52 Fibromuscular Dysplasia

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Overview

Fibromuscular dysplasia (FMD) is a non-inflammatory, non-atherosclerotic, segmental vasculopathy of unknown origin that affects predominantly small and medium arteries. Leadbetter and Burkland first observed FMD in 1938 in the renal arteries of a 5-year-old boy with hypertension.[1] Since then, FMD of the renal arteries has been associated frequently with renal hypertension. In 1964, Palubinskas and Ripley reported the first radiological evidence of an extrarenal location of the disease involving the coeliac axis, and 1 year later Connett and Lansche described the first case of FMD involving the internal carotid arteries. In 1967, Huber and Fuchs reported two patients with radiological evidence of FMD affecting the intracranial arteries.[2,3]

Epidemiology

FMD is often an incidental finding on cervical and cerebral angiographies, and therefore its real incidence is not known; it is seen in between 0.6% and 3.2% of cerebral angiograms, mostly following retrospective reviews of angiographic data.[4] The prevalence of cervicocephalic FMD in autopsy series has been reported only rarely; it was found in 0.02 of autopsies in a clinicopathological study.[5] Women are affected four times more frequently than men, in the majority of cases between 30 and 50 years of age; it rarely occurs in children, where there is no gender preponderance.[4,6,7]

Cephalocervical FMD is generally multifocal, it typically involves the upper cervical segments of the internal carotid arteries (ICAs) and vertebral arteries (VAs), unusual locations for atherosclerotic lesions, the external carotid artery (ECA) and the intracranial vessels may also be affected.[4,8–10] The extracranial ICA is the most commonly affected site of cephalocervical FMD, bilateral involvement is observed in 60% to 80% of patients. Extracranial VA is the second commonly involved site and is frequently associated with ICA involvement. According to collective experience, renovascular disease occurs in 60% to 75% of patients with FMD, cerebrovascular disease in 25% to 30%, and multivessel both renal and cerebral involvement in up to 24%.[6] Renal and carotid arteries are most commonly involved in adults; in children the intracerebral circulation is considered the main area affected.[11] The frequent association of FMD with aneurysmal arterial disease and its occasional familial occurrence may suggest a congenital mesenchymal disorder.[12]

Etiology

FMD is a non-inflammatory, non-atherosclerotic, segmental vasculopathy of unknown origin that affects predominantly small and medium arteries.[6,8,13,14] Different etiological hypotheses have been proposed. Mechanical, ischemic, metabolic and immunological factors may be responsible for the development of FMD.

Stanley supposed that in the context of poor vascular supply of the arterial wall and mechanical stretch–traction stresses, ischemic lesions could result in the architectural changes of medial layer thickening intercalating with thinner areas typical of FMD.[3,15]

Experimental investigations have shown that ischemic insult of the vessel's wall due to impaired vasa vasorum in the presence of significant estrogen levels reproduced comparable vessel dysplasia.[15] However, currently a causative role of oral contraceptive or postmenopausal estrogen therapy cannot be attributed, although hormones may have a facilitating role.[16]

According to Mettinger, the disease may start as a minor lesion of congenital origin predisposing to an abnormal fibroproliferative response to mechanical or circulatory stimuli. The kind of changes that can be considered as being a "minor lesion" are however unclear.[17]

Recent studies have suggested that a deficiency of alpha 1-antitrypsin could be a genetic risk factor for the development of arterial FMD and intracranial aneurysms. This suggests that FMD might represent a relatively non-specific angiographic finding of an underlying biochemical abnormality. If the hypothesis of a biochemical etiology of FMD is true, it might be placed among inherited connective tissue disorders.[13,18–20]

As already stated, the frequent association of FMD with aneurysmal arterial disease and its occasional familial occurrence may suggest a congenital mesenchymal disorder.[12]

Others have implied that FMD is a morphological pattern from different pathological entities, mostly developmental anomalies, initially grouped together by the renal artery involvement.[21]

Pathophysiology and pathogenesis

Pathological features of FMD are smooth muscle hyperplasia, elastic fiber destruction, fibrous tissue proliferation, and general derangement of the arterial wall.[10,22] The lesion is usually segmental, the involved portions show alternating thick and thin areas of vessel wall. The thick areas present degeneration of

the elastic tissue, disruption and loss of the muscular coat, and an increase in fibrous tissue; the dilations are due to atrophy of the vessel wall, and often assume the configuration of mural microaneurysms. Stenotic areas alternating with dilatations produce the classical "string of beads" angiographic pattern.[7,14,23] Unifocal or multifocal stenoses or isolated aneurysmal dilatation are less frequent morphological findings.[7]

Three histological types of FMD have been identified. Concentric rings of fibrous proliferation and/or smooth muscle hyperplasia cause medial thickening and internal elastic lamina disruption, typical of the so-called medial FMD occurring in 90% to 95% of cases. The other less common identified types in the Stanley's classification are intimal and paramedial (adventitial) fibroplasia.[15,22] Intimal and medial hyperplasia occur mostly in women; in men the lesion may sometimes be caused by extra-arterial adventitial and periarterial fibrous bands.[24]

A pathogenic hypothesis states that normal intra-arterial stresses may act on a segment of an artery for longer than on average, and this, with or without a relatively poor blood supply from the vasa vasorum, produces medial damage. This eventually progresses to one or another the histological types of dysplasia, depending on which artery is involved, the level or depth of medial injury, and the mechanism of attempted repair, which may be modified by the hormonal environment.[15]

The symptoms may occur as a result of arterial stenosis, occlusion, thromboembolism, spontaneous dissection, and aneurysm formation and rupture.[8,25]

Clinical features

Cephalocervical FMD can be asymptomatic or manifest as TIAs, cerebral infarction or SAH.[4] Patients may also present with a variety of non-specific symptoms such as pulsatile tinnitus, migraine headache, dizziness, visual disturbances, and cervico-facial hypoesthesia.[14,24]

FMD should be especially suspected in women between the ages of 30 and 50 years with relatively non-specific neurological symptoms or presenting with asymptomatic unilateral or bilateral cervical bruits.[2,24]

FMD is the principal arteriopathy predisposing to spontaneous (non-traumatic) arterial dissection.[23,26–28] Review of the literature indicates that this association may not be as rare as previously believed. Cervicocranial arterial dissections are observed in 15% of cases of FMD. Based on angiographic criteria, it has been shown in up to 25% of cases of spontaneous VA dissection, an association well described in children and young adults; in these cases the dissection is usually medial, involving the V3 segment in 70% of cases.[23,29–31] FMD may also be considered in rare cases of recurrent or bilateral spontaneous carotid dissection, both uncommon occurrences that may be attributed to an underlying predisposing factor.[26,32]

The incidence of intracranial aneurysms in association with FMD varies from 20% to 50%; women are more frequently affected than men, and the aneurysms, often multiple, predominate in the supraclinoid ICA. The prognosis is often poor in relation to a high frequency of spasm.[22,26,33,34]

Recently a metaanalysis was performed using data from 17 previously reported series of patients with ICA and/or VA FMD, and data from a new retrospectively evaluated series; after eliminating the selection bias of patients presenting because of symptoms caused by an aneurysm, they found a 7.3% prevalence of incidental asymptomatic cerebral aneurysms in patients with ICA and or VA FMD, much lower than the prevalence previously reported in the literature.[35]

Finally, FMD, especially in its intracranial location, should be considered as a possible cause in all cases of childhood stroke;[36,37] therefore, the need for an extensive specific investigation in such patients should be emphasized.[3,11,38]

Classification

Pathologically, FMD has been classified according to which arterial lamina is mainly involved. The majority of the cases involve fibroplasia of the media. Fibroplasia of the intima and adventitia occur only rarely.[3]

Three types of FMD have been recognized and classified as follows:

1. Medial, the most common type, consisting of medial hyperplasia and muscle-cell proliferation; this type affects predominantly women and may be associated with mural-aneurysm formation
2. Intimal, accounting for approximately 5% of cases, characterized by an increase in the fibrous elements of the intima producing concentric narrowing of the lumen; this type occurs primarily in children and adolescents, affecting both sexes equally
3. Adventitial, the least frequent type, characterized by narrowing of the vessel lumen by fibrous tissue hypertrophy surrounding the vessel wall.[39]

A morphological classification has been proposed based on angiographic features. Three different radiographic patterns have been identified: the most common, the so-called "string of beads" (type 1), is characterized by a chain of arterial dilatations separated from one another by localized strictures; a second pattern, less common, is unifocal and is characterized by tubular stenosis (type 2); a third angiographic type of FMD has been called atypical; where one wall of the affected segment is spared and the opposite wall presents an ovoid diverticulum, sharply circumscribed at either end by a semi-circumferential narrowing.[14,23] A rare type of FMD of the CCA, in the form of a septum, has also been reported in the literature. The lesion was located at the carotid bifurcation just proximal to the beginning of the ICA, and has been correlated with a greater risk for cerebral ischemia than the typical "string of beads" lesion.[40]

Diagnosis

Angiography is considered the gold standard for the diagnosis of FMD. Angiography findings in patients

with FMD typically include irregularly spaced constrictions of the arterial wall alternating with luminal dilatation from fibrous bands, giving the appearance of a "string of beads." This pattern occurs in 80% to 90% of patients with FMD, and medial fibroplasia is the most common histological type of FMD associated with this angiographic finding. Less common and specific angiographic findings include unifocal or multifocal tubular stenosis, occurring in 6% to 12% of cases; these smooth, concentric tubular lesions can be associated with any histological type of FMD. A third angiographic type, called "atypical" by some authors, shows a diverticulum-like formation of the arterial wall and non-circumferential stenosis.[4,10,11,22,41,42] Intracranially, the "string of beads" radiological appearance is the most common finding; the arterial dilatations corresponding to microaneurysms or microsacculations separated from one another by localized strictures and resulting in narrowing of the lumen by approximately 40%. The tubular form, which resembles a long, concentric stenosis, is usually seen in the cervical ICA.

FMD is multifocal, and therefore extensive angiographic studies should always be performed.[3] A four-vessels study is required whenever a dysplasic pattern is found during arteriography. Cerebral views should not be neglected because of a possible association with intracranial aneurysm. When hypertension is found in a patient suffering from carotid dysplasia, renal arteriogram is recommended.[43]

Magnetic resonance angiography (MRA) is a useful, non-invasive modality for the evaluation of narrowing of the carotid artery bifurcation. However, several artifacts known to affect this technique may lead to an appearance similar to that described for FMD. The patient's motion and swallowing may produce focal regions of apparently abnormal signal intensity within the carotid artery that may mimic FMD. Although angiography is the examination of choice, the non-invasive nature of cervical MRA suggests a role in the screening of patients with TIA, follow-up of known FMD, evaluation of potentially affected relatives, and in the study of children. Furthermore, changes suggestive of FMD may be recognized on MRA examinations performed for other indications, such as evaluation of carotid stenosis.[44]

ICAs affected by FMD are often longer than normal. The carotid bifurcation is usually at the C4/5 level or below; when the bifurcation is high, the ICA is frequently coiled or kinked. FMD affects the middle and distal thirds of the cervical portion of the vessel, and the lesions are most commonly seen opposite to the C1/2 vertebral bodies. The disease spares the proximal and most distal portions of the vessel. FMD is usually bilateral, although one side is frequently more advanced than the other. The finding of a carotid dissection at the time of arteriography should lead to examination of the other vessels known to be at risk for FMD.[15]

Carotid duplex ultrasound is a useful screening procedure. The "string of beads" pattern may appear as multiple areas of shadowing corresponding to the sites of stenosis and dilatation seen in the angiogram.[45] Atypical FMD in the form of a carotid web can be detected by carotid ultrasound. Intraluminal webs of the carotid arteries are rare. Atypical FMD is thought by some to represent a variant of intimal fibroplasia, which involves only one wall of the vessel and produces a smooth or corrugated mass that projects into the vessel lumen, creating the angiographic appearance of a web. The diagnosis of a carotid web should be considered in a young patient who is otherwise healthy and has no history of trauma or systemic disease. It is likely that the intraluminal web produces a low-grade obstruction that may disturb the flow and serve as a focus for thrombus formation and subsequent embolization risk.[46] The carotid duplex examination is advisable in patients with equivocal neurological symptoms or asymptomatic bruits in the neck, or who is affected by FMD on other arteries and intracranial aneurysm.[24]

Differential diagnosis

The angiographic differential diagnosis for the "string of beads" lesions includes stationary arterial waves or circular spastic contractions of the extracranial carotid and vertebral arteries. Stationary arterial wave images, also called "pearl necklace" images, are a phenomenon characterized by an exceedingly regular rippling of the intra-arterial contrast column. Circular spastic contractions are more regularly spaced than the FMD, and occur without the dilatation of intervening segments typical of FMD.[3] It is more difficult to establish a differential diagnosis for the rarer tubular stenosing form of FMD, since it must be distinguished from atheromatous stenosis, tubular occlusion due to vasculitis conditions, congenital hypoplasia and arterial spasm.[3,22,39,47]

Atherosclerotic lesions usually involve the proximal 1–2 cm of the ICA or the origin of the vessels; FMD characteristically spares the origins and proximal segments. General examination, clinical history, laboratory studies and neuroradiological findings may help in the diagnostic process.

The angiographic findings in the atypical FMD are non-specific, and may be indistinguishable from atherosclerosis or post-traumatic aneurysm of the ICA.[41]

Treatment

FMD poses therapeutic problems because the natural history of the disease is not well known and therapeutic approaches have not been standardized. Therefore, choice of treatment should be highly individualized.

Whether anticoagulant and antiplatelet agents are beneficial in asymptomatic patients is unclear, and preventive surgery in case of asymptomatic FMD is not recommended. A conservative approach seems to be appropriate for asymptomatic patients and patients with aspecific symptomatology. Nevertheless, the potential of cerebrovascular FMD as a cause of neurological disease should not be overlooked. Patients with symptoms associated with TIAs and stroke may benefit from antiplatelet therapy, and anticoagulation may be considered for patients with recurrent focal ischemia.[4,10,43,48]

In the instances of symptomatic lesions, patients at high risk for cerebrovascular accidents, or in those cases where the disease is refractory to medical management, more aggressive therapeutic options can be considered. Procedures such as graduated arterial dilatation, endarterectomy, or bypass interventions can be justified if the arteriographic lesion and a correlation with the symptomatology are undeniable.[10,43] Graduated intraluminal dilation of the ICA proposed by Morris et al.[49] has been reported as a successful, safe and simple procedure in many cases.[24,50–52] This technique, consisting of dilation of the stenotic areas by gentle passage of dilators through transverse arteriotomy performed at the origin of the ICA and after the tortuous segments of the artery have been straightened by traction, has proved effective and is performed through an easy surgical access.[25,50,53,54]

Others suggest surgical procedures of resection of the lesions and arterial reconstruction as the ideal treatment. The option of surgical intervention should be considered in the instance of one or more dysplasic aneurysms, even asymptomatic, because of their high embolic potential and the risk of rupture.[43] Dissecting aneurysms with rupture of the intima are best managed with surgical excision, since intraluminal dilatation is a blind procedure that can lead to rupture of the intima.[55]

A surgical decision in cases of acute dysplasic dissection should be weighed against the 50% of cases that resolve spontaneously under medical treatment, from clot resorption and intimal tear healing. In every case, the state of health of the patient and the accessibility of the lesions are determinants in the management decision.

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