Spontaneous Dissections of the Carotid Arteries in a Patient with Recent Coronary Artery Dissection

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Dear Sir,

Multiple dissections of the carotid and coronary arteries are exceptional [1]. We report the case of a young woman without known hereditary connective tissue disorder who presented with spontaneous dissection of the left anterior descending coronary artery, followed by spontaneous dissection of both internal carotid arteries (ICA) 3 weeks later. We discuss the pathogenesis of these multiple dissections.

Case Report

A 44-year-old right-handed woman was admitted to the University of Lyon stroke unit on June 27, 2006, because of a right hemiparesis with aphasia which were present on awakening. Vascular risk factors were limited to obesity (body mass index = 32). The medical history revealed anterior wall myocardial infarction related to spontaneous dissection of the left anterior descending artery 3 weeks before, confirmed by a coronary angiography (fig. 1). She had been treated by coronary angioplasty with a drug-eluting stent (taxus) and had been discharged from the cardiological intensive-care unit under clopidogrel 75 mg daily, aspirin 75 mg daily, bisoprolol 1.25 mg twice daily, ramipril 2.5 mg twice daily, eplerenon 25 mg daily and

pravastatin 20 mg daily. Post-myocardial-infarction echocardiography had demonstrated anteroapical hypokinesia with low left ventricular ejection fraction (50%). On admission, right hemiparesis and aphasia had cleared. The patient had no history of recent infection (no fever, no flu, no respiratory tract symptoms) and reported no prior transient neurological deficit, headache or cervical pain. No Horner's syndrome was observed. The patient had no skin, joint, eye or skeletal abnormalities suggesting a hereditary connective tissue disorder. No family history of arterial dissection or valvular heart disease was noted. Multimodal MRI performed on June 27 showed no recent ischemic lesion on diffusion-weighted imaging. T₂-weighted sequences showed bilateral middle cerebral artery/anterior cerebral artery borderzone hyperintensities. Magnetic resonance angiography showed occlusion of the right ICA beginning 1 cm after the bifurcation and narrow stenosis of the left ICA in the subpetrous segment consistent with the diagnosis of bilateral spontaneous ICA dissection (fig. 2). Collateral supply was efficient through the right posterior communicating artery. No intramural hematoma was observed. No angiographic changes of fibromuscular dysplasia were found. Neck

ultrasound was suggestive of right ICA occlusion and left distal ICA stenosis. Right ophthalmic artery flow was reversed. Vertebral arteries were normal with increased compensatory flow. Erythrocyte sedimentation rate and C-reactive protein level were normal. Intravenous unfractionated heparin (18,000 IU daily aiming at doubling the partial thromboplastin time) was started on June 28 substituted with oral anticoagulation therapy (acenocoumarol 3 mg daily with a target international normalized ratio of 2.0–3.0) on July 4. Control transthoracic and transesophageal echocardiography performed on July 3 showed no persistent hypokinesia. Left ventricular ejection fraction had returned to normal (68%). No aortic arch atheroma was observed. Aortic root diameter was 35 mm. Grade 1 mitral valve insufficiency was present. The patient was discharged from the stroke unit on July 6. The clinical follow-up at 4 months showed normal neurological examination. No recurrent cardiac or cerebral ischemic event had occurred. Control MRI at 4 months showed no recent ischemic lesion and the persistence of the right ICA occlusion while the left ICA had returned to normal (fig. 3). Oral anticoagulation therapy was stopped whereas dual antiplatelet therapy (clopidogrel

Fig. 1. Coronary angiography showing dissection of the left anterior descending artery (arrow).

Fig. 2. Baseline magnetic resonance angiography showing occlusion of the right ICA and narrow stenosis of the left ICA in the subpetrous segment.

Fig. 3. Control magnetic resonance angiography at 4 months showing the persistence of the right ICA occlusion while the left ICA has returned to normal.

75 mg daily and aspirin 75 mg daily) was maintained in order to prevent coronary stent thrombosis.

Discussion

Spontaneous dissections of the carotid and vertebral arteries account for 10–25% of ischemic stroke in young and middle-aged patients [2]. Most dissections are ‘spontaneous’ while minor or ‘trivial’ traumas are observed in approximately 40% of cases [3]. Patients with spontaneous carotid dissection are thought to have an underlying structural defect of the arterial wall, although only a minority present a well-characterized heritable connective tissue disorder (Marfan’s syndrome, Ehlers-Danlos syndrome type IV, autosomal dominant polycystic kidney disease or osteogenesis imperfecta type I).

We considered that the prior medical history and the MRI data (location and aspect of carotid abnormalities on baseline imaging, recanalization of the left ICA on follow-up imaging) were suggestive of bilateral carotid dissection. The absence of a visible intramural hematoma in our patient may be related to the timing of MRI (the exact time of onset of the carotid artery dissections after the initial coronary

artery dissection is unknown and the intramural hematoma may have disappeared). Bilateral cardiac embolism into both carotid arteries seems unlikely. To our knowledge, the association of carotid and coronary artery dissection has only been reported once [1]. On the other hand, there have been reports of dissections of the aorta [4] or of the renal arteries in patients with previous spontaneous carotid dissections, usually in the setting of fibromuscular dysplasia when both carotid and renal arteries are involved [5, 6].

Spontaneous coronary artery dissection is a rare cause of acute coronary syndrome. Most of the cases occur in young or middle-aged women (80%) without vascular risk factors, especially during the peripartum period. The left anterior descending artery and left main coronary artery are most frequently involved (47 and 17%, respectively). The outcome is considered grim, with a rate of survival as low as 33% of patients [7].

The involvement of coronary and carotid arteries in a short time (3 weeks), which was observed in our patient, points to a ‘systemic’ etiology. Our patient had no history of recent infection, and all markers of inflammation were normal. She denied any use of vasoconstricting or illicit drugs. She also had no history and no clin-

ical signs of hereditary connective tissue disorder. We hypothesize that our patient may suffer from a primary generalized arteriopathy related to an unknown connective tissue disorder that predisposes to structural weakness of the vessel wall and therefore increases the risk of dissection. The morphology of connective tissue elements was found to be aberrant in the majority of skin biopsies from patients with spontaneous cervical artery dissection lacking clinical stigmata of a known connective tissue disorder, pointing to a molecular defect in the biosynthesis of the extracellular matrix [8]. Our patient had evidence of an enlarged aortic root (35 mm) on transesophageal echocardiography, consistent with the existence of a generalized defect of the extracellular matrix. A case-control study has suggested that aortic root diameter enlargement (i.e. diameter >34 mm) is associated with an increased risk of spontaneous cervical artery dissection and that mitral and aortic valve dystrophy and mitral valve prolapse are more frequent in patients with spontaneous cervical artery dissection [9]. Our case report reinforces the concept of the presence of a generalized defect of the extracellular matrix in some patients presenting spontaneous cervical artery dissection.

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