









syndrome, transient ischemic attack, ischemic stroke, or may be associated with intracerebral aneurysms with a risk of subarachnoid or intracerebral hemorrhage. The presence of both hemorrhage due to aneurysm rupture and ischemic stroke due to stenosis is characteristic of FMD [2].

For diagnosis, echo-Doppler may reveal irregular stenosis compatible with the diagnosis. However, CT and MRI angiography perform better, particularly because FMD usually affects the middle and distal portions of the carotid and vertebral arteries, which are less accessible with Doppler ultrasound [9]. CT and MRI angiography also have the advantage of detecting associated intracranial aneurysms [6]. Three angiographic aspects are described. The first is multifocal FMD, characterized by alternating stenoses and dilatations, giving the appearance of a “string of beads”. This is the most common aspect of cervical FMD, observed in 80% of cases. The second aspect, characterized by unifocal lesions, which account for approximately 7% of cases and can be challenging to differentiate from atherosclerosis, carotid hypoplasia, or certain types of vasculitis. The third aspect includes essentially atypical forms, characterized by the appearance of a fibrous septum, positioned diaphragm-like at the origin of the internal carotid artery. This form of FMD is rare (4% of cases), and more frequently observed in black patients, histologically corresponding to intimal FMD [3].

Conventional angiography remains the gold standard examination and the technique with the highest spatial resolution for comprehensive lesion assessment in fibromuscular dysplasia, especially for minor lesions or intracranial involvement [10]. However, this technique carries the risk of dissection or embolic complications.

The main differential diagnosis is atherosclerotic disease. In the case of atherosclerosis, lesions typically affect the origin or proximal portions of arteries in older patients with one or more cardiovascular risk factors [7]. Other potential differential diagnoses include systemic vasculitis affecting large- or medium-sized arteries (particularly Takayasu’s disease) or other primary connective tissue diseases affecting arterial walls, such as Marfan syndrome, Ehlers-Danlos syndrome, neurofibromatosis type 1, and so on [5].

In the case of asymptomatic fibromuscular dysplasia (FMD), the consideration of preventive antiplatelet treatment can be discussed [11]. Antithrombotic medications are typically used in patients who have experienced an ischemic stroke, although their effectiveness has not been specifically demonstrated in this context [4].

For cases of acute cervical dissection or meningeal hemorrhage resulting from aneurysm rupture, treatment does not deviate from the general recommendations. Surgical or endovascular revascularization is not recommended for asymptomatic cervical FMD patients. However, in certain individuals with hemodynamic complications, recurrent ischemic strokes, or debilitating pulsatile tinnitus, angioplasty (with or without stenting) may be considered, despite limited literature on this topic [10,12]. The indication for surgical or endovascular treatment of an intracranial aneurysm does not differ from the general approach for these two conditions [13].

### **Implications**

By presenting this case, we aim to raise awareness among healthcare professionals about the varied presentations and localizations of fibromuscular dysplasia. Early recognition and accurate diagnosis of this condition can guide appropriate management strategies and prevent potential complications. Additionally, our report highlights the potential risks associated with conventional angiography, which should be carefully considered in the diagnostic workup.

### **Limitations of the Study**

This case report represents a single patient’s experience and may not be generalizable to all cases of fibromuscular dysplasia. Furthermore, the limitations of retrospective data analysis and the inherent biases associated with a single case study should be acknowledged. Further research and larger studies are needed to validate our findings and explore optimal management strategies for fibromuscular dysplasia.

## Conclusion

Fibromuscular dysplasia (FMD) is a rare vascular disease characterized by abnormalities in the arterial wall, predominantly affecting young women. It can lead to various manifestations such as stenosis, occlusion, aneurysms, or dissection in the middle arteries. All arterial territories can be involved, with renal and cervical arteries being the most commonly affected. Different angiographic presentations exist, including uni-focal or multifocal stenosis, each with its own prognosis. The clinical presentation of FMD is nonspecific and highly heterogeneous, with many patients being asymptomatic or having mild symptoms. The current management approach primarily focuses on medical and endovascular treatments for symptomatic patients.

## Bibliography

1. C Vaisseaux and I Poumons. "Le point sur la dysplasie fibromusculaire". *Cardiologie* 22.2 (2017): 16-20.
2. PF Plouin., *et al.* "Fibromuscular dysplasia". *Orphanet Journal of Rare Diseases* 2.1 (2007): 1-8.
3. M Pasquini., *et al.* "Dysplasie fibromusculaire cervicale et intracrânienne". *La Press Medicale* 40.7-8 (2011): 713-719.
4. E Touzé., *et al.* "Fibromuscular dysplasia of cervical and intracranial arteries". *International Journal of Stroke* 5.4 (2010): 296-305.
5. AC Desbois., *et al.* "Fibromuscular dysplasia". *Revue de Médecine Interne* 36.4 (2015): 271-276.
6. A Persu., *et al.* "Diagnosis and management of fibromuscular dysplasia: An expert consensus". *European Journal of Clinical Investigation* 42.3 (2012): 338-347.
7. A La Batide Alanore., *et al.* "Fibromuscular dysplasia". *La Presse Médicale* 36.6 II (2007): 1016-1023.
8. G Wuerzner., *et al.* "Hypertension et dysplasie fibromusculaire: Au-delà des artères rénales". *Revue Médicale Suisse* 13.574 (2017):1580-1583.
9. LR Caplan. "Fibromuscular dysplasia". *Uncommon Causes Stroke*, 2<sup>nd</sup> Edition (2008): 491-496.
10. S Lenck., *et al.* "Diaphragms of the carotid and vertebral arteries: An under-diagnosed cause of ischaemic stroke". *European Journal of Neurology* 21.4 (2014): 586-593.
11. JW Olin., *et al.* "Fibromuscular dysplasia: State of the science and critical unanswered questions: A scientific statement from the American heart association". *Circulation* 129.9 (2014): 1048-1078.
12. A Persu., *et al.* "European consensus on the diagnosis and management of fibromuscular dysplasia". *Journal of Hypertension* 32.7 (2014): 1367-1378.
13. E Touzé., *et al.* "Cervical and intracranial fibromuscular dysplasia: clinical presentations, diagnosis, prognosis, and management". *Bulletin de l'Académie Nationale de Médecine* 201.7-9 (2017): 1103-1112.

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