Dural Arteriovenous Fistula in Moyamoya Angiopathy

14

Shambaditya Das, Souvik Dubey, and Biman Kanti Ray

Abstract

Moyamoya angiopathy (MMA) is an idiopathic, progressive intracranial occlusive vasculopathy leading to formation of abnormal vascular network of collaterals. Dural arteriovenous fistula (DAVF) has been rarely reported to co-exist with MMA. A paucity of literature has limited the elucidation of the pathophysiological association between the two conditions. It may be hypothesized that a chronic cerebral ischemic state in MMA and subsequent angiogenesis leading to the formation of collaterals might lead to the formation of DAVF. At present, there are no definite guidelines for treatment of DAVF in MMA. However, it may be reasonable to practice conservative management with close follow-up in case of asymptomatic DAVF or those without cerebral venous drainage, while high grade DAVF may be subjected to endovascular or open surgery to prevent future intracranial hemorrhage.

Keywords

Moyamoya angiopathy · Dural arteriovenous fistula · DAVF · Moyamoya disease Angiogenesis

S. Das (✉) · S. Dubey · B. K. Ray
Department of Neurology, Institute of Post Graduate Medical Education and Research, Bangur Institute of Neurosciences, Kolkata, India

14.1 Introduction

Moyamoya angiopathy (MMA) is a chronic progressive occlusive intracranial vasculopathy characterized by angiographic findings of stenosis or occlusion at the terminal portion of the internal carotid artery (ICA) or proximal anterior cerebral artery (ACA) and/or middle cerebral artery (MCA) together with the abnormal vascular network (classically appears to be like "puff of smoke") at the base of the brain [1, 2]. It can present with wide array of transient and fixed neurological manifestations and is a frequently overlooked cause of stroke [3–5]. MMA has been reported with various vascular malformations like intracranial aneurysms, arterio-venous malformations and primitive carotid-basilar anastomosis. Dural arteriovenous fistula (DAVF) is intra-cranial condition characterized by abnormal connections between meningeal arteries and dural venous sinuses, meningeal veins or cortical veins [6]. Both MMA and DAVF have extremely rare occurrence and their co-existence have only been uncommonly reported [6–12].

14.2 Pathophysiology

The pathogenesis of MMA is yet to be fully elucidated. Both genetic and inflammatory factors have been implicated. Adult DAVF is generally an acquired lesion [13–16]. Dural sinus thrombo-

X. Lv (ed.), *Intracranial and Spinal Dural Arteriovenous Fistulas*,
https://doi.org/10.1007/978-981-19-5767-3_14

sis or stenosis has been known play a pivotal role in the pathogenesis of DAVF. Besides, trauma, craniotomy or treatment of another DAVF has also been reported to be associated with formation of DAVF [6, 9].

Because the co-occurrence of MMA and DAVF is quite rare, a robust research related to the exact etio-pathogenetic connection between the two conditions is lacking. The proposed hypothesis is open-ended, wherein MMA can lead to the formation of DAVF and vice-versa [6, 12].

MMA is a chronic vaso-occlusive condition where a persistent cerebral ischemia is inevitable [1]. This contributes to release of pro-angiogenic factors like basic fibroblast growth factor (bFGF), vascular endothelial growth factor, transforming growth factor β-1, hepatocyte growth factor, intracellular adhesion molecules, matrix metalloproteinases and hypoxia-inducing factor 1α. It has been seen that this pro-angiogenic factors especially bFGF and vascular endothelial growth factor are elevated in the dura of both MMA and DAVF. It can be contemplated that this increased pro-angiogenic factors contribute to the angiogenesis, leading to formation of Moyamoya collaterals as well as formation of DAVF [6, 9–12].

Furthermore, cases of DAVF formation following revascularization surgery (especially extracranial-intracranial bypass) in MMA has been seen. It is not unusual that the cortical veins near the bypass graft may have intra-operative traumatic affection, thus, entertaining the possibility of an iatrogenic basis of DAVF in MMA [6, 9, 11]. Liu et al. [9] and Feroze et al. [11] described a similar situation wherein DAVF developed following the revascularization surgery for MMA.

As an extrapolation, it may also be postulated that MMA initiation and progression secondary to DAVF may occur as a consequence to increased turbulence proximal to DAVF, often in the distal ICA causing accentuated intimal hyperplasia and progressive occlusive changes of MMA [10, 12, 17]. While in three of the previously reported seven cases, MMA and DAVF were detected simultaneously, in rest four cases, DAVF was detected later to the diagnosis of MMA. Thus, the plausibility of the mechanism of chronic ischemia of MMA precluding the DAVF formation seems more [6–12].

Another theory suggests that head trauma, known to herald both DAVF and MMA, may lead to altered angiogenesis within the dura and subsequent sinus venous thrombosis and DAVF. This is supported by the case described by Zaletel et al., wherein DAVF and MMA were detected simultaneously following a history of head injury several years ago [12].

14.3 Management

MMA is a progressive disease with recurrent cerebrovascular events and progressive cognitive decline and merits revascularization surgery aimed at improving cerebral blood supply [1, 18]. There are no clear guidelines on the management of concomitant MMA and DAVF, and must be individualized depending on the clinical presentation and invasiveness [6]. While an asymptomatic DAVF and those without cortical venous drainage may be managed conservatively with a close follow-up, a high-grade DAVF should be managed aggressively through endovascular route or open surgery. DAVF with cortical venous drainage is a risk factor for future hemorrhagic events and deserves to be intervened (Fig. 14.1) [6]. Another consideration that the treating neurologist needs to be mindful of while intervening DAVF in the background of MMA with revascularization surgery is that the integrity of the bypass graft might get compromised during fistula disconnection by transarterial glue embolization or microsurgery [11]. While four of the seven reported cases underwent conservative management for DAVF [6, 8, 11, 12], three were intervened due to severe clinical symptomatology or higher Cognard classification, two of them underwent transvenous embolization and one underwent both transarterial and subsequent transvenous embolization [7, 9, 10].

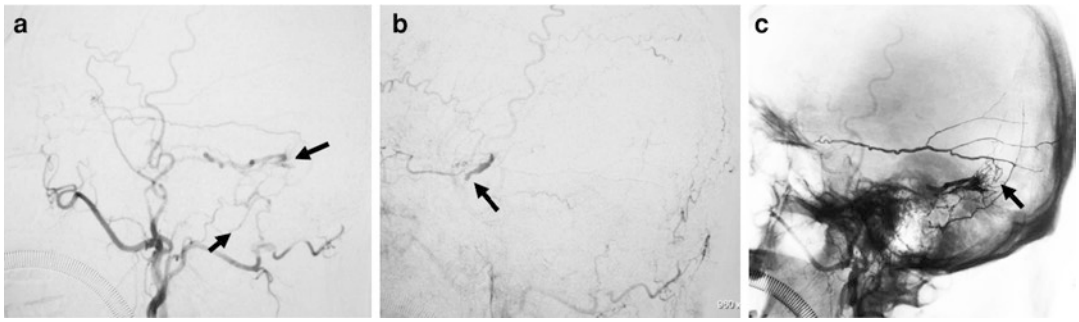

Fig. 14.1 A case of a Borden type-III and Cognard type-III DAVFs involving the transverse sinus co-existed with MMA (arrow in panel **a**). The shunt was obliterated with Onyx injection (arrows in panel **c**). (**a**) Preoperative the left external carotid angiogram. (**b**) Postoperative carotid angiogram showing the obliteration of the distal internal carotid artery and the proximal internal carotid artery was retrogradely filled from the ophthalmic artery (arrow). The DAVF was completely obliterated. (**c**) Postoperative craniogram showing the Onyx casts (arrow)

14.4 Conclusion

MMA can rarely be associated with DAVF during various stages of its disease course. Chronic cerebral ischemia and subsequent angiogenesis might be the missing link between the two conditions. Management of DAVF in the background greatly depends on the symptomatology and invasiveness.

Funding The authors did not receive any funding or sponsorship.

The manuscript has not been published in any preprint server.

Disclosure Dr. Shambaditya Das, (Prof.) Dr. Souvik Dubey, and (Prof.) Dr. Biman Kanti Ray have no disclosures to make.

Authorship Agreement Dr. Shambaditya Das take full responsibility for the data, the analyses and interpretation, and the conduct of the research. I have full access to all the data and have the right to publish any and all data, separate and apart from the guidance of any sponsor.

Conflict of Interest The authors have stated explicitly that there are no conflicts of interest in connection with this article.

References

1. Das DS, Dubey DS, Acharya DM, Ghosh DR, Chatterjee DS, Hazra PA, et al. The disease presentation of Moyamoya angiopathy in Eastern India. J Stroke Cerebrovasc Dis. 2020;29(8):104957.
2. Das S, Dubey S, Acharya M, Chatterjee S, Lahiri D, Das G, et al. Thalassemia and Moyamoya syndrome: unfurling an intriguing association. J Neurol. 2019;266(11):2838–47 [cited 2020 Aug 30]. Available from: https://pubmed.ncbi.nlm.nih.gov/31422456/.
3. Das S, Ray BK, Dubey S. Temporal lobe epilepsy with nocturnal wandering leading to discovery of Moyamoya angiopathy. Acta Neurol Belgica. 2021:1–3 [cited 2021 Dec 2]. Available from: https://link.springer.com/article/10.1007/s13760-021-01830-y.
4. Das S, Ghosh R, Dubey S, Pandit A, Ray BK, Kraemer M. Limb-shaking TIA in Moyamoya angiopathy. Clin Neurol Neurosurg. 2021;207:106783 [cited 2021 Jul 5]. Available from: https://linkinghub.elsevier.com/retrieve/pii/S0303846721003127.
5. Dubey S, Ghosh R, Chatterjee S, Dubey MJ, Ray BK, Das S, et al. Spicy foods triggering clinical symptoms in Moyamoya angiopathy. J Neurosurg Sci. 2021;65(1):85–88 [cited 2021 Mar 18]. Available from: https://pubmed.ncbi.nlm.nih.gov/32550609/.
6. Hou K, Zhao Y, Chen X, Xu K, Yu J. Moyamoya disease concurrent with dural arteriovenous fistula: a case report and literature review. Exp Ther Med. 2020;20(6):161 [cited 2022 Jan 17]. Available from: https://www.ncbi.nlm.nih.gov/pmc/articles/PMC7571339/.
7. Killory BD, Gonzalez LF, Wait SD, Ponce FA, Albuquerque FC, Spetzler RF. Simultaneous unilateral moyamoya disease and ipsilateral dural arteriovenous fistula: case report. Neurosurgery. 2008;62(6):E1375–6 [cited 2022 Jan 17]. Available from: https://pubmed.ncbi.nlm.nih.gov/18824958/.
8. Koduri S, Andrew Wilkinson D, Griauzde JM, Gemmete JJ, Maher CO. Development of bilateral dural arteriovenous fistulae following pial synangiosis for moyamoya syndrome: case report. J Neurosurg Pediatr. 2019;24(1):9–13.
9. Liu AF, Li C, Yu W, Lin LM, Qiu HC, Zhang YQ, Lv XL, Wang K, Liu C, Jiang WJ. Dissection-related

carotid-cavernous fistula (CCF) following surgical revascularization of chronic internal carotid artery occlusion: a new subtype of CCF and proposed management. Chin Neurosurg J. 2020;6:2.
10. Liu P, Xu Y, Lv X, Ge H, Lv M, Li Y. Progression of unilateral moyamoya disease resulted in spontaneous occlusion of ipsilateral cavernous dural arteriovenous fistula: case report. Interv Neuroradiol. 2016;22(3):362–4.
11. Feroze AH, Kushkuley J, Choudhri O, Heit JJ, Steinberg GK, Do HM. Development of arteriovenous fistula after revascularization bypass for moyamoya disease: case report. Oper Neurosurg. 2015;11(1):E202–6.
12. Zaletel M, Surlan-Popović K, Pretnar-Oblak J, Žvan B. Moyamoya syndrome with arteriovenous dural fistula after head trauma. Acta Clin Croatica. 2011;50:115–20 [cited 2022 Jan 17]. Available from: https://pubmed.ncbi.nlm.nih.gov/22034792/.
13. Das S, Ray BK, Ghosh R, Sengupta S, Pandit A, Dubey S. Impact of COVID-19 pandemic in natural course of Moyamoya angiopathy: an experience from tertiary-care-center in India. Egypt J Neurol Psychiatry Neurosurg. 2021;57(1):1–6. Available from: https://ejnpn.springeropen.com/articles/10.1186/s41983-021-00412-2.
14. Das S, Dubey S, Pandit A, Ray BK. Moyamoya angiopathy unmasking systemic lupus erythematosus. BMJ Case Rep. 2021;14(1):e239307 [cited 2021 Jul 5]. Available from: https://casereports.bmj.com/content/14/1/e239307.
15. Mikami T, Suzuki H, Komatsu K, Mikuni N. Influence of inflammatory disease on the pathophysiology of moyamoya disease and quasi-moyamoya disease. Neurol Med Chir (Tokyo). 2019;59(10):361 [cited 2021 Jul 30]. Available from: https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6796064/.
16. Kim JS. Moyamoya disease: epidemiology, clinical features, and diagnosis. J Stroke. 2016;18(1):2–11 [cited 2021 Dec 16]. Available from: https://pubmed.ncbi.nlm.nih.gov/26846755/.
17. Mawad ME, Hilal SK, Michelsen WJ, Stein B, Ganti SR. Occlusive vascular disease associated with cerebral arteriovenous malformations. Radiology. 1984;153(2):401–8 [cited 2022 Jan 17]. https://doi.org/10.1148/radiology15326484172. Available from: https://pubs.rsna.org/doi/abs/10.1148/radiology.153.2.6484172.
18. Mikami T, Suzuki H, Komatsu K, Mikuni N. Influence of inflammatory disease on the pathophysiology of moyamoya disease and quasi-moyamoya disease. Neurol Med Chir (Tokyo). 2019;59(10):1–10 [cited 2020 Aug 30]. Available from: https://www.ncbi.nlm.nih.gov/pmc/articles/PMC6796064/.