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Internal carotid artery fibromuscular dysplasia in arterial hypertension: Management in clinical practice

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Abstract
Fibromuscular dysplasia (FMD) reminds of a rare form of secondary arterial hypertension occurring in young people and involving the renal arteries. FMD may also involve vertebral, subclavian, mesenteric, iliac arteries and carotid arteries. FMD of internal carotid arteries is a rare finding that is frequently incidental and asymptomatic. It usually occurs in middle-aged women and is secondary to media-intima fibrodysplasia. The carotid artery may be elongated or kinked and associated cerebral aneurysms have been reported. Symptoms including transient ischaemic attack or stroke are uncommon and are related to decrease of blood flow or embolization by platelet aggregates. At the onset, differential diagnosis with vasculitis must be placed. Computed tomography or magnetic resonance imaging (MRI) angiography demonstrates bilateral high-grade stenosis with the characteristic ''string of beads'' pattern. Antiplatelet medication is the accepted therapy for asymptomatic lesions. Graduated endoluminal surgical dilation is an outmoded therapy, no longer used in most medical centres. Current percutaneous angioplasty is the preferred treatment for symptomatic carotid FMD, but no randomized controlled trials comparing this methodology with surgery is available. The management of a case of arterial systemic FMD in a 52-year-old women, diagnosed after a hypertensive crysis, is discussed. Imaging methods disclosed stenoses of carotid arteries, of celiac tripod and of superior mesenteric artery. Because of high risk associated to endovascular surgery, medical therapy was started. In the first year of follow-up, no events have been reported.

Key Words: *Carotid artery, fibromuscular dysplasia, hypertension, ultrasonography*

Introduction

First described by Connett & Lansche in 1965 (1), fibromuscular dysplasia (FMD) of internal carotid artery is usually asymptomatic and frequently incidental (2). It occurs in middle-aged women and is secondary to medial or intimal fibrodyplasia; it affects several centimetres of middle carotid artery and is bilateral in 50% of cases. Its angiographic prevalence is 0.5–0.7% (3). Cigarette smoking and history of arterial hypertension are associated with an increased risk of this condition (2).

On imaging, carotid artery may be elongated or kinked, and associated ipsilateral intracranial aneurysms have been reported (3). FMD occurs in association to various connective tissue disorders, and is assumed to be a non-specific entity. Its pathogenetic background is so far unknown, but there are indications that it is related to vasa vasorum paucity, repeated microtrauma, inadequate hormonal background or α-anti-trypsin deficiency (4). The natural history of incidental lesions is unknown, but progression has been reported in about 25% of patients (5). Symptoms including transient ischaemic attack, stroke and subarachnoidal bleeding are uncommon and are related to decreased blood low flow or embolization of platelet aggregates (6).

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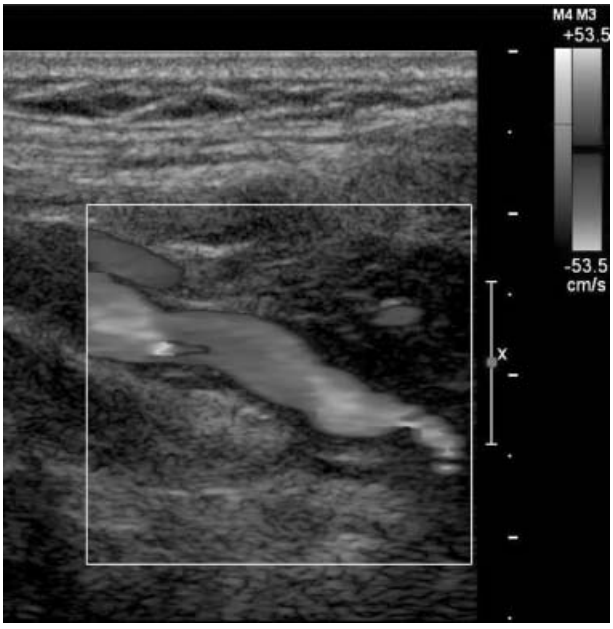

Figure 1. Carotid duplex ultrasonography of the right internal carotid artery shows alternating regions of lumen narrowing and vessel dilation several centimetres distal to the carotid bifurcation.

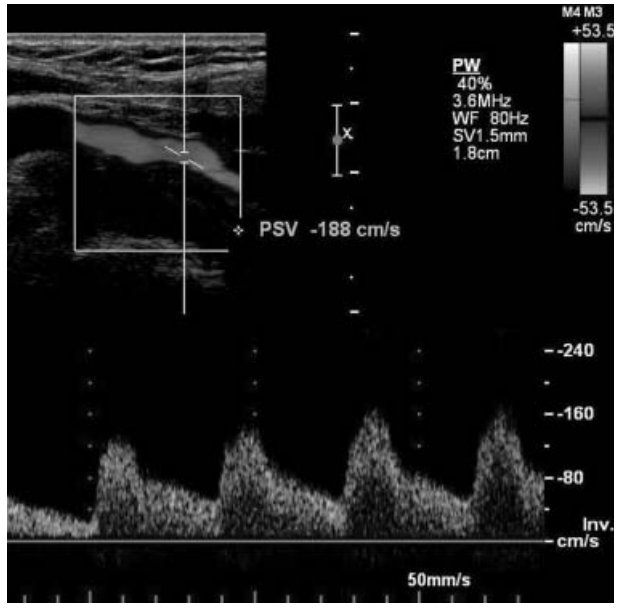

Figure 2. Eco-colour Doppler of right internal carotid artery.

Case report

A 52-year-old woman was admitted to our Hypertension Centre for a hypertension crisis accompanied by headache, palpitation and diaphoresis. She was a moderate smoker and reported a family history of arterial hypertension and cerebrovascular morbidity. She denied any history of diabetes and neurological symptoms. Three days before admission, she had slight temperature and suffered from an episode of temporal pulsatile migraine (right side) associated with a transient right ocular pain followed by diplopia and fugax amaurosis. On this occasion, her physician had found slight increase of systolic and diastolic blood pressure (i.e. 152/94 mmHg) and prescribed nimesulide (50 mg twice daily) for migraine; the patient immediately decided to stop smoking.

At physical examination, she presented with slight palpebral ptosis and lacrimation (right eye), bilateral carotid bruit and blood pressure (BP) of 202/128 mmHg without difference between the arms. Intravenous administration of an α-blocker drug (a bolus of urapidil 50 mg iv, followed by infusion of 25 mg/h) decreased BP levels within 3–4 h to 160/96 mmHg with relief of headache and diaphoresis. Electrocardiogram and chest X-ray were normal.

Screening tests for secondary forms of arterial hypertension such as measurement of plasma renin activity, plasma aldosterone, 24-h urinary free cortisol and metanephrines were negative. Laboratory tests showed a slight normocitic anaemia (haemoglobin 11.2 g/dl) and hypercholesterolaemia (245 mg/dl). Antinuclear antibodies (ANA) were increased (title 1:140), while extractable antinuclear antibodies (ENA), antineutrophil cytoplasmic antibodies (ANCA), C reactive protein, anti-phospholipid antibodies, rheumatoid factor, C3 and C4 complement fraction and cryoglobulins were normal.

Neck ultrasound of the extracranial vessels showed two hypo-echogenic plaques determining an eccentric stenosis of 60% and 65% at the origin of the right and the left internal carotid arteries respectively. The distal limit from plaques beginning was undetectable and operator concludes as a picture like to arteritis (Figures 1 and 2).

During the first 3 days in hospital, her BP was 148/94 mmHg. On the fourth day, the patient underwent carotid-computed-tomography (CCT) angiography of the neck and steroid (prednison 25 mg daily) and antiplatelet medication (acetylsalicylic acid 100 mg daily) were started. Symptoms promptly disappeared, and BP was normalized 2 days after starting therapy with an angiotensin-II-receptor antagonist (olmesartan 20 mg daily) and a diuretic (hydroclorothiazide 12.5 mg daily).

CCT angiography confirmed the presence of bilateral stenosis of the carotid arteries, particularly of the right with the characteristic ''string of beads'' pattern, a morphological feature indicative of FMD (Figure 3).

MRI angiography – extended to mesenteric, renal and iliac arteries – showed a severe stenosis of the celiac tripod and a moderate stenosis of the superior

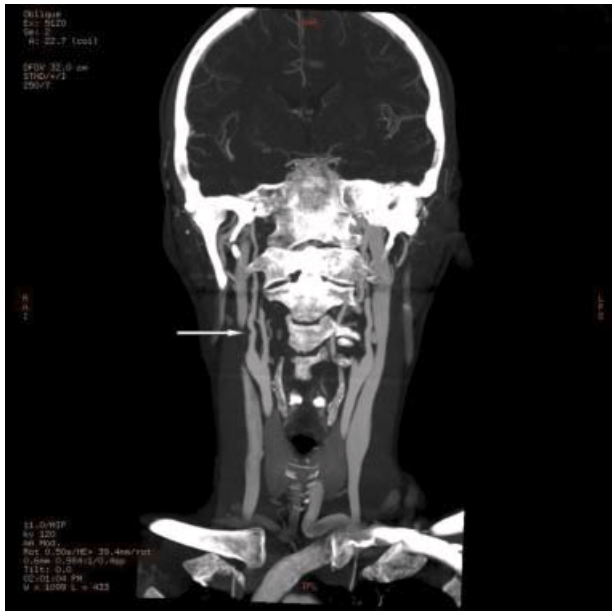

Figure 3. Computed tomography (CT) angiography of extracranial vessels. CT shows irregularity of both internal carotid arteries, particularly of right artery (arrow), with the characteristic "string of beads" pattern.

mesenteric artery. Both renal arteries appeared normal. Cerebral MRI angiography was normal.

Because of the high risk of the endovascular carotid surgical treatment (7) we decides to placed the patient on antiplatelet medication; steroids were stopped and anti-hypertensive therapy was continued. Patient is being now followed up annually with MRI angiography of the extracranial vessels; in the first year of follow-up, no symptoms of angina abdominis were reported.

Discussion

In our patient, the hypertension crisis that brought her to admission into hospital may have been related to the high doses of nimesulide and also to the sudden cessation of smoking in the 4 days before our observation.

It is usually not too difficult to differentiate between atherosclerotic and FMD lesions; the former generally occur at the origin or proximal portion of the carotid artery in older subjects with classic cardiovascular risk factors, while the latter occur in middle or distal carotid artery in young subjects with few cardiovascular risk factors.

On the contrary, it may be difficult to distinguish FMD from vasculitis. FMD is by definition a non-inflammatory disease and is therefore not associated with anaemia or other abnormalities of the acute phase of inflammation. In our case, the presence of fever, anaemia, temporal migraine and ocular disturbances could have suggested Horton's arteritis, although in this particular form of arteritis, carotid involvement is uncommon (2), i.e. instead of frequent in Takayasu's arteritis (8). Increase of ANA suggests an immune mechanism for FMD; moreover, that FMD might be a phenotypic expression of a generalized activation of immune system warrants further investigations.

Antiplatelet medication is the current therapy for asymptomatic FMD. For symptomatic FMD, the distal location of the lesions precludes standard carotid endarterectomy. Graduated endoluminal surgical dilation is an outmoded therapy that no longer is used in most medical centres. Currently percutaneous angioplasty is the preferred treatment for symptomatic carotid FMD but there have been no randomized controlled trials comparing this methodology with surgery (2). For this reason, the main surgical vascular centres prefer conservative treatment at least in subjects with minor events.

Conclusion

Ultrasound evaluation of the carotid arteries has become quite common, particularly in elderly subjects suffering from hypertension or with cerebrovascular symptoms. The case herein presented suggests the opportunity for ultrasound evaluation of the carotid arteries also in middle-aged subjects suffering for the same condition and even in those with family history of arterial hypertension and cerebrovascular diseases. In fact, had ultranosography been performed in our patient, carotid lesions could have been easily detected and the cerebrovascular event avoided.

Although duplex ultrasonography has a lower sensitivity than other imaging techniques studies, it is non-invasive and easy available. If discovered in the carotid arteries, FMD should also be investigated in other vascular beds, as it has been reported at autopsy studies to be the cause of some sudden deaths in young people (9).

Declaration of interest: The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

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