



# Choreiform Movement Disorder Secondary to Intracerebral Cavernoma: Case Report

## *İntraserebral Kavernoma Bağlı Koreiform Hareket Bozukluğu: Olgu Sunumu*

Banu Özen Barut, Bekir Enes Coşkun, Özlem Türksöy\*  
Bülent Ecevit University, Department of Neurology, Zonguldak, Turkey  
\* Bülent Ecevit University, Department of Radiology, Zonguldak, Turkey

### Summary

Although vascular factors are considered primarily in the etiology of hemichorea among the elderly, chorea related to congenital vascular malformations including cavernous angioma are reported very rarely. We found cavernous angioma to be the cause of persistent, unilateral chorea localized to the right lower extremity in a 83 year-old woman. In this article we would like to highlight the etiology of chorea in patients with late onset movement disorders. (*Turkish Journal of Neurology* 2013; 19:60-2)

**Key Words:** Choreia, vascular malformation, cavernoma

### Özet

Unilateral kore nedeniyle değerlendirilen yaşlı olgularda etyolojide vasküler nedenler ilk sırada yer alır. Ancak kaverno anjiyom gibi konjenital, vasküler malformasyonlara bağlı kore oldukça nadirdir. Bu yazıda, 83 yaşında tek taraflı, alt ekstremiteye lokalize kore nedeniyle değerlendirilen ve etyolojide kaverno anjiyom saptanan kadın hastayı sunmak ve bu olgu aracılığı ile geç başlangıçlı hareket bozukluklarında kore etyolojisini tartışmak istedik. (*Türk Nöroloji Dergisi* 2013; 19:60-2)

**Anahtar Kelimeler:** Kore, vasküler malformasyon, kavernom

### Introduction

Chorea, one of many hyperkinetic movement disorders, is named after the word choreia (dance, in Greek) and used to describe the random, sudden and involuntary spasms spreading from one side of the body to the other. These movements involve continuous, short, low-amplitude and irregular muscle contractions, and, although seen mostly in distal extremities, can also be seen in the face and tongue. Severe chorea may impair speech and swallowing functions, as well as posture. In addition, it may cause grimacing and abnormal breathing sounds, too. Patients may partially suppress these movements and usually try to conceal them with semi intentional movements. When movements are mild, they are hard to distinguish from restlessness and may cause a similar feeling of restlessness in the observer. Patients usually complain of clumsiness and loss of coordination including dropping or bumping into objects.

The etiology of chorea is varied and includes genetic, autoimmune, infectious causes, neurodegenerative diseases, vascular or tumoural pathologies causing structural lesions in the basal ganglia, toxic or metabolic causes and side effects of drugs (1,2). Unilateral chorea is known as hemichorea and is usually associated with vascular causes, including cerebral infarctions, arteriovenous malformations and subdural hematomas (2). Cavernomas rarely cause chorea and there are fairly few cases reported in the literature (3,4,5,6,7,8). Here we discuss a case of intracerebral cavernoma identified as a result of investigations in a case of chorea where the initial clinical picture of hemichorea later transformed to limited chorea of unilateral lower extremity.

### Case

Eighty-five year old female patient presented at our neurology clinic with complaints of involuntary movements in lower right extremity that started 2 years ago in the right hand and foot as

**Address for Correspondence/Yazışma Adresi:** Banu Özen Barut MD, Department of Neurology, Bülent Ecevit University, Zonguldak, Turkey  
Gsm.: +90 533 735 12 63 E-mail: bozen73@yahoo.com

**Received/Geliş Tarihi:** 05.05.2012 **Accepted/Kabul Tarihi:** 28.11.2012

involuntary movements. Haloperidol treatment prescribed by another site and used for some time provided partial benefit and the symptoms in her hand resolved. However, the involuntary movements in the lower right extremity have been continuing without any change for two years, the severity of the symptoms has increased in the last 3 months. There was no history of systemic or psychiatric disease in the patient's medical history. There was no drug usage prior to the start of her complaints. There was no history of similar disease in the family. Physical examination findings were normal. Neurological examination showed that cranial nerves were normal. Motor examination showed that upper and lower extremity strength was complete. Continuous, irregular choreiform movements were observed in distal lower right extremity during motor examination, which increased with emotional stress and did not change in form when the patient was distracted or was walking. Muscle tone had clearly decreased in the lower right extremity. Sensory and cerebellar examination was normal. Deep tendon reflexes (DTR) were normally active in all foci, and no pathologic reflexes were observed. Complete blood count, fasting blood sugar, liver and kidney function tests, and erythrocyte sedimentation rate results were within normal limits. Thyroid function tests, antinuclear antibody, VDRL, brucella, HOV and hepatitis markers were all negative.

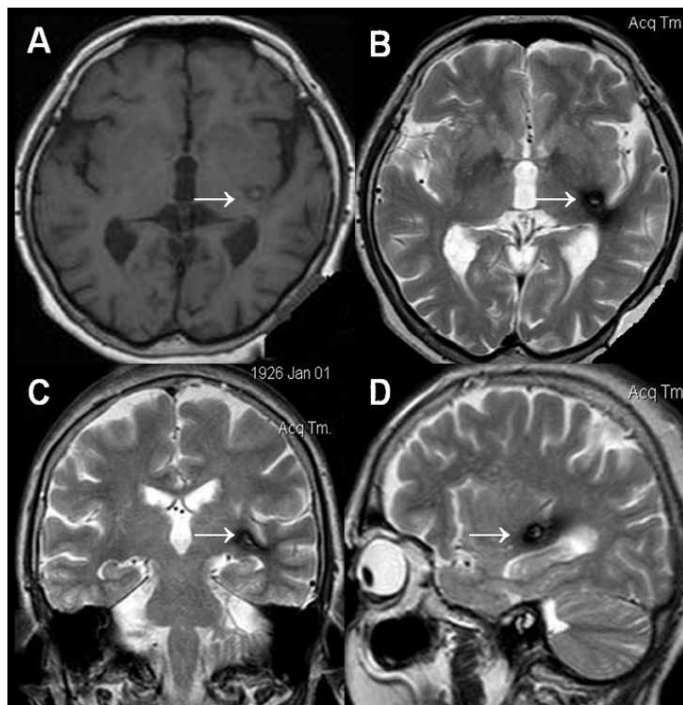
Cranial magnetic resonance imaging (MRI) showed a lesion consistent with a cavernous angioma in the T1-weighted (T1W) and T2-weighted (T2W) sequences, in the posterior crus of the left internal capsule, sublentiform and retrolentiform region, adjacent to the insular cortex and lateral left thalamus, containing hyperintense, late subacute stage hematoma with a diameter of approximately 10 mm and without mass effect, surrounded by a hypointense hemosiderin ring (Image 1). This localization of this lesion was thought to be consistent with the choreiform movement disorder starting as hemichorea on the left and later limiting to the lower left extremity. Following a neurosurgical consultation, follow-up with MRI scan every 3 months was recommended, considering the patient's old age and his symptoms not having much impact on activities of daily living. Olanzapin 5 mg 1x1 was initiated for symptomatic treatment; although the dosage was increased to 10 mg later, it was discontinued at the follow-up visit a month later, due to the sedation side effect and lack of considerable benefit for the patient. Sodium valproate 500 mg 1x1 was initiated later, and the dose was increased to 1000 mg. The patient did not benefit from this treatment after a period of two months. The patient had been using amantadine 200 mg at the time of writing of this article, without any benefit.

## Discussion

Acute or subacute onset chorea in an elderly patient usually suggests secondary causes, including drugs, vascular diseases, metabolic causes, autoimmune diseases, neoplasms, senility and hereditary causes. Unilateral choreiform movements are called hemichorea, account for approximately 0.7% of all movement disorders, and usually co-exist with a structural pathology such as contralateral vascular lesion, tumour or tuberculoma or sometimes with diabetic ketoacidosis (2). Some of the vascular pathologies causing chorea are ischemic or hemorrhagic cerebrovascular diseases, and vascular malformations. Cerebrovascular diseases are

the most common causes of chorea among all movement disorders. Vascular malformations include venous angiomas, arteriovenous malformations and cavernous angiomas (9,10). Vascular anomalies such as cerebral arteriovenous malformations or cavernomas rarely cause chorea (11).

Cavernous hemangiomas or cavernomas are congenital vascular malformations and comprise 8-15% of all cerebral vascular malformations. Cavernomas are vascular cavities that have a thin wall made up of a thin endothelium layer, and that does not contain any smooth muscle or elastin (12). They are usually located in the supratentorial region, subcortical parenchyma, commonly adjacent to the rolandic fissure, in the basal ganglia or brain stem. Most of them are asymptomatic and are diagnosed coincidentally or at a post mortem examination. Symptomatic cavernomas usually become so in the third or fourth decade of life (11). Considering our patient was diagnosed in the ninth decade of his life, this process may even be longer. Cavernomas may cause seizures, headaches and focal neurologic deficits. Movement disorders due to cavernomas are quite rare (12). Among these are cases of hemichorea localized in the caudate nucleus (4,5,6,7,8), a case of chorea resulting from a cavernoma localized in the putamen (3), cases of cavernoma causing dystonia (13,14,15,16,17) and cases of parkinsonism caused by cavernomas (18,19,20,21). Cases of cavernoma causing chorea are rare and lesion is usually localized in the caudate nucleus (4,5,6,7,8). Three of these cases are above the age of 65, similar to our patient. Chorea improved in one of them without treatment, symptoms were too mild to require treatment (6,8). The third case benefited from surgical treatment (4). The



**Image 1:** Cavernous angioma localized in the posterior crus of the left internal capsule, sublentiform and retrolentiform region. A. Axial T1W sequence. B. Axial T2W sequence. C. Coronal T2W sequence. D. Sagittal T2W sequence.

other two cases were 7 and 11 years old; one case benefited from pimozide treatment, and the other from surgical treatment (5,7). The clinical picture in all five cases was described as hemichorea. The lesion in another cavernoma case resulting in hemichorea was identified in the putamen (3). The clinical picture in this 65-year old patient was assessed as hemichorea and the patient benefited from 1000 mg sodium valproate treatment (3). The cavernoma in our case was described to be localized in the posterior crus of the left internal capsule, adjacent to the rolandic fissure and in the retrolentiform region (Image 1). Our patient did not benefit from medical treatment and was assessed to be refractory chorea.

Changes in the hemodynamics of cerebral parenchyma explain the cavernoma becoming symptomatic. These changes occur secondary to hemorrhages and cavernoma may become symptomatic via irritative effect. The change in cerebral hemodynamics may sometimes occur through microhemorrhages (16). We thought the symptomatic transformation of the lesion in our case may be related to the bleeding in the cavernoma and was associated with the hemosiderin ring surrounding the lesion.

There has been attempts to explain the etiopathogenesis of chorea by an impairment in the balance between direct and indirect pathways. As a result of the impact on striatal output projecting on globus pallidus, ie indirect pathway, the inhibition on the neurons in this region increases; this, in turn, increases the inhibition on the subthalamic nucleus and decreases the output from the internal globus pallidus. As a result, thalamo-cortical activity increases, creating the main mechanism thought to underlie hyperkinetic disorders. In our case, we thought that the lesion's adjacent localization to the subthalamic nucleus caused damage to the associated indirect pathway, in turn causing chorea symptoms (8,9).

In conclusion, among vascular causes of chorea, cavernoma angiomas may cause chorea, albeit rarely. The lesion, in these cases, is occasionally localized in the basal ganglia, and patients mostly present with hemichorea. Chorea is sometimes refractory and does not respond to medical treatment.

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