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Imaging and Case Report

Coronary Artery Disease in a Young Adult With Unilateral Moyamoya Disease

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Moyamoya disease (MMD) is a progressive vascular disorder of the brain characterized by progressive stenosis of the internal carotid arteries and their proximal branches.[1] Reduced blood flow in these vessels leads to the development of collateral vessels, and this characteristic of collateral circulation, which resembles something hazy, like a puff of smoke, was termed as "moyamoya" in Japanese.[1,2] Genetic factors have been implicated in the pathogenesis of MMD, with genome-wide and chromosomal analyses identifying an association of chromosomes (3p24-262, 8q233, 6q244, and 17q255) with MMD[3] and a moyamoya susceptibility gene, *RNF213*, identified in Japanese patients.[4]

Case report

A 32-year-old Asian woman with no previous medical history presented with acute-onset left hemiplegia, facial droop, and aphasia and was found to have acute posterior circulation and right middle cerebral artery territory infarcts, for which she was taken to the catheterization laboratory for thrombectomy. During magnetic resonance imaging, the patient was found to have unilateral MMD (Figure 1A-C). The patient complained of chest pain, with mild elevation of the level of troponin (0.092 ng/mL; upper normal value, <0.055 ng/mL). A complete lipid profile showed a total cholesterol level of 115 mg/dL (reference range, <200 mg/dL), high-density lipoprotein level of 30 mg/dL (reference range, >40 mg/dL), low-density lipoprotein level of 43 mg/dL (reference range, <130 mg/dL), and lipoprotein A level of 42.3 mg/dL (reference range, <30 mg/dL). A transthoracic echocardiogram showed a reduced ejection fraction of 35%, with mid and apical left ventricular wall dyskinesis, suggestive of stress cardiomyopathy. Poor R wave progression and negative T waves in the anterior leads were evident in the electrocardiogram. A coronary computed tomography scan was indicative of significant narrowing of the ostium of the left anterior descending artery (LAD) and moderate narrowing of the mid LAD.

The patient subsequently underwent coronary angiography, which revealed severe narrowing of the ostial LAD and at least moderate narrowing of the mid and distal LAD (Figure 1D). Intravascular ultrasound displayed significant plaque burden and calcification in the ostial and mid LAD and a deep myocardial bridge with a mild atheromatic plaque burden in the distal vessel (Figure 1E-G). Based on these findings, percutaneous coronary intervention was performed with the implantation of a 3.0 × 12-mm^2 drug-eluting stent in the ostial LAD and a 2.25 × 26-mm^2 drug-eluting stent in the mid LAD (Figure 1H-J). The patient was discharged after several days of hospitalization with a plan for neurosurgical treatment of MMD later.

Discussion

MMD is a rare cerebrovascular condition that predisposes patients to stroke and is infrequently associated with coronary artery disease (CAD). CAD is usually due to smooth muscle hyperplasia and not due to atherosclerosis, as was evident in the present case. These patients also need to undergo genetic testing for major susceptibility genes because these have been implicated in MMD and extracranial arteriopathy.[5] Furthermore, recent studies have suggested that there is an embryologic background associating MMD and CAD that involves the cephalic neural crest (NC), the "vascular neurocristopathy" hypothesis.[6,7] As such, the cephalic NC, including cardiac NC during embryogenesis, contributes to the concomitant manifestation of vascular diseases in the cardiovascular region, including pathogenesis of MMD. Although it is an intriguing theory, it is still not validated, and further research is needed.

Conclusion

Atherosclerotic CAD, although rare, can present in young patients with MMD and should be high on the exclusion list in these patients.

Declaration of competing interest

The authors declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Keywords: coronary artery disease; intravascular ultrasound; moyamoya; percutaneous coronary intervention.

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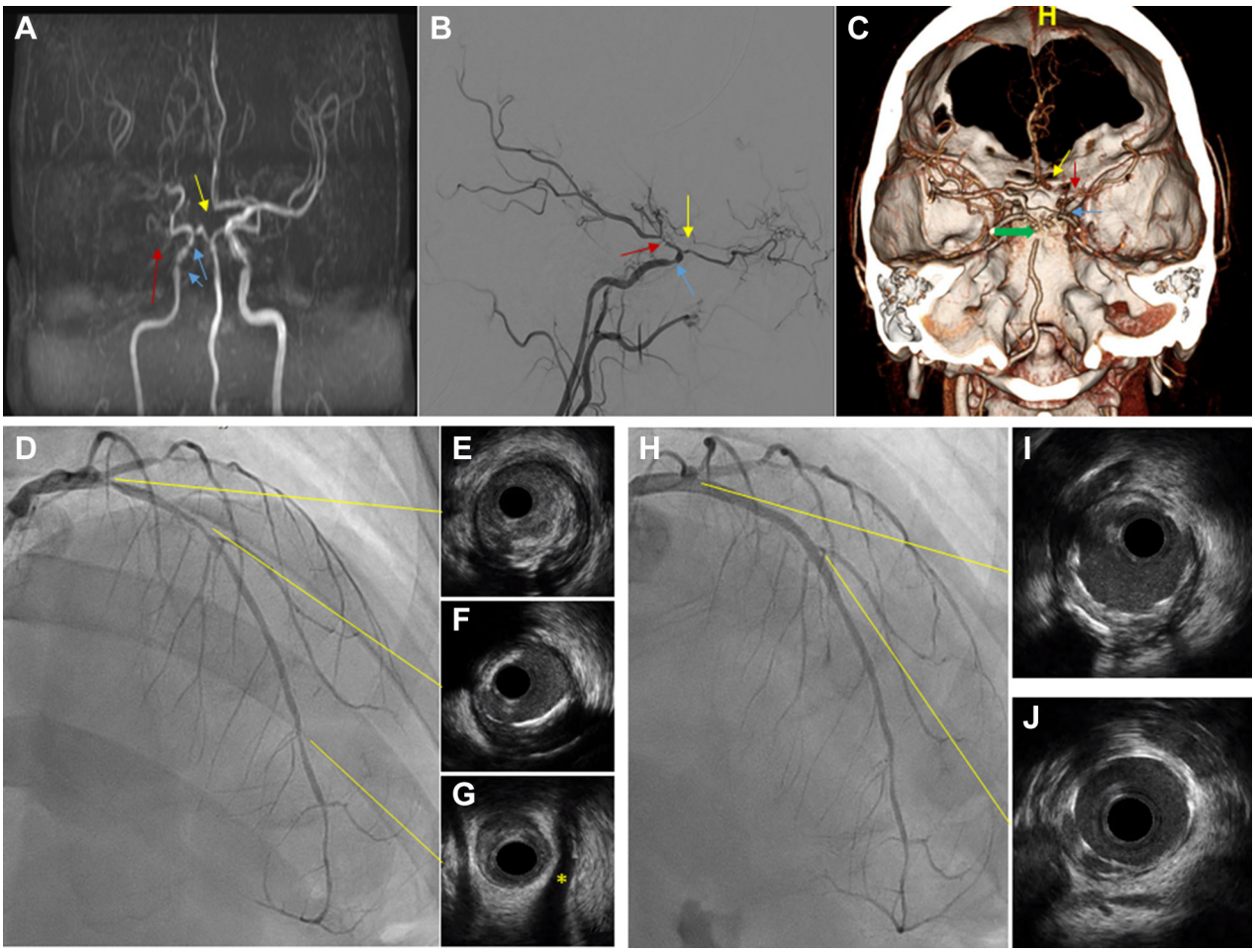

Figure 1. Severe steno-occlusive disease of the distal right internal carotid artery (blue arrows) with diminutive anterior cerebral artery (yellow arrow) and M1 segment of the middle cerebral artery (red arrow) with supply of the distal segments via collaterals and basilar artery occlusion (green arrow); as seen in time-of-flight magnetic resonance angiography (**A**), lateral internal view cerebral angiogram (**B**) and 3D volume rendering (**C**). Coronary angiogram showing severe disease in the ostium, mid, and distal LAD (**D**); co-registered IVUS images of the respective lesion locations (yellow asterisk depicts myocardial bridge) (**E-G**). Coronary angiogram after percutaneous coronary intervention (**H**) and respective IVUS images in the ostial (**I**) and mid (**J**) LAD showing optimal stent expansion and apposition. IVUS, intravascular ultrasound; LAD, left anterior descending artery.

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Ethics statement and patient consent

The research conducted adheres to the ethical guidelines as outlined by the University's policies. Patient consent was obtained to use the images in this case report.

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