



All quiet on the front of fibromuscular dysplasia?

Fibromuscular dysplasia (FMD) is an idiopathic and rare disease that presents with segmental, non-inflammatory, non-atherosclerotic, (multi-)focal narrowing of small and medium arteries. It most commonly can be found in the renal, internal carotid, and vertebral artery, followed by visceral and external iliac arteries. Symptoms depend on the affected vascular bed and can reach from renovascular hypertension to non-specific neurological symptoms, such as headache, tinnitus, or rarely stroke in cerebrovascular manifestation, to ruptured aneurysms or arterial dissection with subsequent ischaemia.

Patients affected are often of younger age and predominantly of female gender. Diagnosis is generally based on physical examination and patient's history and is confirmed by typical angiographic morphology, i. e. "string of beads" in multifocal FMD and concentric smooth focal or tubular stenosis in focal FMD. Options for treatment of hypertension in patients with FMD include antihypertensive medication as well as endovascular and surgical revascularization [1]. However, since large-scaled data and randomized clinical trials are missing, there are no guidelines concerning diagnosis and treatment.

Recently, de Groote et al. investigated 123 patients with FMD in Flanders, Belgium [2] with regard to demographics, distribution of vascular involvement, clinical presentation, and treatment strategy in a multi-centre retrospective registry. FMD diagnosis was confirmed based on medical imaging, and patient selection was based on radiology reports and international classification of diseases (ICD) coding.

Demographic data showed the majority of patients to be female (83.7%) with a mean age of 57 ± 15.8 years at the time of diagnosis. Risk factors at baseline included arterial hypertension (81.7%), tobacco use (37.0%), and hypercholesterolemia (43.7%). Affected vascular sites were most often renal (85.3%), followed by carotid (74.7%) and vertebral (39.8%) arteries. De Groote et al. showed renovascular FMD to be more common in male patients, whereas cerebrovascular FMD was more prevalent in females. However, in a large proportion of FMD patients, multiple vascular beds were affected (41% >2 foci). Thus, symptoms ranged from arterial hypertension (59.5%), to headache (26.4%), dizziness (23.1%) or cerebrovascular events (17.9% transient ischaemic attack (TIA) or stroke, 4.9% subarachnoid haemorrhage). Aneurysms and dissections were reported in 25 (20.3%) and 14 (11.4%) patients, respectively, predominantly in cervical and cerebral arteries. Most patients were pharmacologically treated: antiplatelet medication was prescribed in about 52%, two-thirds (70/104, 67.3%) received one or more antihypertensive drugs, most-

ly β -blockers. However, one-third of patients received interventional treatment (24.7% percutaneous transluminal angioplasty (PTA), 9.4% stenting) for renovascular FMD (PTA in 32.8%) as well as in patients with intracranial carotid aneurysms (coiling in 36.4%).

With the results being rather in line with previous US [3] and French registries, the study does not add much to the currently restricted knowledge on FMD. However, it points out some interesting results that are worth taking a closer look at:

1. FMD diagnosis was made incidentally in 8.3% of patients. Could the prevalence of this „rare disease“ be significantly underestimated?
2. FMD diagnosis was made most frequently by DSA (73.5%). Given the broad availability of non-invasive imaging techniques (CT, MRT), but also ultrasound, one may ask for a selective choice of diagnostic approaches.
3. About a quarter of the commonly relatively young patients presented with a severe condition such as a cerebrovascular event (22.8%). In contrast, little is known on primary prevention and indicators for early diagnosis in FMD.

In summary, there is still a lot to learn about FMD since data on optimal diagnostic and therapeutic work-up are scarce. European and nationwide registries [4] are a first important step to approach this probably underrated disease that often strongly affects the patient's quality of life and prognosis.

References

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