MOYAMOYA SYNDROME WITH ARTERIOVENOUS DURAL FISTULA AFTER HEAD TRAUMA

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SUMMARY – Moyamoya vascular pattern and dural arteriovenous fistula (dAVF) are rare vascular abnormalities and both can be secondary to head trauma. The role of dural angiogenesis in the pathophysiology of vascular malformation is rather unclear. We report a unique case of moyamoya vasculopathy simultaneously associated with dAVF after heavy head trauma. It seems that both moyamoya syndrome and dAVFs are associated with dural angiogenesis induced by head trauma. The interrelationship between vascular abnormalities is complex and unclear.

Key words: *Moyamoya disease; Craniocerebral trauma; Intracranial arteriovenous malformations; Angiogenesis*

Introduction

Moyamoya syndrome is an uncommon cerebrovascular disorder that is characterized by progressive stenosis of the terminal portion of the internal carotid artery and/or its main branches. Intracranial dural arteriovenous fistula (dAVF) is also a rare vascular lesion. The etiology of both entities is unclear, although they have been tried to be connected with several conditions and diseases. Thus, both entities could appear after head trauma, however, to our knowledge, simultaneous occurrence of both conditions has not yet been reported. We describe a case of moyamoya vasculopathy and AVF in a patient having sustained severe head trauma.

Case Report

A 71-year-old right-handed forester was admitted to the Neurology Department due to burning headache, located deep in the right hemisphere, that had gradual onset and had started four days earlier. Twenty-four years before, he suffered a major head and chest injury after he was crushed under a tree. He was comatose for a couple of weeks. Upon recovery, slight left hemiparesis remained. At that time, computed tomography scan did not show any pathological abnormalities. He had mild arterial hypertension but did not take any medication. Over the past couple of months, he noticed that he could not empty his bladder properly.

On admission, he was sitting in a wheelchair and was unable to walk unattended. His blood pressure was 187/97 mm Hg, heart rate 69 beats *per* minute and body temperature 37.5 °C. He was abulic, disoriented and unwilling to cooperate. On the mini mental test, he scored 14/30. He had mild central paresis of the left facial nerve and spastic hemiparesis with brisk reflexes and extensor plantar response on the left side. In addition, paratonia of the right limbs and poor dexterity was present. The gait was slow, widely based and hemiparetic. CT showed subarachnoid hemorrhage in both frontal and parietal lobes (Fig. 1). Magnetic resonance imaging showed bilateral subcortical ischemic lesions distributed mainly in the territory supplied by the internal carotid artery (Fig 3). Cerebral angiography revealed severe narrowing of both internal carotid

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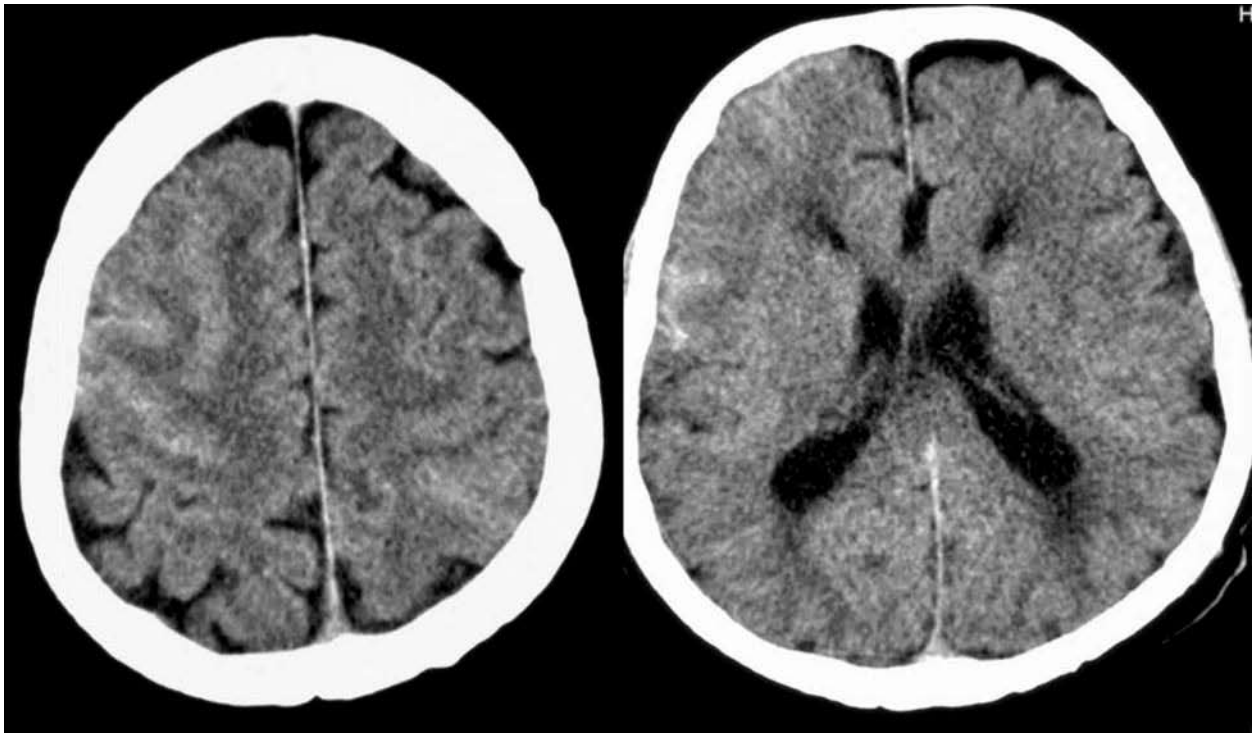

Fig 1. Computed tomography in axial plane shows bleeding into the subarachnoid space of the frontal and parietal lobes on both sides (subarachnoid hemorrhage).

arteries and occlusion of the M1 segment of both middle cerebral arteries. Collateral perforating lentiform arteries were prominent (Fig. 3). Angiography of the right common carotid artery showed collateral cerebral circulation from external carotid artery and a dAVF that was fed by branches of occipital artery. The AVF drained into the right transverse sinus. Angiography of the left vertebral and basilar arteries revealed prominent and tortuous perforate and posterior choroidal arteries presenting a typical moyamoya pattern. Apart from arterial hypertension, he had no risk factors for cerebrovascular diseases. Color duplex sonography of the neck arteries showed small, partially calcified, and hemodynamically insignificant plaques in both carotid bifurcations. The patient was treated conservatively using calcium channel antagonist, analgesics and antihypertensive therapy. Subarachnoid hemorrhage spontaneously reabsorbed. The cognitive status remained the same. We did not decide for operative therapy due to the preexisting cognitive impairment. The patient was discharged in stable clinical condition.

Discussion

We describe an unusual case of bilateral moyamoya syndrome associated with dural arteriovenous fistula in an elderly Caucasian patient admitted to neurology department due to subarachnoid hemorrhage. Extensive review of the literature revealed only a few cases of simultaneous presentation of moyamoya vasculopathy and arteriovenous fistula. There is only one report of unilateral moyamoya syndrome and dural arteriovenous fistula and only a few reports of moyamoya disease in combination with different arteriovenous malformations[1,2]. The main question about the mentioned entities is the pathophysiological mechanism that remains unexplained. Our case report, in which moyamoya syndrome and dural arteriovenous fistula occurred simultaneously years after head trauma, may suggest some new speculations concerning the pathophysiological mechanisms that partially or completely underlie both entities.

In our patient, clinical presentation was by no means uncommon for both entities. The subarachnoid

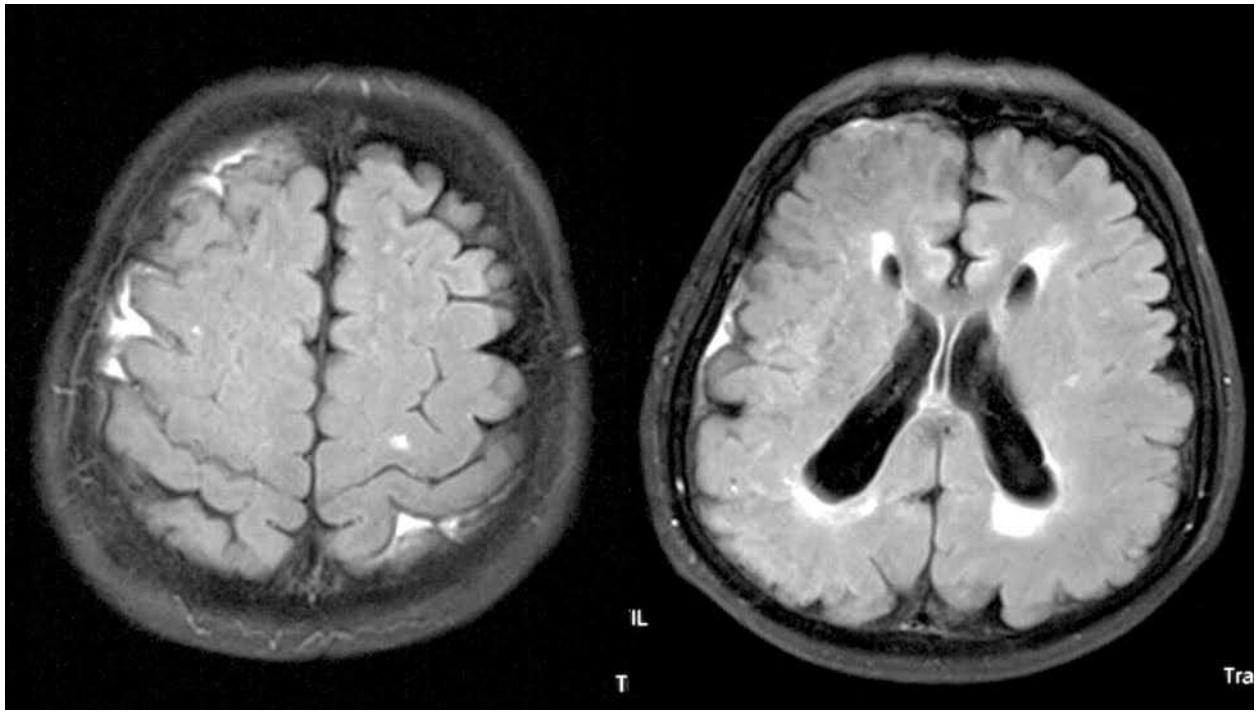

Fig 2. MRI flair sequence in axial plane shows subarachnoid hemorrhage and ischemic lesions bilaterally.

hemorrhage could be attributed either to the dural arteriovenous fistula or to the moyamoya syndrome. The rupture of the fragile collateral vessels associated with stenosis of the internal carotid artery resulting in intracranial hemorrhage is common in patients with moyamoya syndrome[3]. The cognitive decline could be explained by the widespread subcortical ischemic lesions, which were probably also part of the pathological process occurring in moyamoya syndrome.

Although moyamoya disease is commonly found in East Asia, the moyamoya vasculopathy has been observed throughout the world[4]. It has been shown that it has two peaks of occurrence: one at 5 years of age and another one, lower peak at about 40 years of age[5]. Importantly, our patient was significantly older. Therefore, we could convincingly assume that moyamoya vasculopathy and dural arteriovenous fistula in our patient were acquired rather than congenital.

The precipitating factor for both entities in our patient could have been previous severe head trauma. It has already been reported that head trauma is associated with dural arteriovenous fistula[5]. Our patient had a dural arteriovenous fistula located on the side of the previous head trauma. On the other hand, head trauma could have been a secondary cause of moyamoya vasculopathy as well. We could assume with certainty that the altered angiogenesis occurred within dura after the trauma. Altered dural angiogenesis can be accompanied by sinus venous thrombosis[7], facilitating the occurrence of dural arteriovenous fistula. Furthermore, altered angiogenesis could induce the occurrence of moyamoya vasculopathy.

The relationship between moyamoya disease and dural arteriovenous fistula is not clear. It seems that dural arteriovenous fistula could cause the moyamoya pattern and *vice versa*. According to Mawad *et al.*[8], the predilection of vascular occlusion for the internal carotid may be related to the increase in blood turbulence proximal to the dural arteriovenous fistula. Namely, the increased turbulent flow related to the hemodynamics of the arteriovenous malformation could lead to intimal hyperplasia, endothelial thickening, and progressive occlusion of the feeding vessels. Nevertheless, the fistula in our patient was supplied entirely from the external carotid artery, thereby reducing blood flow through the internal carotid artery. The idea that vascular occlusive disease leads to dural arteriovenous fistula seems to be more appealing.

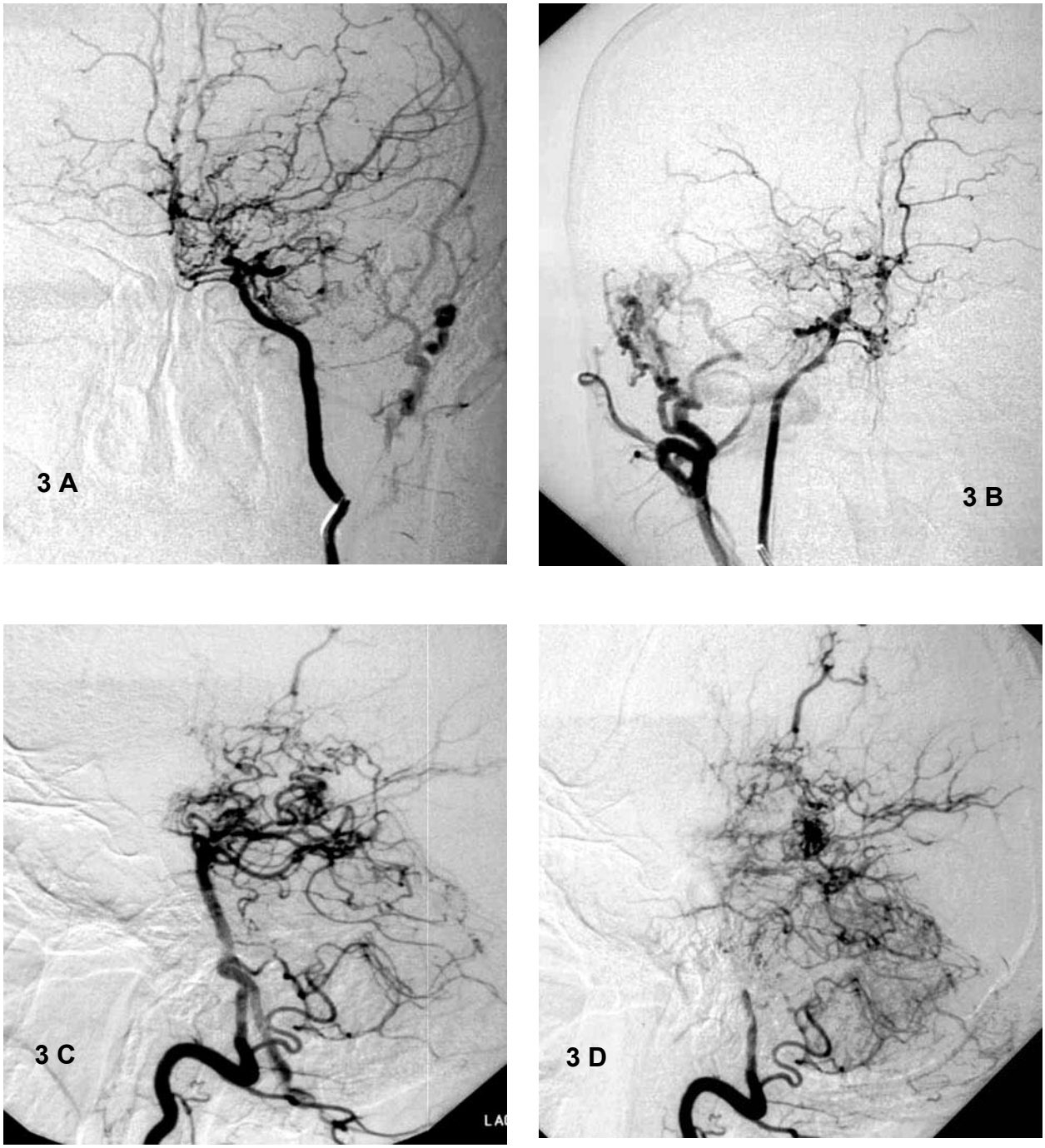

Fig. 3 Angiography: (A) angiographic injection of the left internal carotid artery showing severe narrowing of the left internal carotid artery and occlusion of the left M1 segment; (B) angiographic injection of the right internal carotid artery showing severe narrowing of the right internal carotid artery, occlusion of the right M1 segment and collateral cerebral circulation from external carotid artery. Collateral perforating lentiform arteries are prominent. dAVF is fed by branches of occipital artery and drained into the right transverse sinus; (C) and (D) angiographic injection of the left vertebral and basilar artery showing prominent and tortuous perforating arteries in posterior choroidal arteries presenting typical moyamoya pattern.

Several previous studies have described an association between moyamoya disease and arteriovenous malformations indicating causal relationship[1,2]. Previous studies have also shown that the levels of several factors including basic fibroblast growth factor (bFGF)[9], transforming growth factor β-1, hepatocyte growth factor, vascular endothelial growth factor[10], matrix metalloproteinases, intracellular adhesion molecules, and hypoxia-inducing factor 1α are elevated in the dura of patients with moyamoya disease[11,12]. It could be speculated that such ischemic environment may have contributed to the formation of dural arteriovenous fistula in our patient. Recent investigations have shown separately that both moyamoya syndrome and dural arteriovenous fistula could be associated with dural angiogenesis promoted by head trauma[13,14]. Thus, angiogenesis in response to the release of angiogenic factors induced by trauma may simultaneously lead to the development of moyamoya vasculopathy and dural arteriovenous fistula as well.

Although somehow speculative, our explanation(s) might help explain or at least bring a new view on the unknown etiology of these entities. However, our case could also represent the extremely rare coincidence of two rare phenomena. In conclusion, based on our particular case, we suggest that both moyamoya syndrome and dural arteriovenous fistula might be associated with dural angiogenesis due to severe head trauma.

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Sažetak

MOYAMOYA SINDROM S ARTERIOVENSKOM FISTULOM DURE NAKON OZLJEDE GLAVE

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Vaskularna struktura moyamoya i arteriovenska fistula dure (dAVF) su rijetke krvožilne nepravilnosti koje mogu nastati kao posljedica ozljede glave. Uloga duralne angiogeneze u patofiziologiji vaskularne malformacije prilično je nejasna. Opisujemo jedinstven slučaj moyamoya vaskulopatije istodobno udružene s dAVF nakon teške traume glave. Čini se da su i sindrom moyamoya i dAVF udruženi s duralnom angiogenezom izazvanom ozljedom glave. Međuodnos vaskularnih nepravilnosti je složen i nejasan.

Ključne riječi: *Bolest moyamoya; Kraniocerebralna ozljeda; Intrakranijske arteriovenske malformacije; Angiogeneza*