

## Intracerebral fibromuscular dysplasia presenting as a transient ischemic attack: A case report

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### Abstract

**Background:** Fibromuscular dysplasia is a nonatherosclerotic, noninflammatory systemic arterial disease primarily affecting the renal and carotid arteries. Prevalence is rare and is estimated to be seen in less than 0.4% of the population. Most common initial presentations include hypertension, renovascular or cervical bruit, and headache. Prognosis is favorable but is dependent on diagnosis and treatment of the clinical manifestations of the affected vasculature.

**Case Report:** Patient is a 60 year old African American man with a past medical history of multiple sclerosis, hypertension, lumbar and cervical spinal stenosis, type II diabetes, and chronic obstructive pulmonary disease who presented to the emergency department with neurologic signs suggestive of a stroke. Computed tomography and magnetic resonance imaging were negative for acute stroke. However, CT angiography showed signs of luminal irregularities in the internal carotid arteries bilaterally suggestive of fibromuscular dysplasia. This is a patient with significant comorbidities, many of which can cause neurologic deficits. Possible etiologies included multiple sclerosis flare, vasculitis, spinal radiculopathy, or conversion disorder. Finally, a conventional angiogram of the head, neck, and abdomen confirmed fibromuscular dysplasia of the internal carotid arteries with diffuse extension into the intracerebral vasculature.

**Conclusion:** This was an interesting case where the etiology of the patient's acute and inconsistent neurologic deficits was difficult to determine. Final diagnosis is a transient ischemic attack resulting from a new diagnosis of cerebrovascular fibromuscular dysplasia. This is an atypical presentation of fibromuscular dysplasia and one of the few cases showcasing disease extension into the intracerebral vasculature.

## Keywords

Fibromuscular dysplasia; transient ischemic attack; conventional angiography; intracerebral.

## Abbreviations

FMD: Fibromuscular Dysplasia; MS: Multiple Sclerosis; COPD: Chronic Obstructive Pulmonary Disease; OSA: Obstructive Sleep Apnea; CT: Computed Tomography; GCA: Giant Cell Arteritis; TA: Takayasu Arteritis

## Introduction

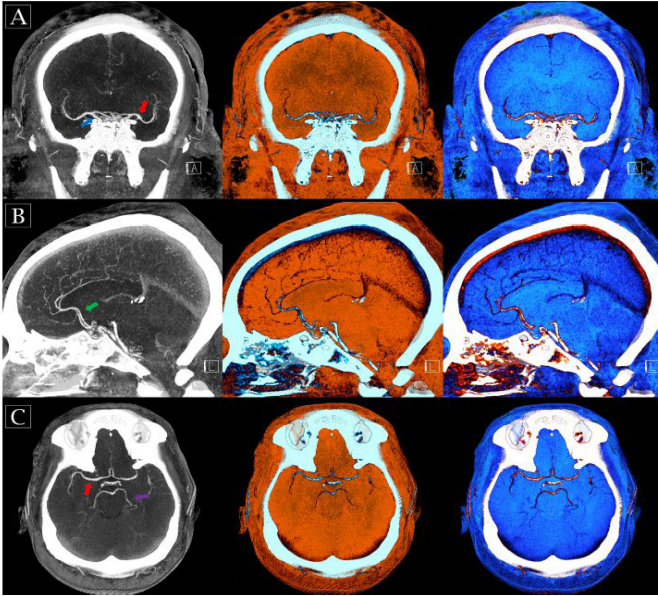
Fibromuscular Dysplasia (FMD) is a rare noninflammatory, nonatherosclerotic vascular disease seen in less than 0.4% of the population that primarily affects medium to small sized arteries [1,2]. FMD is classified into two subtypes, multifocal and focal. Multifocal FMD, which is seen in 90% of patients, is seen as the classic 'string of beads' appearance on the affected arteries. These findings are caused by alternating medial fibrosis and arterial dilation through multiple arterial segments [3]. Focal FMD, primarily seen in children, is characterized by tubular stenosis isolated to a single arterial segment [4]. Data from the United States FMD Registry estimates approximately 80-90% of patients are Caucasian women and the mean age of diagnosis is 52 years of age [5]. The renal, carotid, and vertebral arteries are most affected, but findings can be seen in any arterial bed. Renal artery involvement was seen in approximately 75% of patients, with renovascular hypertension as the most common presenting symptom [6]. Cerebrovascular FMD most commonly involves the carotid arteries bilaterally (95%) and the vertebral artery (60 - 85%) [7]. Most cases are asymptomatic, but common manifestations include headache, carotid bruit, and pulsatile tinnitus. Common complications of cerebrovascular FMD are transient ischemic attack (TIA), ischemic stroke, and arterial dissection or aneurysm. Treatment is primarily focused on preventing thromboembolic complications, arterial dissection and aneurysm rupture through hypertension control and antiplatelet therapy [6]/.

## Case Report

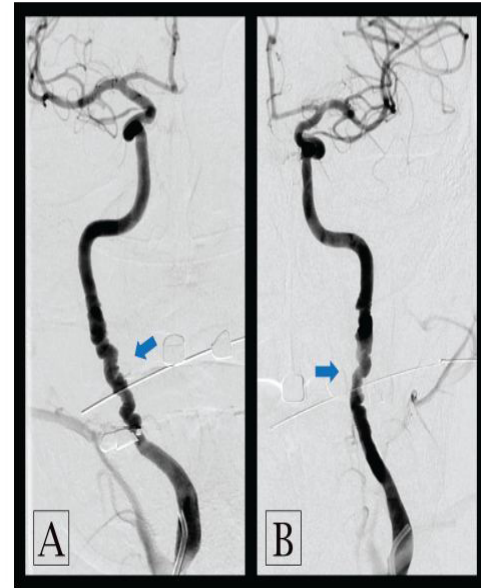
This patient is a 60 year old African American man with a past medical history of Multiple Sclerosis (MS), hypertension, type II diabetes (A1C 6.4), cervical and lumbar spinal stenosis, chronic obstructive pulmonary disease (COPD), and obstructive sleep apnea (OSA) who presented with acute neurologic dysfunction concerning for a stroke. He described waking up in a normal state of health for his daily exercise, returned to sleep, and was awoken two hours later by symptoms of right sided weakness, slurred speech, and tingling of the tongue/lips/mouth. Later he developed blurry vision in the right eye, tingling on left side of his face, and complained of shooting pains down his right arm and leg. Additionally, patient had also presented to the ED two weeks prior for multiple unrelated complaints including chest pain, abdominal pain, and gait instability resulting in a negative overall workup. In this instance, he promptly presented to the emergency department and was treated as a Stroke Alert.

Computed tomography (CT) without contrast was negative for intracranial bleed and MRI was negative for acute stroke. However, CT angiogram of the head and neck showed irregular luminal contours of the distal internal carotid arteries bilaterally and diffuse small caliber intracranial vessels in the anterior and

posterior circulations with mild luminal irregularities [Figure 1]. No stenosis or other abnormal findings were present.



**Figure 1:** Computed tomography angiogram of the head showing coronal (A), sagittal (B), and (C) axial reconstructions. Internal carotid (blue arrow), middle cerebral (red arrow), anterior cerebral (green arrow), and posterior cerebral (purple arrow) arteries show narrowing and dilation of the intravascular lumen consistent with intracerebral fibromuscular dysplasia. Images were further enhanced with Perfusion (orange column) and Flow (blue column) color look up table (CLUT) presets using Horos imaging software.



**Figure 2:** Conventional angiogram of the right (A) and left (B) internal carotid arteries showing narrowing and dilation of the intravascular lumen (blue arrows) compatible with fibromuscular dysplasia.

The luminal irregularities seen on imaging raised suspicion for FMD. But due to its low incidence, atypical disease demographic, and this patient's other comorbid neurologic conditions, the medical team looked for more probable causes of his neurologic dysfunction. Initial differential diagnosis included:

- (1) multiple sclerosis flare
- (2) vasculitis
- (4) cervical/lumbar radiculopathy
- (5) conversion disorder
- (6) FMD.

Patient was admitted to the Internal Medicine service and was started on a 5-day course of methylprednisolone. His imaging was negative for a stroke and no white matter plaques resembling MS were visualized. However, his neurologic findings were diffuse and did not follow a focal pattern, raising suspicion of a MS flare. He reports only one past MS flare over 30 years ago and he has been well controlled on weekly interferon-beta since his diagnosis. On day 2 of admission, his presenting symptoms had resolved and was now exhibiting left sided upper/lower extremity weakness with intermittent tremors. Over the following days his neurologic symptoms gradually improved, but his neurologic exam remained inconsistent.

His imaging findings were also concerning for a vasculitis. Polyarteritis Nodosa is a necrotizing vasculitis of medium-sized arteries characterized by alternating fibrosis and aneurysm, similar to those seen on this patient's CT angiogram. Neurologic symptoms can be seen, but multiple organ systems are generally

affected [8]. A renal duplex ultrasound showed adequate flow in the renal arteries bilaterally and the patient denied symptoms of postprandial abdominal pain consistent with significant mesenteric artery stenosis. The patient's visual changes were also concerning for Giant Cell Arteritis (GCA) or Takayasu Arteritis (TA). Inflammatory markers were only mildly elevated; ESR 47 and CRP 0.84. An ESR <50 mm/h and absence of temporal artery tenderness to palpation made GCA less likely [9]. A CTA chest was ordered and showed no aortic arch fibrosis or vascular narrowing consistent with TA [10]. ANA, ANCA, complement studies were also negative.

Per chart review, patient had undergone an L1-L4 spinal fusion for severe lumbar stenosis one year prior. He now experiences chronic pain and is scheduled 5 mg oxycodone three times a day by a pain management physician. In the ED he complained of shooting pains down his right UE & LE, resembling cervical and lumbar radiculopathy. MRI spine was ordered and showed marked cervical spondylosis with central canal and foramina narrowing. No post-surgical changes seen in the lumbar spine. These findings may have explained the UE weakness and paresthesias, but they fail to explain the bulbar symptoms and LE deficits seen on presentation.

Finally, conversion disorder was briefly considered. This diagnosis explains the multiple unrelated neurologic symptoms and inconsistent neurologic exam. Patient noted to the medical team that his father was recently placed in hospice care and he was distressed by the thought of his father passing. However, the patient has no history of psychiatric disorders, which are present in approximately 90% of patients with conversion disorder [11]. This diagnosis was unlikely.

After the 5-day course of methylprednisolone, patient had fully recovered to his baseline neurologic status with no complications during his hospital stay. His hypertension remained difficult to control throughout his hospital stay. Ultimately, he was placed on metoprolol tartrate, amlodipine, and losartan. Patient had a history of angioedema from Lisinopril, but the benefits of blood pressure control outweighed the low risk of ACE-ARB cross reactivity inducing angioedema. The treatment team had also noticed the patient preferred to keep his room at 95°F (35°C), which was unusual during the summer months. His TSH and T4 were below the normal range, indicating central hypothyroidism. He had no history of intracranial pathology or overt signs of hypopituitarism, so further workup was deferred to outpatient follow-up.

After a broad investigation revealed inconclusive findings, the patient was sent for conventional angiogram of the head, neck, and abdomen; as this is the gold standard of diagnosis for FMD [6]. The findings showed focal areas of narrowing and dilation without significant stenosis in the left and right cervical internal carotid arteries. Similar vascular findings were shown on his CT angiogram of the head, confirming a rare diagnosis of Cerebrovascular Fibromuscular Dysplasia with diffuse intracranial vascular involvement. No abnormalities were found in the renal arteries or vertebral arteries [Figure 2].

## Discussion

This was an example of an interesting case of a man with multiple comorbidities who presented with symptoms concerning of a stroke/TIA. His original imaging was concerning for an undiagnosed vascu-

lar abnormality a potential cause of his neurologic symptoms. But considering the low likelihood of FMD or a vasculitis in this patient, more common etiologies were investigated first. After a comprehensive workup, a new diagnosis of cerebrovascular fibromuscular dysplasia inducing a low-flow TIA was concluded. With the patient now recovered to his neurologic baseline, the next question to be answered is “what is the next step?”

In 2019 the European Society of Hypertension and the Society for Vascular Medicine released the first comprehensive series of guidelines regarding the diagnosis, classification, and treatment FMD [6]. The guidelines focus on two mainstays of treatment: (1) hypertension control (2) thromboembolism prevention. This patient suffered from moderately uncontrolled hypertension in the absence of renal artery stenosis. His hypertension was likely primary with OSA as a contributing factor. His home medications included maximum doses of amlodipine and metoprolol tartrate with widely variable blood pressures ranging from 180s/100s to 120s/70s. He also had a history of ACE angioedema. But the team decided to add Losartan to his regimen. Recent studies have shown ACE-ARB cross reactivity is unlikely and the benefits of blood pressure control outweighed the low risk of recurrent angioedema [12]. The guidelines also encourage the use of a daily low-dose aspirin for thromboembolism prevention. Dual antiplatelet therapy was deferred as randomized control trials have demonstrated the benefits for stroke prevention do not outweigh the risks of bleeding [13].

This particular case showcased a rare presentation of FMD involving the internal carotid arteries bilaterally with diffuse extension into the intracerebral vasculature. Bilateral carotid FMD is a common finding, with prevalence closely following renal artery FMD [14]. Few case reports have been published regarding intracerebral FMD [15,16]. These patients share similar risk factors to other forms of FMD. Continued hypertension control and single antiplatelet therapy is still recommended. Prognosis is favorable in this patient. Continued reporting and utilization of the US Registry for Fibromuscular Dysplasia will provide greater insight into this rare disease and open the door to more effective therapies.

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