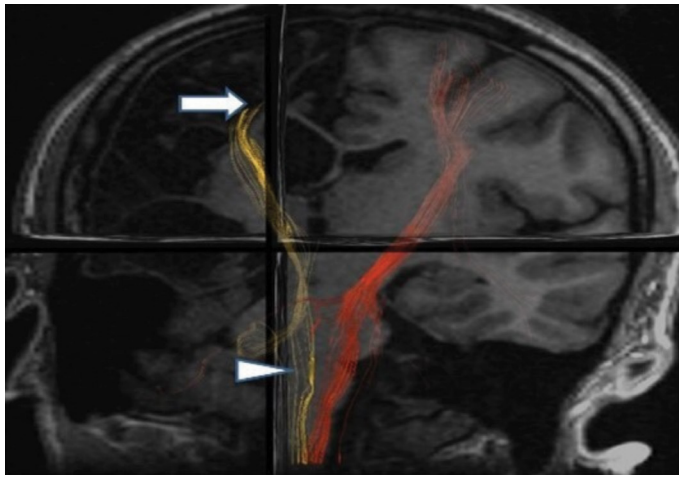

Figure 3. On the coronal view of the 3D fiber-tractography, the white matter fibers of the corticopontocerebellar pathway on the lesion side are thinner (arrowhead), and the origin of the tract is more inferior (arrow) owing to the encephalomalacia and parenchymal loss

CCA is a secondary change in the contralateral cerebellar hemisphere that occurs with damage to supratentorial brain tissues, decreasing blood flow and metabolism in the acute phase and resulting in atrophy in the chronic phase (5). Although its pathophysiology is not clearly understood, the most likely cause has been associated with the corticopontocerebellar tract's involvement connecting the cerebellum and the contralateral cerebral hemisphere and the severing of neuronal functional ligaments (10). DTI is a helpful MRI technique in demonstrating these pathways. The basis of this technique is to determine the structure of the tissue by measuring the *in vivo* diffusion rate and the direction of the water molecules (6). Axon and myelin are involved in the signal transmission in the brain's white matter nerve cells.

Myelin prevents the diffusion of water molecules due to its fat content (11). Isotropic diffusion is seen in tissues (gray matter) where water molecules move equally in all directions without diffusion restriction. In contrast, anisotropic diffusion is observed in tissues with diffusion restriction (white matter). DTI is a guide in many diseases, such as neurodegenerative, demyelinating, and metabolic diseases, epilepsy and brain tumor surgery, and ischemic vascular diseases (6). In this case, corticopontocerebellar fiber loss was observed secondary to the parenchymal loss in the right hemisphere.

DDMS cases are rarely accompanied by CCA. Cerebellar atrophy that develops secondary to cerebral injury may be ipsilateral or contralateral, depending on the injury's development period. In the literature, the damage is generally reported contralaterally when it occurs during the postnatal period in similar cases (4). In cases with cerebral hemiatrophy of unknown etiology, the onset of symptoms, imaging methods, and laboratory markers are integral to diagnosis. Further studies, including DTI, are needed to elucidate the physiopathology of CCA accompanying DDMS.

Ethics

Informed Consent: The patient's written informed consent to this case study was obtained.

Peer-review: Externally and internally peer-reviewed.

Authorship Contributions

Surgical and Medical Practices: M.F.Y., Ş.B., Concept: M.F.Y., Ş.B., İ.Ö., Design: M.F.Y., Ş.B., İ.Ö., Data Collection or Processing: M.F.Y., Ş.B., İ.Ö., Analysis or Interpretation: M.F.Y., Ş.B., İ.Ö., Literature Search: M.F.Y., Ş.B., Writing: M.F.Y., Ş.B.

Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: The authors declared that this study received no financial support.

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