Case Report / Olgu Sunumu
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Chorea gravidarum as a presentation of moyamoya disease

Moyamoya hastalığının bir sunumu olarak kore gravidarum

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ABSTRACT

Neurological findings in moyamoya disease (MMD) are various, mostly related to cerebrovascular events. Movement disorders such as dystonia, chorea or dyskinesia are rarely reported. Chorea gravidarum is a very rare movement disorder in MMD. In this article, we present a 26-year-old female patient whose first symptom was chorea and who was diagnosed as demyelinating disease first, while she was later diagnosed as MMD after magnetic resonance angiography and digital subtraction angiography.

Keywords: Chorea gravidarum, movement disorders, moyamoya disease.

ÖZ

Moyamoya hastalığında (MMH) nörolojik bulgular çeşitli olup çoğunlukla serebrovasküler olaylar ile ilişkilidir. Distoni, kore veya diskinezi gibi hareket bozuklukları nadiren bildirilir. Kore gravidarum MMH'de çok nadir bir hareket bozukluğudur. Bu yazıda, ilk semptomu kore olup önce demyelinizan hastalık tanısı konulurken daha sonra manyetik rezonans görüntüleme ve dijital substraksiyon anjiyografiyi takiben MMH tanısı konulan 26 yaşında bir kadın hasta sunuldu.

Anahtar Sözcükler: Kore gravidarum, hareket bozuklukları, moyamoya hastalığı.

Moyamoya disease (MMD) is a rare disease characterized by bilateral stenosis or occlusion of the arteries around the circle of Willis and angiographic findings of abnormal vascular distribution. Neurological findings are various and associated with cerebrovascular events developed due to vascular pathological changes. The first finding is often transient ischemic attack, cerebral infarct or intracranial hemorrhage and also seizures can be seen rarely.[1,2] Movement disorders such as dystonia, chorea or dyskinesia are rarely reported in patients with MMD.[2-6] In this article, we present a case of MMD observed with chorea gravidarum which is much less seen.

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CASE REPORT

Our patient was admitted to our clinic with the complaints of curling and bending of the left arm and leg when she was 18 years old and six months pregnant. Brain magnetic resonance imaging (MRI) revealed bilateral subcortical hyperintense lesions in the frontal region. The patient was searched in terms of demyelinating disease and atypically located posterior reversible encephalopathy syndrome. Lumbar puncture was performed; the findings of cerebrospinal fluid were normal. Movement disorder was assessed as chorea gravidarum. As treatment, haloperidol was given and chorea was completely recovered with this treatment which was discontinued after one month. The patient did not present to the further follow-ups and we could not hear from her.

After eight years of no follow-up, the patient was admitted to our clinic with the complaint of numbness and weakness in the left arm and leg when she was 26 years old. Neurological examination, hemogram, biochemistry, lipid profile, thyroid function tests were normal at admission. Contrasted brain MRI showed cavum septum pellucidum variation and non-enhancing patchy signal increases, the largest of which measuring 20×12 mm, which involved gray and white matter areas in the bilateral frontal and parietal lobes, prominent on the left side (Figure 1a, b). The patient's MRI findings were assessed in favor of ischemic lesions while brucella, salmonella, syphilis, hepatitis, human immunodeficiency virus tests, tumor markers, vasculitis markers, Epstein-Barr virus immunoglobulin M (IgM) and cytomegalovirus

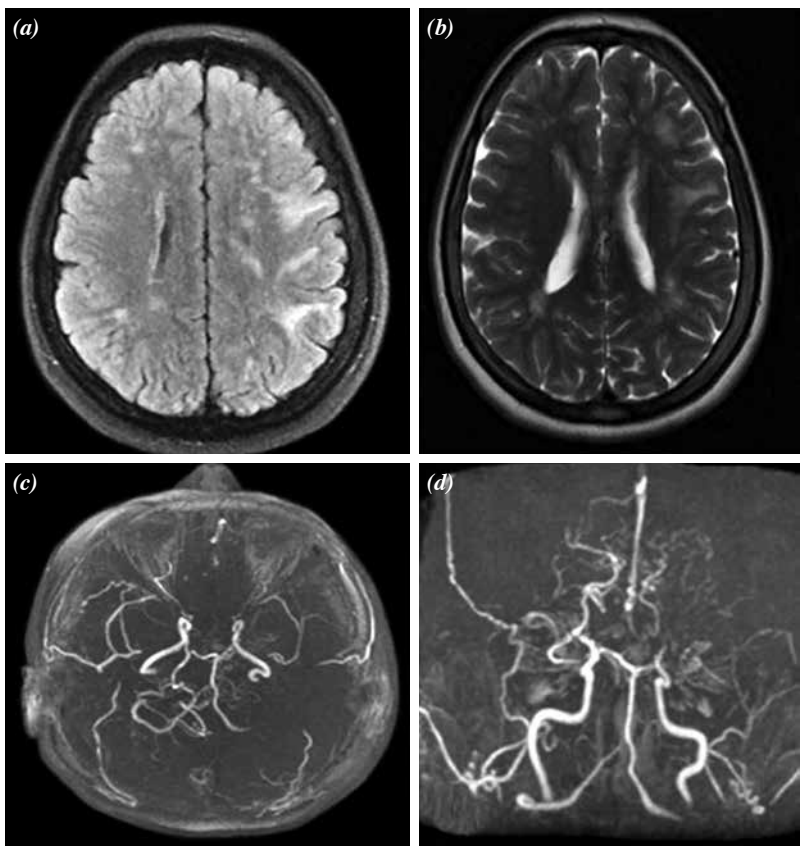

Figure 1. ***(a, b)*** *In brain magnetic resonance imaging flair and T2 axial sections, hyperintense lesions are noted, prominent on left.* ***(c, d)*** *Brain magnetic resonance angiography shows that major vessels are not adequately observed as from distal of internal carotid artery.*

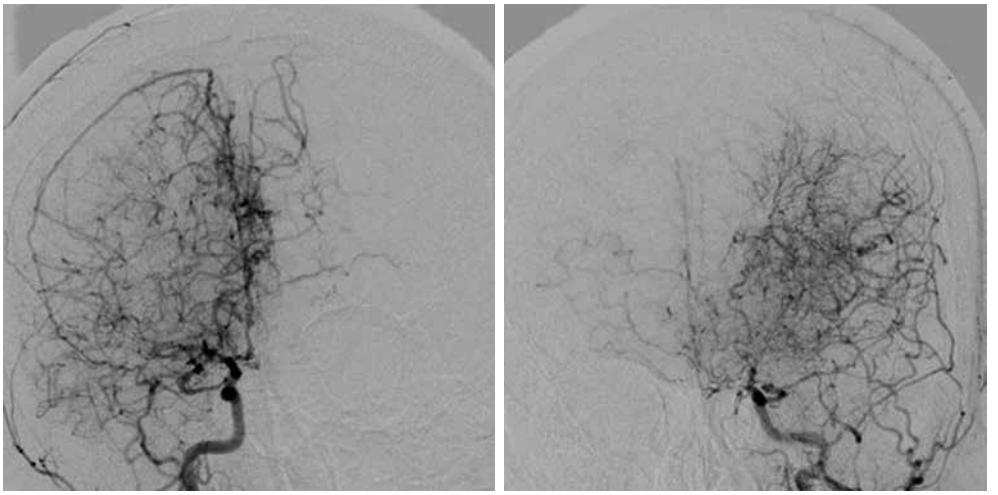

Figure 2. *In digital substraction angiography, branches of the right and left anterior cerebral and middle cerebral arteries thinning towards distal. Distal segments show "clouds of smoke" collateral circulation vessels.*

IgM tests in blood were detected to be negative. Brain MR angiography was performed for etiology. Left cerebellar hemisphere, medial cerebral hemisphere and anterior and posterior cerebral artery flows were not observed. Weak flow in the right medial cerebellar artery and posterior cerebellar artery, enlarged intracranial perforating arterial structures of intracranial anastomoses in bilateral thalami and basal ganglia localization, extracranial and intracranial anastomoses via bilateral ophthalmic artery and dural perforating arteries were observed (Figure 1c, d).

These MR angiographic findings of the patient were evaluated in favor of MMD and the patient underwent cerebral digital subtraction angiography (DSA). Intracranial internal carotid arteries were gradually thinned towards the distal and the calibrations of the medial cerebral artery (MCA) and the anterior cerebral artery (ACA) were decreased, opaque passage was observed in right and left MCA and ACA from proximal to distal and several enlarged collaterals formed a typical smoke appearance (moyamoya pattern; puff of smoke appearance) (Figures 2 and 3). The patient was diagnosed with MMD. Low-dose acetylsalicylic acid was started. A written informed consent was obtained from the patient.

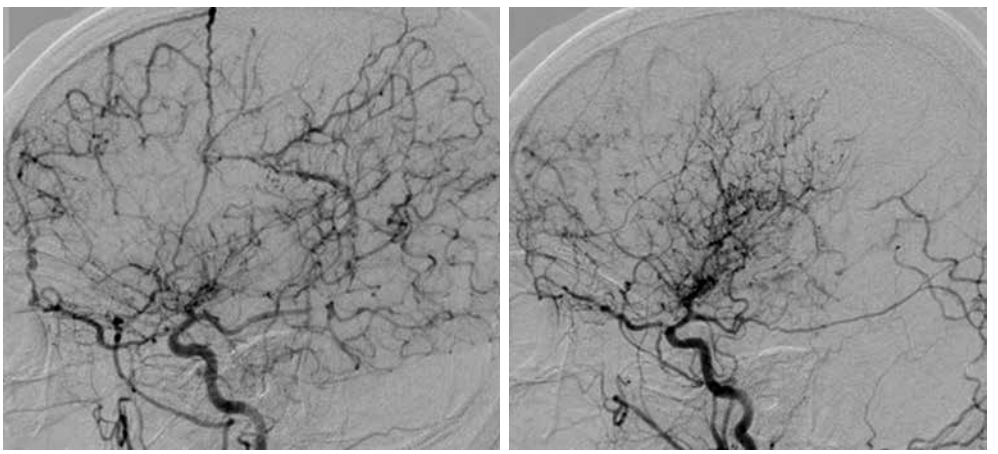

Figure 3. *Extracranial-intracranial anastomoses are commonly seen in digital subtraction angiography.*

DISCUSSION

Moyamoya disease is common in children as a transient ischemic attack or seizure, while in adults, it is observed as hemorrhagic stroke.[6,7]

Involuntary movements are rare in moyamoya patients, observed in only 3-6% of patients, and more common in children. The most common movement disorders are chorea, hemidystonia, athetosis and paroxysmal dyskinesia.[6,8]

Chorea gravidarum is a rare disease of pregnancy and usually occurs in the first trimester of pregnancy.[9-12] About half of the cases are idiopathic. It has been suggested that the main pathophysiology in idiopathic chorea gravidarum is the increased dopaminergic sensitivity in basal ganglia due to estrogen levels.[9,13] However, other underlying etiologies such as rheumatic fever, antiphospholipid syndrome, other autoimmune diseases, Wilson's disease, thyrotoxicosis, Huntington's disease, subcortical infarction, thrombophilic disease and medication related chorea should also be considered.[9,10,12,13] There are a limited number of case reports in the literature with chorea gravidarum and MMD.

Baik et al.[6] collected patients with MMD and movement disorders from the literature and hospital archives. They identified chorea gravidarum in two of the 42 patients they obtained. Three patients also reported that chorea was developed during oral contraceptive use. They hypothesized that hypoperfusion or ischemia in basal ganglia may reveal chorea with the presence of a hormonal disorder in patients without infarct in this area.

Kim et al.[9] reported a case in which chorea gravidarum developed after hyperemesis gravidarum and examinations revealed MMD. They hypothesized that hyperemesis gravidarum-related hyperventilation caused transient cerebral ischemic disorder. Moreover, Jung et al.[8] presented a case of MMD in which chorea developed in right hand after consumption of hot meal. They suggested that hyperventilation due to hot meal consumption caused hypocarbia which resulted in vasoconstriction and vasospasm.

Furthermore, Baik et al.[6] found in their review that, in cerebral perfusion studies, cerebral blood flow in basal ganglia and cerebral cortex is decreased in four of five patients who do not have ischemic lesions in brain MRI, but chorea.

The cause of development of chorea in a pregnant moyamoya patient may be bleeding or infarct located in the basal ganglion or subthalamic region. If no pathology is detected in these areas by imaging methods, there may be dopaminergic hypersensitivity in the basal ganglion due to gender hormones, transient circulatory problems due to impaired vascular structure, or if the patient has a hyperemesis gravidarum, hyperventilation may cause vasospasm and hypoperfusion in the basal ganglia.[6,8,9]

The first presentation of MMD in our patient was chorea gravidarum. However, since the brain MRIs performed at that period were interpreted as demyelinating, MMD was not diagnosed and the complaints of the patient were improved by haloperidol treatment. With brain MRI and brain MR angiography images of the patient at admission to our clinic, we thought that the patient may have MMD, and cerebral DSA findings also supported MMD diagnosis with typical appearance. Our patient already had cerebral perfusion impairments due to MMD, and we believe that hormonal fluctuations due to pregnancy severely worsen this perfusion, cause transient hypoperfusion in the basal ganglia, thalamocortical pathways, and may play a role in the development of chorea in pregnancy.

Chorea gravidarum along with MMD is a very rare condition. The first clinical finding of our patient was chorea gravidarum eight years before, and the brain MRI scans taken at that time were thought to be demyelinating or posterior reversible encephalopathy syndrome-like lesions and the case was mostly studied from this perspective. She could be diagnosed after eight years, when she applied to the stroke clinic. In patients under chorea gravidarum etiology investigation, lesions in MRI should be considered as MMD, even if not typical, in terms of ischemia and vascular imaging should be performed.

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