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Case Report

Intriguing Vascular Enigma Unveiled: Chronic Headache in a Young Patient Revealing Incidental Fibromuscular Angiodysplasia

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Abstract

We are reporting the case of a young 23-year-old patient with chronic treatment-resistant cephalalgia. The CT scan revealed abnormal arteries related to a fortuitous discovery of fibromuscular dysplasia with no sign of complications. Fibromuscular dysplasia is characterized as a segmental, non-atheromatous disease of the arterial wall that leads to stenosis of medium-sized arteries. The renal artery is the most frequently affected, with a prevalence ranging from 60% to 100%. Involvement of the carotid or vertebral arteries is less common, occurring in approximately 10% to 35% of cases. This case is an illustration of an uncommon localization of this rare arterial pathology that emphasizing the significant role of radiology in its diagnosis and management highlighting the risks associated with conventional angiography.

Keywords: *Fibromuscular Dysplasia; Cerebral Arteries; CT-Angiography, Diagnostic Imaging, Case Report*

Introduction

Fibromuscular dysplasia is a rare idiopathic non-atherosclerotic and non-inflammatory segmental disease of the muscular arterial wall, leading to stenosis of medium arteries [1]. It affects primarily women between 30 and 50 years old. All arterial territories can be affected; however, the renal and cervical arteries are the most frequently involved. It is often asymptomatic but it may be discovered when complications occur [2]. The etiology of fibromuscular dysplasia is currently unknown.

Conventional angiography remains the gold standard examination due to its high spatial resolution, providing a comprehensive assessment of fibromuscular dysplasia lesions, particularly in cases of minor lesions or intracranial involvement [10]. However, it is important to note that this technique carries inherent risks. As a result, CT and MRI angiography have emerged as safer alternatives for exploration, and a preferable choice for exploration due to their lower risk of complications.

Case Report

Our case is a 23-year-old male with no medical history coming to emergencies with chronic headache that did not respond to medication. Physical and neurological examinations were normal with no deficit or any other associated sign. A brain CT scan was performed, revealing spontaneously hyperdense appearance of the left internal intracranial carotid artery and homolateral posterior cerebral artery. Contrast injection demonstrated a tortuous and dilated left internal carotid artery, extending from its cervical portion to the first segment of the left middle cerebral artery M1 as well as the posterior cerebral artery of the same side. A developed arterial circulation was found in the posterior arterial network. There was no sign of bleeding. Conventional angiography was performed to search for small and peripheral aneurysms, revealing many vascular loops and ecstasy of the left internal carotid artery as well as the first portion of the middle cerebral artery. An alternating of stenosis and dilatation was found in the posterior cerebral circulation in the same side, causing a string of breads appearance. No aneurysm, dissection or subarachnoid hemorrhage was found. Based on these findings, the diagnosis of fibromuscular dysplasia was established.

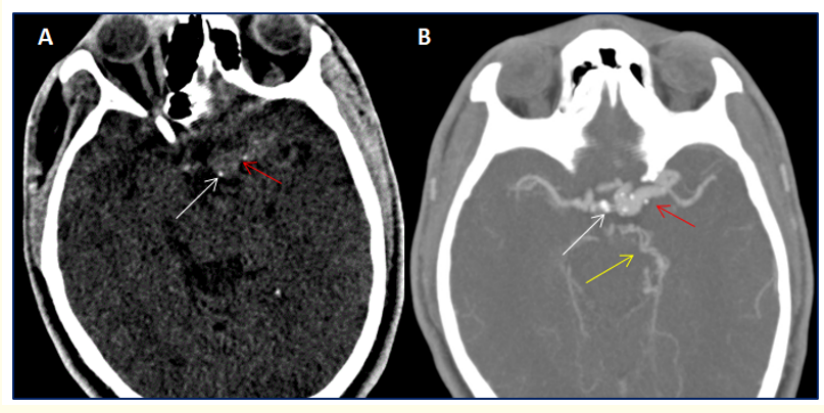

Figure 1: *(A) Axial CT sections shows a spontaneously hyperdense appearance of the internal carotid artery and the proximal portion of the middle cerebral artery (Red arrow) containing small calcifications (White arrow), revealing after contrast injection. (B) an enlargement and twisted appearance of these arteries (Red arrow) as well as of the homolateral posterior cerebral circulation (Yellow arrow).*

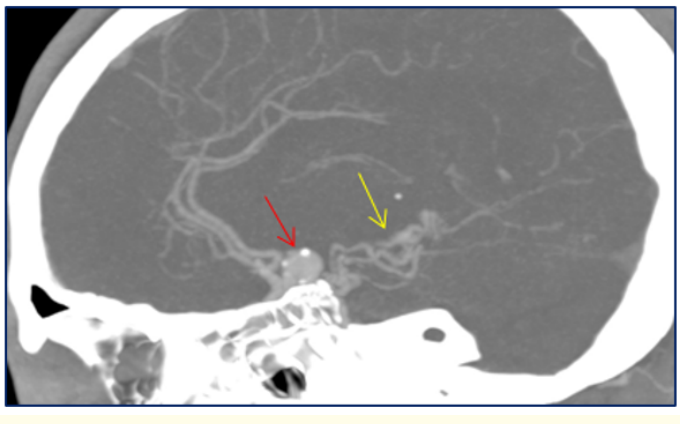

Figure 2: *Oblique CT scan slice demonstrating ectatic left internal carotid (Red arrow) with tortuous path of posterior cerebral artery (Yellow arrow).*

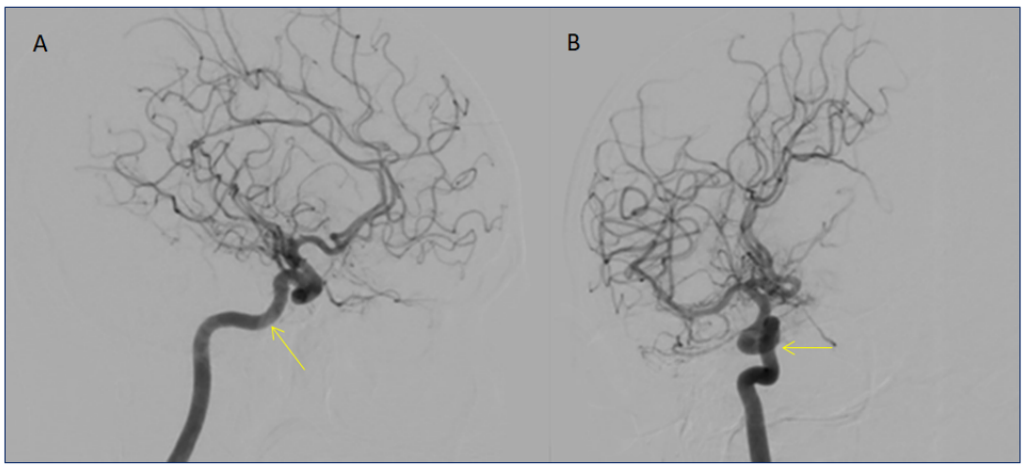

Figure 3: *Front (B) and side (A) view opacification angiography of the right carotid artery (Yellow arrow) revealing no abnormalities of the cervical and intracranial arteries.*

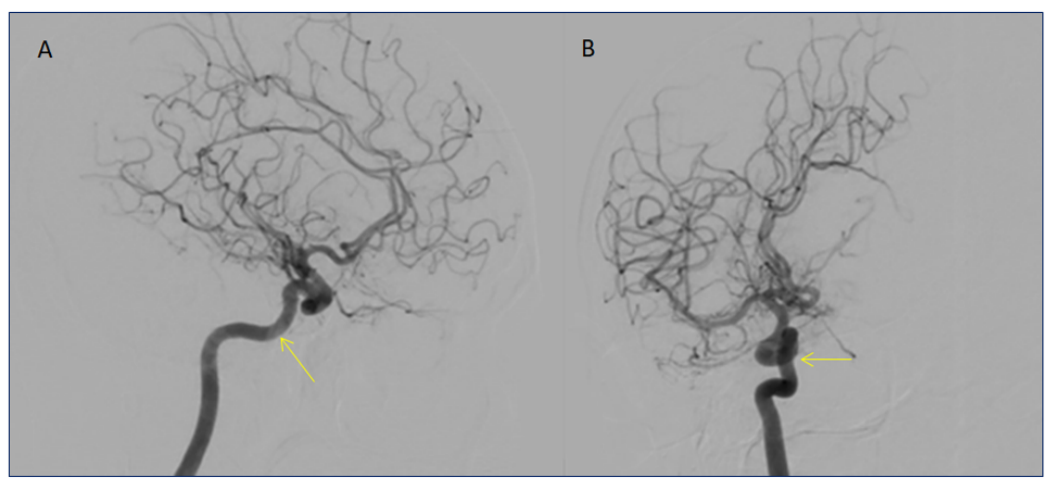

Figure 4: *Front (A) and oblique (B) view of angiography opacification of the left vertebral artery showing a twisted and enlarged segmental aspect of the left posterior cerebral artery with no evident aneurysm detected.*

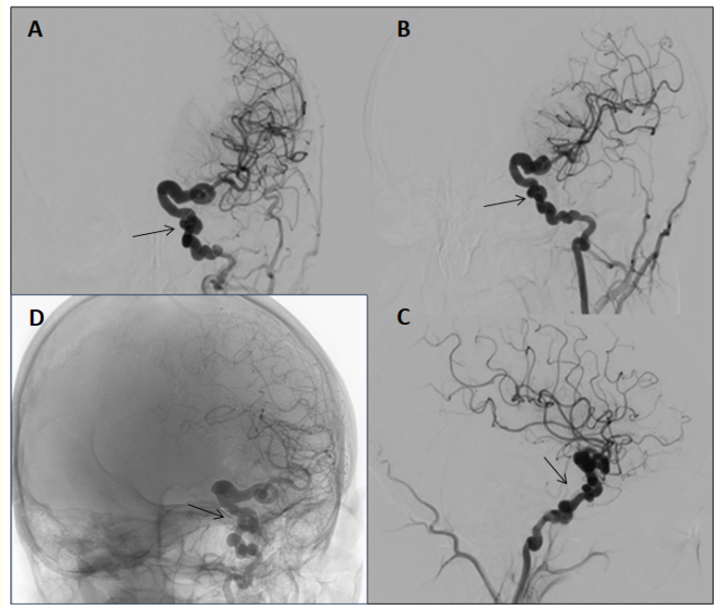

Figure 5: *Opacification angiography of the left carotid artery with frontal (A), oblique (B) and profile (C) views, revealing a widening and twisting pattern of the internal carotid artery from its cervical portion extending to its termination. No specific image indicative of an aneurysm was detected during the examination.*

Discussion

Fibromuscular dysplasia (FMD) is an idiopathic, segmental, non-atherosclerotic and non-inflammatory disease that affects the muscular layer of the arterial wall. It is more common in young and middle-aged women, although it can also occur in men at any age [1]. The etiology of FMD remains unknown. Genetic factors seem to be involved [2], the disease more common in younger women, suggesting a potential involvement of hormonal factors, although the mechanisms are not fully understood. smoking is more prevalent in patients with FMD and appears to be associated with a more severe form of the disease, but the underlying mechanisms of this association are unknown [3]. Mechanical factors may also contribute to the etiology of FMD [4].

Renal artery involvement is the most frequent (60 to 100%) and presents as stenosis of the renal arteries, which can be asymptomatic or complicated by hypertension or impaired renal function. Carotid or vertebral artery involvement is less common (10 to 35%) [5]. The lower frequency of cervical and intracranial fibromuscular dysplasia compared to renal FMD may be explained by the fact that renal FMD is often symptomatic due to arterial hypertension [4]. Involvement of other arterial territories is even rarer, such as mesenteric involvement found in 5 to 26% of patients [6].

Symptoms suggestive of FMD are nonspecific. Depending on the affected arterial territory and the severity of fibromuscular dysplasia, symptoms can vary widely, ranging from simple headaches to ischemic or hemorrhagic stroke with permanent neurological deficits. Headache is present in 60% of patients with cerebrovascular FMD [5,7,8]. Complications of cervicocranial FMD may manifest as Horner's

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.

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