

Moyamoya disease and arteriovenous fistula of the epiaortic vessels

K. Savio · D. Mittino · E. Terazzi · I. Divenuto ·
R. Fossaceca · A. Carriero · F. Monaco

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Abstract The association of Moyamoya disease (MD) with other cerebrovascular disease has been reported in literature but not the occurrence of MD and arteriovenous fistula of the epiaortic vessels. A 61-year old Italian woman was admitted to our Department because of the sudden onset of Broca's aphasia. At general examination she presented a murmur above the right clavicle. Brain magnetic resonance imaging (MRI) showed an ischemic lesion involving left frontal and temporal lobes. Cerebral digital subtraction angiography (DSA) showed moyamoya vessels and an arteriovenous fistula between vertebral artery and internal jugular vein in right laterocervical region. This is the first report of the association of MD with arteriovenous fistula of the epiaortic vessels.

Keywords Moyamoya disease · Vascular malformations · Arteriovenous fistula

Introduction

Moyamoya disease (MD) is an occlusive cerebrovascular disease characterized by bilateral steno-occlusion at the terminal branch of the internal carotid artery (ICA) [1].

Compensation for occlusion results in rich arterial collaterals at the base of the brain, the so called moyamoya vessels, which means “puff of smoke” vessels in Japanese [2, 3].

The first idiopathic form was described in 1957 in Japan and subsequently worldwide, even though the highest incidence is reported in Japanese [2, 4].

The age distribution of patients with MD forms two characteristic peaks in children and adults [3]. In an Italian study of 1999, 34 patients with MD were studied; also in Italy there are two peaks in age distribution: under 10 years and in the 30s [5].

Etiology of the disease remains unknown, but this condition is frequently familial, suggesting the involvement of genetic factors in its pathogenesis. Contribution of infections has also been pointed out, although specific pathogens have not been identified [6].

As known, MD causes transient ischemic attacks (TIAs), cerebral infarction or intracranial bleeding in both children and adults [6].

Superficial temporal artery–middle cerebral artery anastomosis is considered the standard surgical treatment for MD [1].

Although MD is typically an arterial disease of the brain, concomitant vascular lesions in extracranial organs such as the heart and the kidney have also been described [7].

The association of MD with other cerebrovascular disease, such as aneurysms, cavernous malformations and venous malformations, has been reported in literature [8].

Recently, the simultaneous occurrence of unilateral MD and ipsilateral dural arteriovenous fistula have been described in a 44-year old man [9].

The occurrence of MD and arteriovenous fistula of the epiaortic vessels has never been described.

K. Savio (✉) · D. Mittino · E. Terazzi · F. Monaco
Department of Neurology, Amedeo Avogadro University, corso
Mazzini 18, Novara, Italy
e-mail: katiasavio@libero.it

I. Divenuto · R. Fossaceca · A. Carriero
Department of Radiology, Amedeo Avogadro University, corso
Mazzini 18, Novara, Italy

Case report

We report the case of a 61-year old Italian woman with a history of rheumatic heart disease and hypertension, who was admitted to Department of Neurology in December 2007 after the sudden onset of Broca's aphasia. The day before she underwent a surgical intervention for complications from an uretero–vaginal fistula, previously treated with ureteral stent. During the urological intervention blood transfusions were necessary because of severe bleeding.

At neurological examination she had Broca's aphasia and right hemiparesis. At general examination she presented a murmur above the right clavicle.

At laboratory test she had mild anemia with normal MCV and MCH, linked to the previous bleeding.

Electrocardiography and chest X-ray were normal

Computer tomography (CT) scan of the brain showed an ischemic lesion involving left frontal and temporal lobes that was confirmed by brain magnetic resonance imaging (MRI).

Electroencephalography showed left slowing in background activity, mainly in temporal and occipital regions.

Ecocolordoppler of epiaortic vessels showed carotid arteries intimal hyperplasia without stenoses of carotid, vertebral and subclavius arteries.

Echocardiography showed mitral calcification causing valve insufficiency

Magnetic resonance angiography (MRA) of the intracranial vessels showed severe stenotic changes in the right internal carotid artery (ICA) and moderate stenotic changes in the left one; collateral networks were evident bilaterally in the basal ganglia. MRA of the epiaortic vessels showed a probable arteriovenous fistula between vertebral artery and internal jugular vein in right laterocervical region (Fig. 1).

Transcranial color-coded sonography showed the presence of very slow flow at the terminal segment of the two MCA and no others pathological findings.

Patient underwent cerebral digital subtraction angiography (DSA), that showed a stenosis of the supraclinoid portion of right ICA and a severe stenosis of the left ICA after the origin; stenosis were also found at right and left anterior and middle cerebral arteries (ACA and MCA) with the presence of extensive leptomeningeal collaterals, in particular in the left hemisphere, consistent with MD (Fig. 2). DSA also confirmed the presence of an arteriovenous fistula between V2 portion of vertebral artery and internal jugular vein in the right side of the neck (Fig. 3).



Fig. 1 Extracranial and intracranial MRA. “Puff of smoke” aspects of the intracranial vessels (white arrow). Arteriovenous fistula between vertebral artery and internal jugular vein in right laterocervical region (white dashed arrow)

During hospitalization therapy with acetylsalicylic acid was started.

At discharge Broca's aphasia and right hemiparesis improved.

In July 2008 she successfully underwent endovascular closure of laterocervical arteriovenous fistula at Neurosurgery Department of Policlinic Hospital in Milan. She is waiting for superficial temporal artery–middle cerebral artery anastomosis.

Discussion

The association of MD with other vascular malformation is not so rare.

Akasaki et al. [10] reported of an asymptomatic MD associated with coronary and renal artery stenoses, while St Goar et al. [11] presupposed moyamoya of the heart.

Gatti et al. [7] have recently reported of a 56-year old white male with MD who presented a coronary fistula. He had an aortic valve stenosis and he underwent aortic valve replacement; at selective coronary angiography, two right coronary artery-to-right ventricular cavity fistulas were observed, without obstructive lesions.

Januschek et al. [8] have recently reported of a progressive moyamoya syndrome associated with de novo formation of ipsilateral venous and contralateral cavernous malformations in a 33-year old man.

Fig. 2 Cerebral DSA (lateral and frontal view). Right PCA supplies MCA and ACA (black arrow). Left PCA poorly supplies left MCA and ACA (dashed black arrow). “Puff of smoke” aspect (white arrows)

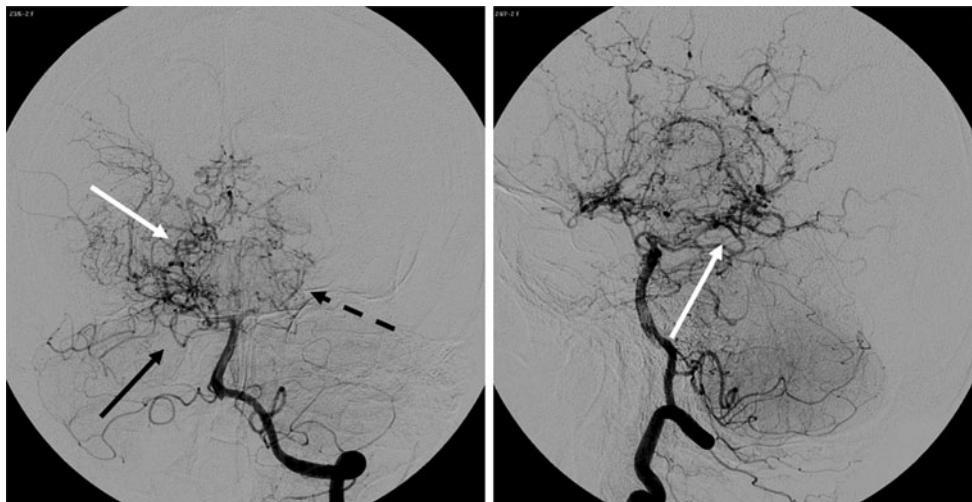


Fig. 3 Cerebral DSA. DAVF between vertebral artery and internal jugular vein in right laterocervical region (black arrows)



Kerchner et al. [12] reported the case of a young woman with occurrence of a cerebral cavernous malformation and contralateral moyamoya-like occlusion of the right MCA.

Wu et al. [13] reported of a case of a 36-year old man with a cerebral arteriovenous malformation treated with radiosurgery who developed MD after this therapy.

Recently Killory et al. [9] have described the case of a 44-year old man with simultaneous unilateral MD and ipsilateral dural arteriovenous fistula. Previous studies have shown that angiogenic factors are elevated in dura mater of MD patients, which may represent a mechanism for the formation of dural arteriovenous fistula.

Conclusion

To the best of our knowledge, this is the first report of the association of MD with arteriovenous fistula of the epiaortic vessels.

As outlined by previous reports, we believe that other concomitant vascular malformations should always be considered in patients with MD.

References

1. Fujimura M, Kaneta T, Tominaga T (2008) Efficacy of superficial temporal artery–middle cerebral artery anastomosis with routine postoperative cerebral blood flow measurement during the acute stage in childhood moyamoya disease. *Childs Nerv Syst* 24(7):827–832
2. Kuriyama S, Kusaka Y, Fujimura M, Wakai K, Tamakoshi A, Hashimoto S, Tsuji I, Inaba Y, Yoshimoto T (2008) Prevalence and clinicoepidemiological features of moyamoya disease in Japan. *Stroke*. 39:42–47
3. Houkin K, Kamiyama H, Abe H, Takahashi A, Kuroda S (1996) Surgical therapy for adult moyamoya disease: can surgical revascularization prevent the recurrence of intracerebral haemorrhage? *Stroke*. 27:1342–1346
4. Aoyagi M, Fukai N, Yamamoto M, Nakagawa K, Matsushima Y, Yamamoto K (1996) Early development of intimal thickening in

- superficial temporal arteries in patients with moyamoya disease. *Stroke*. 27:1750–1754
5. Battistella PA, Carollo C (1997) Clinical and neuroradiological findings of moyamoya disease in Italy. *Clin Neurol Neurosurg* 99(Suppl 2):S54–S57
 6. Research Committee on Moyamoya Disease in Japan (2007) Radiological findings, clinical course and outcome in asymptomatic moyamoya disease. Results of multicenter survey in Japan. *Stroke* 38:1430–1435.
 7. Gatti G, Benussi B, Gon L, Zingone B (2007) Aortic valve replacement in an adult white male with moyamoya disease and coronary artery fistulas. *J Cardiothorac Vasc Anesth* 21(1): 166–168
 8. Januschek E, Fujimura M, Mugikura S, Tominaga T (2008) Progressive moyamoya syndrome associated with de novo formation of the ipsilateral venous and contralateral cavernous malformations: case report. *Surg Neurol* 69(4):423–427
 9. Killory BD, Gonzales FL, Wait SD, Ponce FA, Albuquerque FC, Spetzler RF (2008) Simultaneous unilateral moyamoya and ipsilateral dural arteriovenous fistula: case report. *Neurosurgery*. 62:E1375–E1376
 10. Akasaki T, Kagiya S, Omae T, Ohya Y, Ibayashi S, Abe I, Fujishima M (1998) Asymptomatic moyamoya disease associated with coronary and renal artery stenoses—a case report. *Jpn Circ J* 62:136–138
 11. St Goar FG, Gominak SC, Potkin BN (1999) Bilateral aortoostial coronary artery disease: moyamoya of the heart? *Am J Cardiol* 83:1296–1299
 12. Kerchner GA, Smith W, Lawton MT, Singh V (2006) Co-occurrence of a cavernous malformation and contralateral moyamoya. *Neurology*. 66:1601–1602
 13. Wu TC, Guo WY, Wu HM, Chang FC, Shiao CY, Chung WY (2008) The rare association of moyamoya disease and cerebral arteriovenous malformations: a case report. *Korean J Radiol* 9(Suppl):S65–S67