Moyamoya disease and systemic sclerosis (MoSys syndrome): a combination of two rare entities - comment to the authors

Sirs,

It is with great interest that we read the article recently published by Wegner *et al*. on the rare coexistence of systemic sclerosis and Moyamoya disease (1). At our center we recently observed the same rare coexistence in a diffuse cutaneous systemic sclerosis patient. In 2008, one of our female patients was diagnosed with early diffuse SSc. ANA was positive at high titer (1/640) but without identification of specificity. She was treated with pulses intravenous cyclophosphamide (6 fortnightly pulses at a fixed dose of 500 mg) in 2009 and with rituximab (2 fixed doses of 1000 mg with a 2-week interval) in 2010 (with retreatment after 6 and 18 months) because of rapidly progressive cutaneous involvement and interstitial lung disease (ILD). Due to insufficient effect, she received autologous haematopoietic stem cell transplantation (HSCT) according to the ASTIS protocol (2) in 2012. In 2013 this patient developed acquired haemophilia A, the first case to occur in an SSc patient (3). She was retreated with rituximab (4 fixed doses of 600 mg with a weekly interval), combined with oral steroids. Because of severe ILD (FVC 41%; VC 52%; TLC 52%; DLCO 31%), she received a double lung transplant in 2014. In 2015, she experienced a sudden onset of dysarthria and left sided facial palsy. She was diagnosed with a stroke which was later confirmed by neuroimaging. Since the patient presented 48 hours after symptom onset no acute stroke treatment could be initiated. Symptoms resolved without sequelae. There was no evidence for cardiac arrhythmia and transthoracic echocardiography was normal. Coagulation tests were normal with no evidence of thrombophilia. Computed tomography (CT) angiographic imaging demonstrated a stenosis in the right carotid artery with complete occlusion of the M1 segment of the right medial cerebral artery (MCA) with a filiform accessible A1 segment of the right anterior cerebral artery (Fig. 1A). CT perfusion mapping demonstrated a clear reduction in the perfusion of the right hemisphere (Fig. 1B). Magnetic resonance (MR) imaging, performed several days after the acute event, revealed an acute putaminocapsulo-caudate vascular infarction with strongly reduced blood flow in the distal right carotid artery and the anterior and medial cerebral artery on MR angiography (Fig. 1C-D). MRI angiography performed 1 year later showed normal findings on the left carotid T segment, but progressive narrowing of the right supraclinoid carotid artery, the A1 segment of the anterior cerebral artery and the M1 segment of the middle cerebral artery (Fig. 1E-F). As Moyamoya syndrome typically occurs bilaterally, we considered these findings compatible with a Moyamoya phenomenon, or Moyamoya-like disease, as was the case in the 2 previously published cases occurring in limited systemic sclerosis patients (4, 5) and the case presented in this journal by Wegner et al (1). This is the second case of Moyamoya-like disease to occur in the context of diffuse cutaneous systemic sclerosis. We agree with the authors that Moyamoya-like disease should be considered as a potential cause of ischaemic events in SSc patients.

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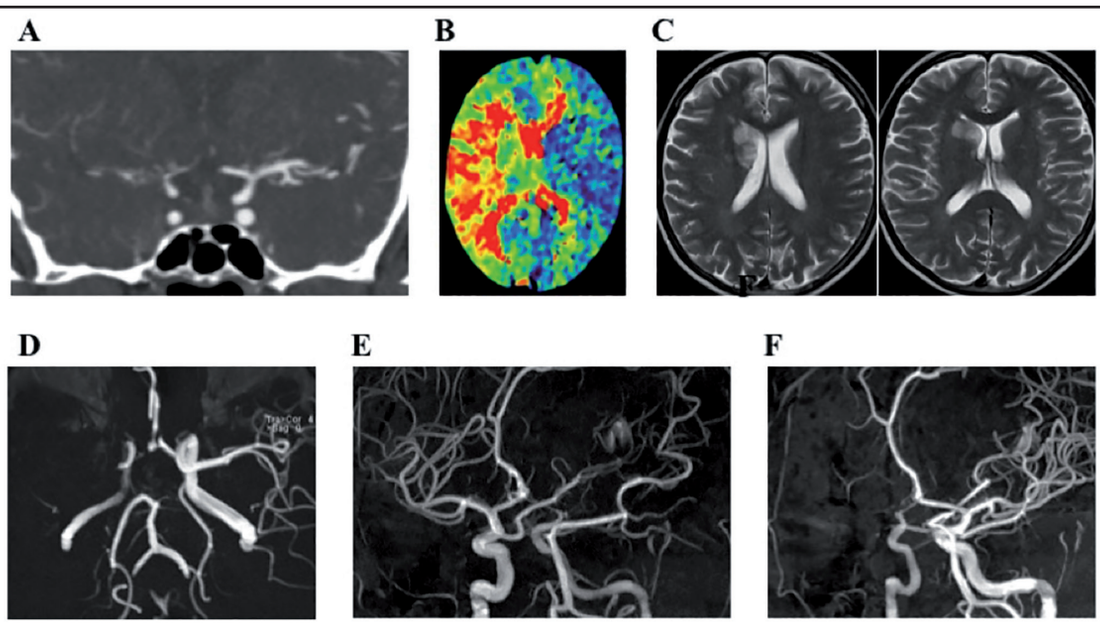

Fig.1. (**A**) Computed tomography (CT) angiographic imaging demonstrating a stenosis of the right carotid artery with complete occlusion of the M1 segment of the right medial cerebral artery. (**B**) CT perfusion mean transit map, highlighting the reduced right hemispheric perfusion. (**C**) Magnetic resonance (MR) imaging with a putaminocapsulo-caudate vascular infarction. (**D**) Magnetic resonance angiography few days after the acute event with reduced blood flow in the distal right carotid artery and the anterior and medial cerebral artery. (**E**) Magnetic resonance angiography 1 year after the acute event: oblique 3D reformated view of the normal carotid T segment on the left side. (**F**) Magnetic resonance angiography 1 year after the acute event: oblique 3D reformated view of the carotid T segment on the right side showing progressive narrowing of the supraclinoid carotid artery, the A1 segment of the anterior cerebral artery and the M1 segment of the middle cerebral artery.