

Unusual Form of Fibromuscular Dysplasia of the Renal Artery in a Young Man: A Case Report

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Abstract: *Fibromuscular dysplasia (FMD) of the renal artery is a rare, idiopathic and asymptomatic disease. The suspicion and the radiological evidence is disrupted by the occurrence of renovascular arterial hypertension in the subject under 30 years of age and is resistant to antihypertensive treatments. According to the literature, the prevalence in the general population is between 30 to 50 years of life and quite common in women than in men. Fibromuscular dysplasia most commonly affects the renal, carotid and vertebral arteries, but it can theoretically affect any artery. Its prevalence is estimated between 4 and 6% in the renal arteries and between 0.3 and 3% in the cervico-encephalic arteries. Radiologists play an important role in the diagnosis of fibrous dysplasia, and a good knowledge of the signs of fibrosclerotic dysplasia will certainly help to reduce the delay between the first symptoms and the diagnosis. In order to assess and demonstrate the interest or the place of Radiology in the FMD diagnostic management, we report a case of fibromuscular dysplasia in a 27-year-old patient followed for ather hypertension since 2016 and resulting uncontrolled with often modification of antihypertensive treatment coming to triple therapy. An angio-scanner demonstrated the diagnosis: characteristic image string-of-beads aspect corresponding to the dilated sections of the right renal artery and the wire between the beads with narrowed sections. The confirmation was made by the arteriography which had shown stenosis of appearance in rosary. Endoluminal revascularization with placement of two stents was performed as a definitive treatment. Follow-up was simple for a period of one year with normalization of blood pressure.*

Keywords: Fibromuscular dysplasia, renal artery, angio-scanner, arteriography, angioplasty

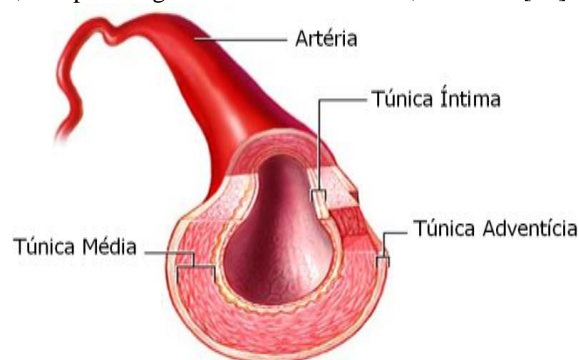
1. Introduction

The term fibromuscular dysplasia (FMD) is applied to a group of idiopathic, segmental, non-inflammatory and non-atherosclerotic diseases of the arterial wall, resulting in stenosis of small and medium-sized arteries [2,3]. The first description dates back to 1938 on an operative specimen (nephrectomy) [3] and the term "fibromuscular dysplasia" is used for the first time in 1958 [4].

Epidemiological data show that FMD reaches, in decreasing order of frequency, renal arteries (3-6%), supra-aortic trunks (0.3-3%), other visceral arteries and limb arteries. Several authors believe that these prevalence values are underestimated because FMD is frequently asymptomatic [13-14]. In a review combining the results of 4 studies including 3,181 asymptomatic patients undergoing renal angiography before donation of a kidney transplant, 4.4% (139 subjects) presented with FMD lesions [15]. The symptomatic presentation of renal artery DFM is renovascular hypertension. The prevalence of this presentation is estimated at 10% of renovascular hypertension cases and less than 4/1000 in the middle-aged hypertensive population (approximately 8 new cases per 100 000 active persons per year) [5].

By analyzing pathology and pathophysiology we find that the attack can be at the level of the intima, the media or the adventitia, each type having its peculiarities to arteriography. The most common form concerns the media and gives the classic appearance in "stack of plates or beads strung" at angiography. The intimal form gives rather an isolated narrowing [5]. Estrogen impregnation given the predominance of women. The predominance of lesions of the right renal artery suggests a mechanical component because the right kidney is more mobile than the left kidney; the mechanism may involve vasa vasorum compression

leading to ischemia. Smoking could also be a risk factor. Autosomal dominant inheritance with incomplete penetrance and variable expressivity is suggested in 6 to 10% of familial types; no specific gene has been identified, however [16].



The exam of choice (with diagnostic and therapeutic value) to establish the diagnosis is arteriography, which shows the pathognomonic picture: staggered stenosis, in "stack of plates or beads strung". Despite this, an aspect of isolated stenosis does not exclude dysplasia but this aspect is often confused with atheromatous stenosis much more common, at least in an older patient. In a young person, this aspect must be reminiscent of a fibro-muscular origin, especially if the narrowing is central (and not eccentric as is a non-circumferential plaque atheroma). Angio-CT and angio-IMR are alternatives to conventional arteriography, with good diagnostic reliability [9,10]. Vascular Doppler ultrasound is difficult to distinguish between atheromatous stenosis and dysplasia [5].

The angiographic classification of Kincaid et al. based on a series of 125 patients is more commonly used. Its purpose is to distinguish [17]:

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- I. Multi-focal type (multiple stenosis or strung beads chain).
- II. Uni-focal type (short stenosis less than 1 cm long).
- III. Tubular type (stenosis more than 1 cm long).
- IV. Mixed type.

Another classification that is not frequently used in daily practice because several types can coexist in the same patient and / or the same artery, it is the radiological classification based on anatomopathological correlations: 1-Medial FMD (60-70%) image in "strung beads" corresponding histologically to areas of fibrous hypertrophy of the media and rupture of the internal elastic lamina. It's the most frequent, it's observed mainly towards quarantine; 2-Perimedial or sub-adventitial FMD (10-20%), tubular dysplastic stenosis histologically the outer layers of the media are the site of extracellular matrix hyperplasia pushing back the adventitia and reducing the vessel lumen; 3-The intimal FMD (5%) histologically unifocal stenosis is characterized by circumferential intimal thickening without involvement of the other tunics. [15, 17]

The appearance of string-of-beads aspects is the most characteristic aspect of the FMD and indicates the presence of medial lesions. The other angiographic aspects are less specific to a histological type. [15, 17]

Treatment of patients with FMD may include: medical treatment, endovascular treatment and surgical treatment.

The purpose of this article is to review and demonstrate the interest or the place of Radiology in the diagnosis of Fibromuscular Dysplasia (FMD).

2. Material and Methods

Mr. F.H., 27 years old with no medical-surgical history and no history of abdominal trauma, who has had arterial hypertension for two years and is resistant to triple therapy and associated with headaches. The clinical examination was poor except for the elevation of blood pressure and abdominal exacerbation, where there is a synchronous vascular murmur at audible heartbeat at the umbilical region, radiating to the right flank. The biological assessment revealed hypokalemia and hyperkaliuria. A Doppler ultrasound of the renal arteries showed an ostial segment of the normal right renal artery with well parallel edges but a remodeled truncal segment with rosary aspect associated with an accelerated blood flow of the narrowed zone characteristic of a fibro-muscular dysplasia. An angiogram showed a characteristic image of strung beads corresponding to the dilated sections of the right renal artery and the wire between the beads with narrowed sections, (see Fig. 1). Arteriography was done and showed stenosis of appearance rosary (Cf Fig 2). The patient underwent percutaneous endo-luminal angioplasty with the placement of two stents, which made blood pressure normal without the need for antihypertensive therapy. The post-therapy follow-ups are simple, living patient and asymptomatic over a retreat of more than two (2) years.

3. Discussion

There are no formal radiological diagnostic criteria. Despite that, it is well established that the string-of-beads aspect found in renal or other frequent locations mentioned above on arteriography, CTA or MRA is highly suggestive of FMD. [28]. Another frequent finding suggestive of an FMD diagnosis is the presence of a "web-like" defect at the origin of the internal artery [3, 20].

In such situations, lesions may be discrete at the cervical level and typical at the renal level. Thus, imaging of the renal arteries will help to establish the diagnosis. The opposite situation is also true, and performing imaging of the renal arteries is recommended in a patient presenting discrete but suggestive lesions in the cervico-encephalic arteries so as to confirm the diagnosis, and vice versa [20].

The symptomatic form is estimated at 10% of cases of renovascular hypertension, pathology on which FMD in our patient was revealed. FMD affects women more with 9:1 ratio. Usually is diagnosed in adulthood, between 30 to 50 years, but it can sometimes appear in childhood [3]. Its occurrence in the young man is exceptional as related to our case. It affects all populations, regardless of their geographical origin.

According to the pathophysiology, the etiology of FMD remains unknown, in spite of it different factors being related: the estrogenic impregnation which justified predominance feminine; mechanical component that justified predominance in the right renal artery (our case); the tobacco ; genetic (autosomal dominant transmission in 6 to 10% of familial types without any specific gene identified) and immunological (HLA-Drw6) [5]. Vascular involvement can be at the level of the intima, the media or the adventitia. The most common form, about 80% of the cases concerns the media and gives the classic aspect in "stack of plates or pearls threaded" with the angiography, most in women between 30 and 50 years [6]. Despite the arterial injury in our case of unilateral localization that accounts for less than half of cases, the vast majority of bilateral locating in more than 50% of cases.

The diagnosis of FMD is based primarily on imaging. Non-invasive diagnostic methods include, in ascending order of precision: Doppler ultrasound, magnetic resonance angiography (MRA), CT angiography [9]. The standard examination for the diagnosis of FMD is conventional angiography, but this invasive procedure is only used for patients in whom revascularization is considered [10].

In our case the diagnostic approach began with renal echo-Doppler showing an acceleration of blood flow, indicating stenosis of the renal arteries with renal failure. This makes it possible to study the size of the kidneys, which is a good criterion for assessing the severity of the disease and its follow-up [19].

Magnetic resonance angiography (MRA) and angio-CT have high specific characteristics for the detection of renal artery stenosis due to FMD (92 and 84% respectively), but have a relatively low sensitivity (64 and 62% respectively) [15]. A

recent study comparing contrast-enhanced angio-MRI with conventional angiography showed a sensitivity of 97% and a specificity of 93% for the diagnosis of FMD. MRI was more sensitive to detecting the appearance of the pearl chain (97%) than to detect stenosis > 50% (68%) [20].

There are different therapeutic modalities for the FMD. We used the approach percutaneous angioplasty with placement of two stents in accordance with the recommendations (indicated in case of stenosis) with a good evolution. Currently, it is considered by the majority of authors as the most reasonable therapeutic option and is reserved for patients with recurrent symptoms, hemodynamic instability, despite medical treatment. However, the results of long-term percutaneous angioplasty are still poorly understood [18].

Medical treatment may be considered as for example antiplatelet therapy is the rule even though the reasons are essentially empirical, with no evidence of effectiveness in the evolution of this type of injury [5]. Surgery is reserved for patients with accessible symptomatic lesions and low perioperative risk because long-term anatomical outcomes are good. [18]

4. Conclusion

FMD is a disease that affects women more than men. Imaging plays an important role in the diagnosis of FMD based mainly in its pathognomonic sign «the string-of-beads aspect». To suggest the diagnosis and to continue the investigations it is important to recognize the arterial hypertension which resist the treatment.

5. Conflicts of Interest

The authors do not declare any conflict of interest.

6. Contributions from the Authors

All authors contributed to this article. All authors have read and approved the final version of the manuscript.

Legend

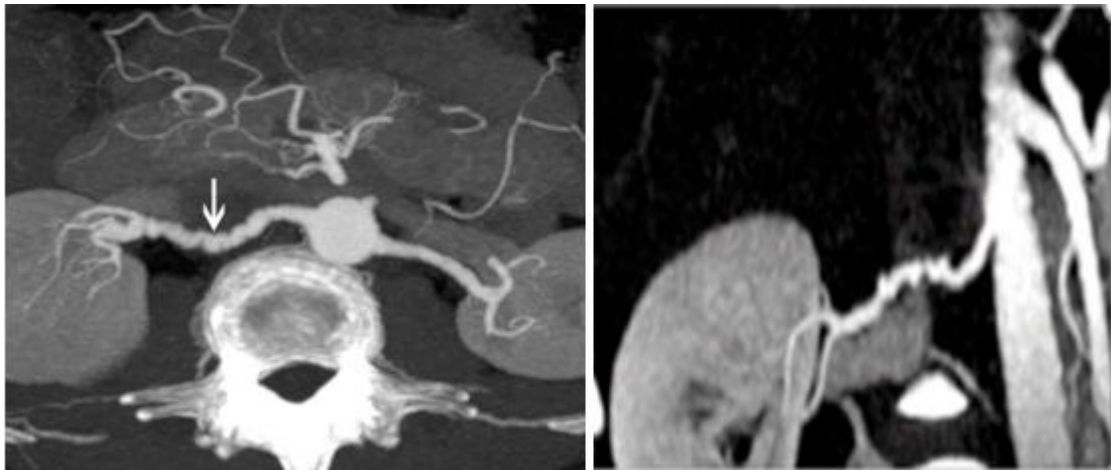


Figure 1: Abdominal angio-scanner showing the image "string-of-beads aspect" of the right renal artery (arrow)

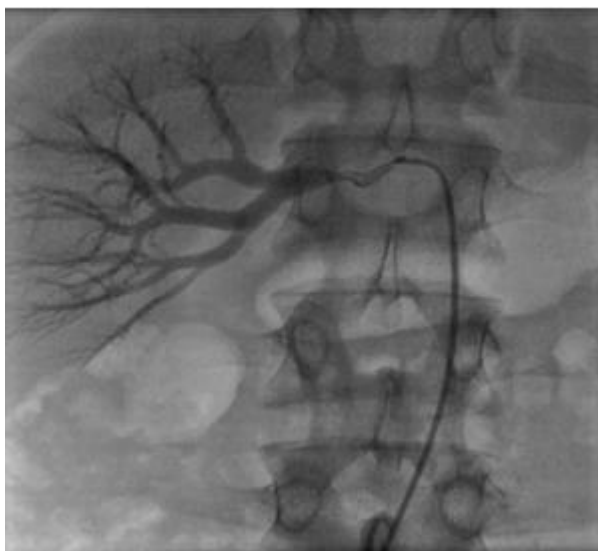


Figure 2: Arteriography showing a stenosis-dilatation of the right renal artery testifying FMD-right renal artery.

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