www.surgicalneurologyint.com

ScientificScholar®
Knowledge is power
Publisher of Scientific Journals

Surgical Neurology International

Editor-in-Chief: Nancy E. Epstein, MD, Clinical Professor of Neurological Surgery, School of Medicine, State U. of NY at Stony Brook.

SNI: Neuroanatomy and Neurophysiology

Editor
Seyed Ali Khonsary, MD
University of California at Los Angeles, Los Angeles, CA, USA

SNI
Open Access

Review Article

Extracranial carotid localized fibromuscular dysplasia: A case report and literature review

Mustafa Ismail[1], Teeba A. Al-Ageely[1], Mohamed Almustafa A. Alzerkani[1], Younus M. Al-Khazaali[1], Haneen A. Salih[2], Aktham O. Al-Khafaji[1], Zahraa M. Kareem[1], Alkawthar M. Abdulsada[3], Hayder R. Salih[4], Samer S. Hoz[5]

[1]Department of Neurosurgery, University of Baghdad, College of Medicine, [2]Department of Biology, University of Al-Mustansiriyah, College of Sciences, Baghdad, Iraq, [3]Department of Neurosurgery, Azerbaijan Medical University, Baku, Azerbaijan, [4]Department of Neurosurgery, Neurosurgical Teaching Hospital, Baghdad, Iraq, [5]Department of Neurosurgery, University of Cincinnati, Cincinnati, United States.

E-mail: Mustafa Ismail - mustafalorance2233@gmail.com; Teeba A. Al-Ageely - teebaalageely@gmail.com; Mohamed Almustafa A. Alzerkani - mohamedalmustafaabbas@gmail.com; Younus M. Al-Khazaali - ymjr.iq@gmail.com; Haneen A. Salih - haneenalaa353@gmail.com; Aktham O. Al-Khafaji - akthamalkhafaji@gmail.com; Zahraa M. Kareem - zahraa1999majeed@gmail.com; Alkawthar M. Abdulsada - alkawthar.alhchaimi@gmail.com; Hayder R. Salih - hayderneurosurg85@gmail.com; *Samer S. Hoz - hozsamer2055@gmail.com

***Corresponding author:**
Samer S. Hoz,
Department of Neurosurgery,
University of Cincinnati,
Cincinnati, United States.
hozsamer2055@gmail.com

Received : 10 October 2022
Accepted : 14 October 2022
Published : 28 October 2022

DOI
10.25259/SNI_937_2022

Quick Response Code:

ABSTRACT

Background: Fibromuscular dysplasia (FMD) is a noninflammatory and nonatherosclerotic arteriopathy that is characterized by irregular cellular proliferation and deformed construction of the arterial wall that causes segmentation, constriction, or aneurysm in the intermediate-sized arteries. The incidence of FMD is 0.42–3.4%, and the unilateral occurrence is even rarer. Herein, we report a rare case of a localized extracranial carotid unilateral FMD associated with recurrent transient ischemic attacks (TIAs) treated by extracranial-intracranial bypass for indirect revascularization. The specific localization of the disease rendered our case unique.

Methods: We conducted a review of the PubMed Medline database search using the following combined formula: ((FMD [Title/Abstract]) AND ((isolated [Title/Abstract]) OR (localized [Title/Abstract]))) AND Internal carotid artery (ICA) (Title/Abstract). Additional resources were included by screening the reference list of the selected papers.

Results: A total of six cases were found, and all accounted for localized FMD affecting the ICA. The age range was between 19 and 52, the male-to-female ratio was (2:4), and all of the cases consisted of unilateral carotid FMD, mainly on the left side with a left-to-right ratio of 5:1. The management and outcome of these cases varied according to the case and associated complications.

Conclusion: Extracranial localized FMD of the ICA is a rare subtype of FMD that has little documentation in the literature. In our case, it was a localized extracranial carotid unilateral FMD associated with recurrent TIAs. The appropriate treatment was using the intracranial-extracranial bypass.

Keywords: Encephalo-duro-arterio-myo-synangiosis, Extracranial – intracranial bypass, Fibromuscular dysplasia

INTRODUCTION

Fibromuscular dysplasia (FMD) is a noninflammatory and nonatherosclerotic arteriopathy that is identified by irregular cellular reproduction and deformed construction of the arterial wall, causing segmentation and constriction (with either central stenosis or the more common beads figure), or aneurysm of intermediate-sized arteries.[6] The cervicocephalic arteries represent the

second most frequent site for FMD (25–%), following the renal artery (60–75%).[3,10]

The incidence of FMD is approximately (0.42–3.4%) and its unilateral occurrence is even rarer.[7] The middle one-third of the carotid arteries is included in over 90% of cases of cervicocephalic FMD.[10] The second most common location is extracranially-located FMD which mainly occurs bilaterally with narrowing at C1 and C2 levels.[2] FMD is usually diffuse and most commonly affects the vertebral arteries and middle and distal portions of the internal carotid artery (ICA) and, occasionally, intracranial arteries.[7] Only 9% of FMD cases affect the extracranial ICA or vertebral arteries.[20]

Most patients with FMD are asymptomatic, and the lesion is discovered incidentally. Some patients have a myriad of symptoms, including headaches, transient ischemic attacks (TIAs), and even stroke.[22] Management focuses on symptomatic relief. Surgical management should be attempted for symptomatic patients or patients who have not responded to medications.[18] Three similar cases with the same management as ours are found in the literature regarding extracranial-intracranial (EC-IC) bypass for extracranial FMD of the ICA.[11,21,23]

In this paper, the authors report a case of a 53-year-old male with focal extracranial carotid unilateral FMD associated with recurrent TIAs. The uncommon localization of the stenosis made the approach seemingly suitable for this case.

CASE DESCRIPTION

A 53-year-old male was referred to Neurosurgical Teaching Hospital in Baghdad, Iraq, with a history of recurrent TIAs with the left upper limb weakness grade 3/5. Magnetic resonance imaging [Figure 1] revealed periventricular hemorrhage along with unilateral cerebrospinal fluid -filled cleft in the right hemisphere extending from the frontal horn of the lateral ventricle to the Sylvian fissure. Diagnostic cerebral angiography catheterization [Figure 2] of the right ICA showed significant stenotic segments of the cervical ICA, and a decision for endovascular stenting was made. During therapeutic catheterization angiography, the stenosis was seen up to the intracranial portion of the ICA, giving a "beads-on-a-string" sign. A final diagnosis of focal FMD of the right extracranial ICA was made. In addition, the coexistence of a hypodynamic circulation due to a weak middle cerebral artery (MCA) blood flow shifted the treatment plan to an EC-IC bypass.

The patient underwent encephalo-duro-arterio-myo-synangiosis (EDAMS) surgery under intraoperative neurophysiological monitoring, including somatosensory evoked potentials and motor evoked potentials. Postoperatively, the patient was conscious with stable vital signs and left-sided weakness grade 3/5, which is the same as

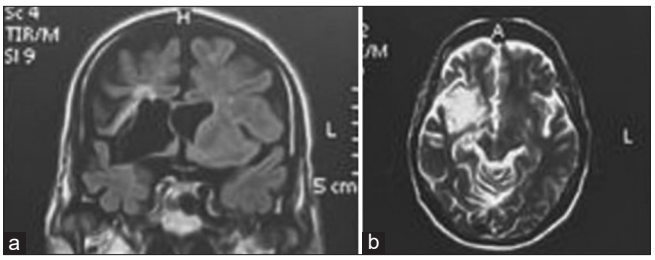

Figure 1: (a) Preoperative T1-weighted magnetic resonance imaging (MRI) image (coronal view) showing periventricular old ischemia alongside cerebospinal fuild (CSF)-filled cleft in the right-sided hemisphere. (b) Preoperative T2-weighted MRI image (axial view) revealing periventricular longstanding ischemic changes and unilateral CSF-filled cleft in the right hemisphere extending from the frontal horn of the lateral ventricle to the Sylvian fissure.

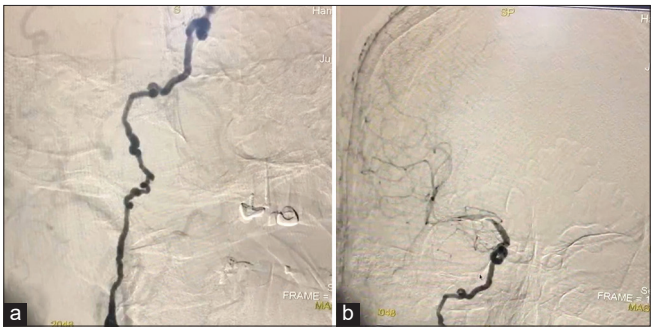

Figure 2: (a) Preoperative digital subtraction angiography demonstrating fibromuscular dysplasia-related multifocal stenosis along the right extracranial internal carotid artery (ICA). (b) Preoperative digital subtraction angiography demonstrating the "beads-on-a-string" sign appearance along the right extracranial ICA.

preoperatively. A postoperative CT scan revealed no signs of new ischemia or stroke, and the patient was discharged after 5 days. Six months of follow-up revealed left-sided weakness Grade 4 with no new findings on imaging.

MATERIALS AND METHODS

We conducted a PubMed Medline database search using the following combined formula: ((FMD [Title/Abstract]) AND ((isolated [Title/Abstract]) OR (localized [Title/Abstract])))) AND ICA [Title/Abstract]. Additional resources were obtained by screening the reference list of the selected articles.

RESULTS

A total of 16 results were found, eight of them were case reports, and only six of them accounted for localized FMD affecting the ICA. According to the reported cases, the age range was between 19 and 52, the male-to-female ratio was 2:4, and all of the cases consisted of unilateral carotid FMD, with the majority on the left side with a left-to-right ratio of

(5:1). The management and outcome of these cases differed according to the case [Table 1].[4,5,8,16,17,19]

DISCUSSION

FMD is a rare nonatherosclerotic and noninflammatory vasculopathy resulting in arterial narrowing and the potential formation of small- and medium-sized vessel aneurysms.[14,15] FMD is usually diffuse and commonly involves the vertebral arteries and middle and distal portions of the ICA, and occasionally, intracranial arteries.[24] Localized FMD is rare in the literature; however, there are few reports of isolated intracranial FMD affecting the ICA, basilar artery, ACA, MCA, and PCA.

Extracranial localized FMD cases also have few reports in the literature.[13] FMD is classified into four main angiographical categories. They include the multifocal type, the most common type displaying the "beads-on-a-string" appearance due to multiple stenoses. Unifocal FMD has long concentric stenosis giving it the "tubular" shape, while short-isolated segments of stenosis characterize the focal type. Finally, up to 20% of patients have mixed-type stenosis.[14] In our case, the patient had localized extracranial carotid artery FMD of a multifocal subtype giving the string of beads appearance.

Because many patients are asymptomatic, it can be found incidentally on imaging or physical examination while investigating for any cervical bruit.[1,12] Such lesions can result in rare but severe complications in approximately 10% of cases, such as decreased cerebral perfusion due to the narrowing of the lumen of the affected vessels leading to TIAs, and thrombus formation, which may result in distal embolization, dissection, and rupture.[24]

Management of FMD depends mainly on the clinical manifestations and the pertinent anatomy of the artery involved. Conservative management of asymptomatic carotid artery FMD is advised, with monitoring and anticoagulant therapy. When FMD results in the occlusion of the artery by thrombus formation, dual antiplatelet therapy is used along with anticoagulant medications to prevent ischemic injury. Revascularization with a bypass is an option for a good outcome with fine patency of blood flow intracerebrally.[12]

Interventional management is usually indicated in patients with symptomatic extracranial carotid FMD. This includes surgical dilatation with or without endarterectomy for the carotid arteries. The most commonly used option is percutaneous transluminal balloon angioplasty with or without the application of stents or cerebral protection devices.[1,24] This method prevents ischemia and neurological deficits in patients with recurrent symptoms despite medical therapy.[1] Microsurgical clipping and endovascular therapy by stenting or coiling are reserved for cases complicated by aneurysms.[15]

In the present case, the localized peculiarity of the stenosis is similar in concept to Moyamoya disease (MMD), another vasculopathy characterized by progressive bilateral stenosis, and occlusion of the supraclinoid ICA leading to the development of collateral vessels. Diverse surgical techniques have been performed to manage MMD to avoid ischemic injury and such techniques may be employed in managing localized FMD of the ICA. The most commonly used methods include direct revascularization by superficial temporal artery to MCA bypass and indirect revascularization methods such as encephalo-myo-synangiosis, encephalo-duro-arterio-synangiosis, and EDAMS.[9]

Table 1: Reported cases of localized FMD in the ICA, the selected treatment options, and the outcome.

Author/year	Age/gender	Site	Management	Outcome
Schievink *et al*. 1994[17]	26/Female	Left ICA* FMD** with dissection	-	Poor/died due to thoracoabdominal aortic dissecting aneurysm
Escamilla *et al*. 2000[8]	37/Female	Left extracranial ICA proximal occlusion of the ACA*** and MCA****	-	-
Birnbaum *et al*. 2005[5]	19/Female	Proximal ACA and MCA	-	-
Poppe *et al*. 2007[16]	52/Male	Right ICA with dissection	Aspirin and clopidogrel	Good
Spengos *et al*. 2008[19]	50/Male	Left distal extracranial and proximal intracranial segments of ICA	Antiplatelet agents	Good
Bilman *et al*. 2021[4]	44/Female	Left ICA FMD with dissection	Bypass	Good
Ismail *et al*. 2022 (Current study)	53/Male	Right cervical ICA	Bypass with EDAMS*****	Good

*ICA: Internal Carotid Artery, **FMD: Fibromuscular dysplasia, ***ACA: Anterior cerebral artery, ****MCA: Middle cerebral artery, *****EDAMS: Encephalo-duro-arterio-myo-synangiosis

We reviewed the literature for the reported cases of localized FMD involving the ICA. Only six cases consisted of localized carotid artery FMD; almost all of these cases involved the left-sided ICA. Treatment choices are individualized to each case, and with the evolving capabilities of endovascular procedures, outcomes for symptomatic FMD may vary significantly.

In the present case, the patient had localized FMD of the right extracranial ICA. The uncommon localization of the stenosis with the coexistence of low-flow MCA made the approach seemingly suitable for this case. Indirect revascularization by EDAMS was performed to prevent further ischemic attacks. Management of FMD varies due to the clinical manifestations observed. The surgeon's decision for the operative intervention method differs according to the case. Suggestions for managing localized arterial stenosis in FMD of the extracranial ICA are minimal and rarely reported.

CONCLUSION

The scarce nature of craniocerebral FMD poses particular diagnostic and therapeutic challenges. Traditional surgical and endovascular management options exist, but individualized planning is essential with each case, especially with ICA occlusion or weak flow. In our case, the localized stenosis of the extracranial ICA is a rare subtype of FMD, rendering our case unique since isolated FMD involving the ICA has only a few documents in the literature.

Declaration of patient consent

Patients' consent not required as patients' identities were not disclosed or compromised.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest

REFERENCES

1. Al-Nouri O, Pan J, Mannava K. Endovascular treatment of concomitant carotid fibromuscular dysplasia and atherosclerotic disease after failed open surgical treatment. Vasc Endovasc Surg 2019;53:250-4.
2. Bartels E, Bartels S, Poppert H. Diagnosis of non-atherosclerotic carotid disease. Perspect Med 2012;1:244-9.
3. Begelman SM, Olin JW. Fibromuscular dysplasia. Curr Opin Rheumatol 2000;12:41-7.
4. Bilman V, Apruzzi L, Baccellieri D, Sanvito F, Bertoglio L, Chiesa R. Symptomatic internal carotid artery dissection and kinking in a patient with fibromuscular dysplasia. J Vasc Bras 2021;20:e20200243.
5. Birnbaum LA, Sherry R, Pereira E. Isolated intracranial fibromuscular dysplasia presents as stroke in a 19-year-old female. Arq Neuropsiquiatr 2005;63:318-20.
6. Brinza EK, Gornik HL. Fibromuscular dysplasia: Advances in understanding and management. Cleve Clin J Med 2016;83:S45-51.
7. Cardounell SZ, Gonzalez L. Carotid Artery Fibromuscular Dysplasia. Treasure Island, FL: StatPearls; 2021.
8. Escamilla F, Espigares A, Hervas R, Fernandez MD, Vela R, Garcia T. Fibromuscular dysplasia with moyamoya phenomenon in a patient with Alport's syndrome. A Type IV collagen disorder. Rev Neurol 2000;30:736-40.
9. Isono M, Ishii K, Kamida T, Inoue R, Fujiki M, Kobayashi H. Long-term outcomes of pediatric moyamoya disease treated by encephalo-duro-arterio-synangiosis. Pediatr Neurosurg 2002;36:14-21.
10. Leary MC, Finley A, Caplan LR. Cerebrovascular complications of fibromuscular dysplasia. Curr Treat Options Cardiovasc Med 2004;6:237-48.
11. Lopez-Gonzalez MA, Zhao X, Ramanathan D, Eastin TM, Minwoo S. High flow bypass for right giant cavernous internal carotid artery aneurysm with fibromuscular dysplasia of cervical internal carotid artery: Microsurgical 2-D video. Surg Neurol Int 2020;11:177.
12. Maramattom BV, Thomas J, Panicker D, Gopalakrishnan CV. Extensive fibromuscular dysplasia in a young girl treated with bilateral STA-MCA bypass. Neurol India 2022;70:792-3.
13. McDonald A. A case of isolated intracranial fibromuscular dysplasia. Cureus 2020;12:e8755.
14. Olin JW, Pierce M. Contemporary management of fibromuscular dysplasia. Curr Opin Cardiol 2008;23:527-36.
15. Plouin PF, Perdu J, La Batide-Alanore A, Boutouyrie P, Gimenez-Roqueplo AP, Jeunemaitre X. Fibromuscular dysplasia. Orphanet J Rare Dis 2007;2:28.
16. Poppe AY, Minuk J, Glikstein R, Leventhal M. Fibromuscular dysplasia with carotid artery dissection presenting as an isolated hemianopsia. J Stroke Cerebrovasc Dis 2007;16:130-4.
17. Schievink WI, Björnsson J, Piepgras DG. Coexistence of fibromuscular dysplasia and cystic medial necrosis in a patient with Marfan's syndrome and bilateral carotid artery dissections. Stroke 1994;25:2492-6.
18. Sharma AM, Kline B. The United States registry for fibromuscular dysplasia: New findings and breaking myths. Tech Vasc Interv Radiol 2014;17:258-63.
19. Spengos K, Vassilopoulou S, Tsivgoulis G, Papadopoulou M, Vassilopoulos D. An uncommon variant of fibromuscular dysplasia. J Neuroimaging 2008;18:90-2.
20. Stanley JC, Gewertz BL, Bove EL, Sottiurai V, Fry WJ. Arterial fibrodysplasia: Histopathologic character and current etiologic concepts. Arch Surg 1975;110:561-6.
21. Stewart MT, Moritz MW, Smith RB 3rd., Fulenwider JT, Perdue GD. The natural history of carotid fibromuscular dysplasia. J Vasc Surg 1986;3:305-10.
22. Subramanian A, Aggarwal G, Agarwal D, Lalwani S. Internal carotid artery fibromuscular dysplasia in a child: Incidental

postmortem finding after head injury. J Lab Physicians 2017;9:60-3.
23. Takenaka K, Shimizu K, Ando T, Sakai N, Yamada H. Fibromuscular dysplasia with cerebral infarction in children case report. Neurol Med Chir (Tokyo) 1989;29:912-6.
24. Touzé E, Oppenheim C, Trystram D, Nokam G, Pasquini M, Alamowitch S, *et al*. Fibromuscular dysplasia of cervical and intracranial arteries. Int J Stroke 2010;5:296-305.

How to cite this article: Ismail M, Al-Ageely TA, Alzerkani MA, Al-Khazaali YM, Salih HA, Al-Khafaji AO, *et al*. Extracranial carotid localized fibromuscular dysplasia: A case report and literature review. Surg Neurol Int 2022;13:498.