Case report

Archives of Psychiatry Research 2023;59:129-132
DOI:10.20471/may.2023.59.01.18

Female with Fibromuscular Dysplasia

Melanija Pintarić[1], Rafaela Vukasović[1], Arijana Lovrenčić- Huzjan[1,2,3]

[1]Department of Neurology, University Hospital Centre Sestre milosrdnice, Zagreb, Croatia, [2]School of Dental Medicine, Zagreb, Croatia, [3]School of Medicine, University of Zagreb, Zagreb, Croatia

Abstract - Fibromuscular dysplasia (FMD) is a noninflammatory, nonatherosclerotic disorder that cause arterials stenosis, occlusion, aneurysm, dissection, and artery tortuosity. The etiology of FMD remains unknown. The most frequently involved arteries are the renal, internal carotid, vertebral arteries and visceral. Among adults, FMD is more common in females. Most common symptoms are renovascular hypertension, headache, pulsatile tinnitus, neck pain, and abdominal pain. We present a 62-year old female patient who was diagnosed with grade 3 arterial hypertension at the age of twenty -nine. During diagnostic treatment, narrowing of the right renal artery was determined and percutaneous transcutaneous angioplasty of the right renal artery was performed. Diagnosis of fibromuscular dysplasia was suspected. After this procedure, the value of blood pressure was normalized. At the age of forty, the patient was re-introduced with antihypertensive, and CT angiography showed 50 % lumen stenosis of the right renal artery. She was referred to a neurologist. The patient has been complaining about frequent headaches, pulsatile tinnitus in the right ear, increased fatigue, and occasional abdominal pain for many years. Neurosonological testing showed partially wall thickening in the left common carotid artery and multiple stenosis and dilatation of the left internal carotid artery. The left vertebral artery has multiple stenoses and dilatations in the distal V1 segment. The findings indicated changes in fibromuscular dysplasia and the diagnosis of FMD was confirmed.

Key words: fibromuscular dysplasia; hypertension, renovascular; renal artery obstruction

e-mail: apr.kbcsm@gmail.com • www.http://apr.kbcsm.hr

Introduction

Fibromuscular dysplasia (FMD) is a noninflammatory, nonatherosclerotic disorder that cause arterials stenosis, occlusion, aneurysm, dissection, and artery tortuosity [1]. The etiology of FMD remains unknown. The most frequently involved arteries are the renal, internal carotid, vertebral arteries and visceral [2]. Among adults, FMD is more common in females. Disease presentation is diverse, depending on the arterial segment involved and the severity of disease. Most common manifestation of involvement of renal arteries is renovascular hypertension, and manifestations of cerebrovascular FMD are headache, pulsatile tinnitus, neck pain, and abdominal pain or more severe like TIA and stroke [3]. In the past, FMD was classified according to the arterial layer affected (intima, media or adventitia) [4,5]. Nowadays FMD is always diagnosed radiographically so pathological classification has been replaced by an angiographic classification [6].There are two angiographic subtypes of FMD. Multifocal FMD is more common, angiography shows „string of beads“, and fo-

Correspondence to:
Melanija Pintarić, MD, Department of Neurology, University Hospital Centre, Sestre milosrdnice, Vinogradska cesta 29, 10 000 Zagreb, Croatia, Phone :+ 385 98 9069 351
E-mail: melanijabarbir@gmail.com

Figure 1. CT- angiography showing 50 % lumen stenosis with irregular soft plaques located at initial part of the right renal artery (arrow)

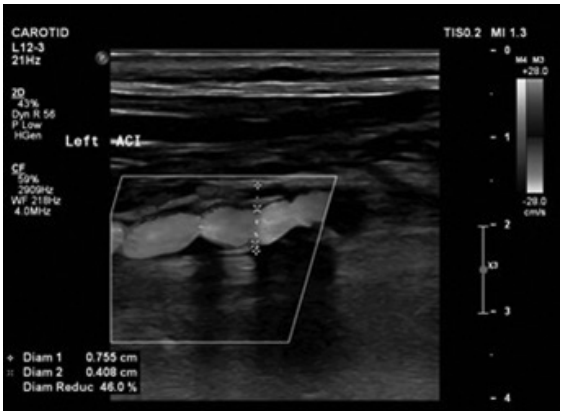

Figure 2. Duplex ultrasound of the left carotid artery showing wall thickening

cal FMD is less common and shows „circumferential or tubular stenosis“ [7].

Subjects and Methods

We present a case of a 62-year old female patient who was diagnosed with grade 3 arterial hypertension (maximum blood pressure values up to 230/140 mmHg) at the age of twenty - nine. During diagnostic treatment, narrowing of the right renal artery was determined and percutaneous transcutaneous angioplasty of the right renal artery was performed. Diagnosis of fibromuscular dysplasia was suspected. After this procedure, the value of blood pressure was normalized. At the age of forty, the patient was re-introduced with antihypertensive drugs with inadequate regulation of arterial pressure, and she was referred to further treatment. Nephrological examination was performed, and Doppler ultrasound of the renal arteries showed acceleration of the flow over 150 cm/s at initial part of the right renal artery. CT angiography showed 50 % lumen stenosis with irregular soft plaques located at initial part of the right renal artery (Figure 1). Continuous measurement of arterial pressure was performed and confirmed uncontrolled hypertension. She was referred to a neurologist. According to the anamnesis, the patient has been complaining about frequent headaches, pulsatile tinnitus in the right ear, increased fatigue, and occasional abdominal pain for many years. Neurosonological testing showed partially wall thickening in the left ACC (0.86 mm) and multiple focal stenosis of the left internal carotid artery (ACI) (Figure 2). The left vertebral artery has multiple stenoses and dilatations in the distal V1 segment (Figure 3). The findings indicated changes in fibromuscular dysplasia and the diagnosis of FMD was confirmed.

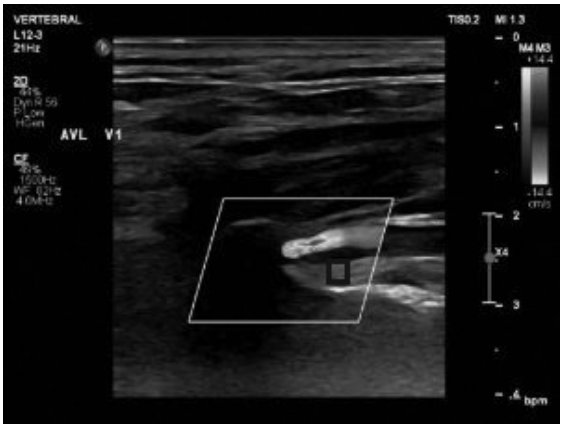

Figure 3. Duplex ultrasound of left vertebral artery showing stenosis and dilatations in V1 segment

Discussion

Approximately two-thirds of patients have multiple arteries involved [1]. Approximately 65 % of patients with renal artery FMD also have carotid or vertebral FMD. Similarly, pa-

tients with carotid or vertebral FMD are likely to have renal involvement [2]. Older literature indicate that carotid artery involvement was present in only 20 % - 30 % of patient with FMD, because previously it was not practice to image other arterial territories after renal FDM was diagnosed. First it is important for physician to think, and to suspect of FMD. FMD should be suspected particularly in a woman under the age of 60 years with sever hypertension, when the onset of hypertension is before the age of 35 years, when patient complains of persistent headache or pulsatile tinnitus. Also, dissection of a peripheral (carotid, vertebral, renal) artery or aneurysm in a visceral, carotid or vertebral artery should raise suspicion of FMD [8]. In patient with suspected FMD the diagnosis is confirmed by diagnostic imaging.

The aim was to present specific symptoms of a patient with hypertension and renal fibromuscular dysplasia in whom cervical fibromuscular dysplasia was established as a reflection of involvement of another arterial territory. In patients with renal artery stenosis at a younger age, it is necessary to estimate the involvement of other arterial territories because approximately 65 % of patients with renal artery FMD also have cervical FMD. Because FMD has tendency to occur in multiple arterial territory, and because patients with FMD have a high prevalence of arterial aneurysm and dissection, they should undergo one time of imaging from head to pelvic.

Acknowledgements

None.

Conflict of Interest

None to declare.

Funding Sources

None.

References

1. Gornik HL, Persu A, Adlam D, Aparicio LS, Azizi M, Boulanger M, et al. First International Consensus on the diagnosis and management of fibromuscular dysplasia. Vasc Med. 2019;24:164-89.
2. Olin JW, Froehlich J, Gu X, Bacharach JM, Eagle K, Gray BH, et al. The United States Registry for Fibromuscular Dysplasia: results in the first 447 patients. Circulation. 2012;125:3182-90.
3. Kim ESH, Olin JW, Froehlich JB, Gu X, Bacharach JM, Gray BH, et al. Clinical manifestations of fibromuscular dysplasia vary by patient sex: a report of the United States registry for fibromuscular dysplasia. J Am Coll Cardiol. 2013;62:2026-8.
4. Stanley JC, Gewertz BL, Bove EL, Sottiurai V, Fry WJ. Arterial fibrodysplasia. Histopathologic character and current etiologic concepts. Arch Surg. 1975;110:561-6.
5. Harrison EG Jr, McCormack LJ. Pathologic classification of renal arterial disease in renovascular hypertension. Mayo Clin Proc. 1971;46:161-7.
6. Olin JW, Gornik HL, Bacharach JM, Biller J, Fine LJ, Gray BH, et al. Fibromuscular dysplasia: state of the science and critical unanswered questions: a scientific statement from the American Heart Association. Circulation. 2014;129:1048-78.
7. Savard S, Steichen O, Azarine A, Azizi M, Jeunemaitre X, Plouin PF. Association between 2 angiographic subtypes of renal artery fibromuscular dysplasia and clinical characteristics. Circulation. 2012;126:3062-9.
8. Kadian-Dodov D, Gornik HL, Gu X, Froehlich J, Bacharach JM, Chi YW, et al. Dissection and aneurysm in patients with fibromuscular dysplasia: findings from the U.S. Registry for FMD. J Am Coll Cardiol. 2016;68:176-85.