

# Fibromuscular Dysplasia of the Vertebral Artery Presenting with Lateral Medullary Syndrome: A Case Report

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**Abstract-** We have recently encountered a rare case of fibromuscular dysplasia (FMD) of the vertebral artery (VA) presenting as lateral medullary syndrome. A 39-year-old male was admitted to our hospital due to vertigo, dysarthria and numbness of the left face and the right limbs. A magnetic resonance imaging (MRI) and magnetic resonance angiography (MRA) of brain revealed lateral medullary infarction in the territory of the left posterior inferior cerebellar artery (PICA). The angiography of the VA revealed tubular stenosis of the left extracranial VA and a focal vascular kinking as well as web in the right extracranial VA, confirming the diagnosis of FMD. We present this rare case to emphasize that FMD could be one of the risk factors causing lateral medullary syndrome in young people.

**Key Words:** Fibromuscular dysplasia (FMD), Lateral medullary syndrome, Magnetic resonance imaging (MRI), Magnetic resonance angiography (MRA), Posterior inferior cerebellar artery (PICA)

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## INTRODUCTION

Lateral medullary syndrome is a special form of infarction that is caused by occlusion of the posterior inferior cerebellar artery. The clinical signs include dysarthria, ipsilateral limbs ataxia, vertigo, ipsilateral Horner's syndrome, ipsilateral pain and temperature sensory loss in the face and contralateral pain and temperature sensory loss in the limbs and trunk. The corticospinal tract is spared and the muscle power remained normal.

In general, the etiology of lateral medullary syn-

drome under 50 years of age differs from those of geriatric patients. Stroke in the first two decades of life were more frequently associated with infectious and inflammatory conditions, whereas stroke in the next two decades is more likely associated with structural abnormalities or hypercoagulability-related conditions<sup>(1)</sup>. The diagnostic management of young stroke patients thus usually involves special cardiac and neuroradiological investigations<sup>(2)</sup>.

Fibromuscular dysplasia (FMD) is one of the structural abnormalities which have been associated with young stroke. FMD is a non-atheromatous, non-inflama-

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tory, segmental arteriopathy of unknown etiology primarily affecting small and medium sized arteries. Fibroplasia of the tunica media is most common<sup>(3)</sup>. It involves primarily the internal carotid and renal arteries and less often the vertebral, iliac, subclavian, and visceral arteries<sup>(4)</sup>. The cerebral vessels are involved in 25-30% of the cases of FMD<sup>(5)</sup>. However, FMD of these cephalic vessels is often asymptomatic and detected as an incidental angiographic finding<sup>(6)</sup>. In this article, we report a case of FMD involving the VA, manifested as arterial dissection and lateral medullary syndrome.

### CASE REPORT

A 39-year-old male who experienced a sudden onset of left neck pain, followed by persisted vertigo, vomiting, left facial numbness, hoarseness and swallowing difficulty for about 3 days before admission. He sought for medical support and received medications from a otolaryngologist initially. Due to the persistent symptoms he was transferred to our emergent unit where brain computed tomography revealed no gross abnormality. Then he was admitted to the ENT (ears, nose, and throat) ward where left vocal cord palsy was found by laryngoscope. As a diagnosis of brain stem infarction seems likely during the neurological consultation, the patient was transferred to the neurological ward in the next day and the young stroke screening procedures were carried out.

Physical examination revealed normal findings including vital signs. Neurological examination revealed hypoesthesia over left facial and right limbs and trunk, mild ptosis with miosis of the left eye (Horner's syndrome), and deviation of uvula to the right side. However, eye movement, muscle power in four limbs, deep tendon reflexes, and plantar responses were all normal. Blood biochemistry tests revealed no abnormalities, but blood cell counts revealed leukocytosis (WBC: 16400/ul; N/L:85/9.2) which was probably ascribable to urinary tract infection.

He denied past history of any systemic disease except for a sudden onset of right side deafness about 2 years ago. There is no recent trauma history. However,



Figure 1. Brain MRI (FLAIR) revealed an acute infarction of the left dorsolateral medullary region (arrow).

his elder sister died at the age of 40 due to an unknown type of cerebral accident, with no history of previous systemic disorders or prominent risk factors for stroke. The other parts of the family history was unremarkable.

We arranged MRI and MRA of the brain to evaluate the infarction. Brain FLAIR (fluid attenuated inversion recovery image) MRI revealed an acute infarction of the left dorsolateral medullary region (Fig. 1), the territory of the left posterior inferior cerebellar artery (PICA). Brain MRA showed progressive tapering of the left distal VA and axial T1-weighted MRI demonstrated a high-signal intramural hematoma and consequent narrowing of the left VA, indicating dissection of the vessel (Fig. 2). We therefore arranged vertebral angiography to assess the underlying structure abnormalities of the vessels. We found tubular stenosis of the left extracranial VA and focal vascular kinking as well as web in the right extracranial VA (Figs. 3A and 3B). FMD was diagnosed according to these findings. The results of the other young stroke studies including serum electrolytes, renal and liver function tests, erythrocyte sedimentation rate,

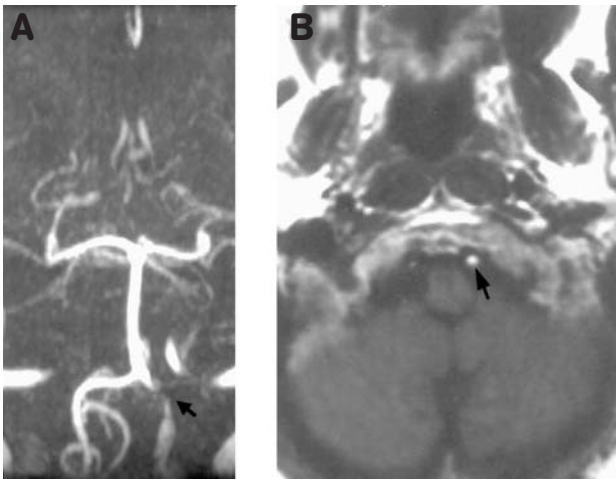


Figure 2. (A) Brain magnetic resonance angiogram shows dissection with progressive tapering of the left distal vertebral artery (arrow). (B) Axial T1-weighted magnetic resonance image showed a high-signal intramural hematoma of the left vertebral artery (arrow).

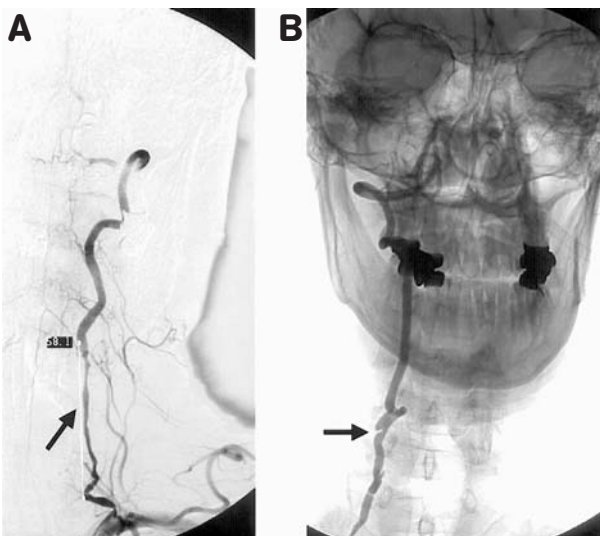


Figure 3. (A) Left vertebral angiography showed tubular stenosis (58.1 mm in length) of the left extracranial vertebral artery (arrow); (B) Right vertebral angiography revealed a focal vascular kinking and web (arrow) in the right extracranial vertebral artery.

echocardiography, Duplex ultrasound and color Doppler flow imaging, coagulation profiles (including protein C, protein S, antithrombin III, lupus anticoagulant, anticardiolipin antibody, and autoimmune parameters), syphilis, and human immunodeficiency were unremarkable

except for the mild hypercholesterolemia (281 mg/dl). The symptoms of dysphagia, saliva drooling and vertigo improved gradually but hoarseness persisted. Anti-platelet drug was given to prevent recurrent stroke. The patient discharged 9 days after admission in a stable condition.

## DISCUSSION

Leadbetter and Burkland first described a patient with hypertension and renal artery stenosis secondary to FMD in 1938<sup>(6)</sup>. Progressive disruption of the arterial wall by FMD may have several consequences. FMD-related arterial disease has been reported in up to 20% of cervical carotid artery dissections<sup>(7-9)</sup>. FMD may also underlie arterial dissection in the vertebral, middle cerebral, anterior cerebral, or superior cerebellar arteries, and could be the cause of “unexplained” or “spontaneous” dissections in some patients. In this case, the “spontaneous” VA dissection did lead us to consider FMD as a possible pathological mechanism underlying the stroke.

A pathological classification of FMD was proposed by Harrison and McCormack<sup>(10)</sup> in 1971 and was revised by Stanley and colleagues<sup>(11)</sup> in 1975. Three main types of FMD have been identified: intimal fibroplasias, medial FMD, and periarterial or periadventitial fibroplasias.

Although FMD should be a pathologic diagnosis, the diagnosis can be made with a high degree of accuracy on the basis of the angiographic appearance<sup>(4,12)</sup>. In this patient, FMD was diagnosed chiefly by the angiographic findings. The long tubular stenosis may be indicative of the subtype of intimal fibroplasias. In young patients of this subtype FMD, long tubular stenosis is more common, whereas smooth focal stenosis predominates in older patients.

The histologic abnormalities may cause three patterns of pathology of the arterial wall<sup>(13-15)</sup>. Multifocal stenosis, alternating with mural dilatations (beaded appearance), is the most common pattern and occurs in 80% to 90% of patients. In 6% to 12% of patients, there is a longitudinal stenosis (tubular appearance). When FMD involves the arterial wall in a noncircumferential manner, there may be outpouching or diverticulum of the

wall (4% to 6% of patients) or, rarely, an asymmetrical septal appearance, leading to a weblike stenosis.

The “web-shaped” tissue of right vertebral artery in this case was an unusual and interesting finding of FMD. There were nine cases that have been reported to show an internal carotid web, but none had the web in vertebral arteries<sup>(16)</sup>. In the report the author suggested that the patient with this particular lesion had a higher risk for stroke than those with the usual “string of beads” lesion. In these patients, surgical treatment should be considered. But in our patient the web was in VA and lacks of clinical experience. So medical treatment was applied first due to the high risk of surgical intervention, and closely follow-up of the clinical conditions is mandatory.

The natural history of FMD is not clear but most cases have a relatively benign clinical course. The available data suggests a low incidence of stroke in both asymptomatic and symptomatic patients with FMD<sup>(14)</sup> and the progression seems to be less pronounced in the cerebral than in the renal vessels<sup>(4)</sup>.

The pathogenesis of FMD remains unknown. The pathology of this disease has been described by Schieveink et al.<sup>(17)</sup>. The narrowed arterial segments show degeneration of the elastic tissue and irregular arrays of the fibrous as well as the smooth muscle tissues in mucous ground substances. The dilatations probably result from atrophy of the coat of the vessel wall<sup>(4)</sup>, and the vasculopathy of web formation may be due to intimal thickening with duplication of the internal elastic lamina<sup>(18)</sup>. Several related hypotheses have been proposed<sup>(4,16,19,20)</sup>, including (1) possible hormonal effect due to female predominance, (2) possible contributions from trauma or repeated microtrauma (the mechanical hypothesis), (3) the genetic hypothesis (It was reported that one third of the patients had a family history of FMD inherited as an autosomal dominant trait. There was also increased occurrence of stroke in the pedigrees.), and (4) the hypothesis of ischemia of the blood vessel wall. There may still be other unknown contributory factors. It is unlikely that all subtypes of FMD have the same underlying causes. FMD may be a multifactorial disorder.

The treatment of FMD is largely supportive once the

symptoms become manifest<sup>(20)</sup>. Surgical intervention should be reserved for patients with evidence of progressive cerebral ischemia<sup>(4,21)</sup>. Various surgical techniques have been used for cerebrovascular FMD such as resection and grafting, angioplasty, bypass, graduated intraluminal dilatation, and more recent advances in instrumentation such as stenting<sup>(4,21,22)</sup>. However, there is still a lack of clearcut management guidelines for such cases.

FMD is a rare cause of young stroke, but should still be considered in young patients with spontaneous arterial dissection and stroke. We present this rare case to emphasize the significance of FMD as one of the risk factors causing lateral medullary syndrome in young people and hope to add to a more clear guideline for the diagnosis and management of such cases.

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