Bilateral carotid lesions with rich collateral vascular networks are characteristics of Moyamoya disease. Renal and coronary artery stenoses may be coexistent with Moyamoya disease. We report a female patient with myocardial infarction and a previous history of Moyamoya disease and pituitary gigantism. Upon reviewing the literature, we found no previous reports of myocardial infarction associated with Moyamoya disease, especially in young female patients.

Case Report

A 19-year-old female patient was referred from the Department of Endocrinology for further evaluation of chest pain and dyspnea. She had a history of Moyamoya disease at age 5 years and with pituitary gigantism at age 13. She complained of dyspnea, and had New York Heart Association functional class III and chest pain at rest. Her blood pressure was 120/80 mmHg. Her height was 167 cm (97th percentile) and her face was coarsely enlarged with an associated voice change. The third and fourth heart sounds, both with basal rales, were heard on chest auscultation. The chest X-ray taken on admission showed marked cardiomegaly with pulmonary edema. An electrocardiogram demonstrated anterior myocardial infarction. Laboratory tests revealed a normal lipid profile and normal cardiac enzymes with creatine kinase (CK)-MB of 21 U, myoglobin of 21.7 ng/ml, and Troponin-T and Trononin-I of 0.02 and 0.52 ng/ml, respectively. The erythrocyte sedimentation rate was 35 mm/h. Her serum level of growth hormone was increased to 14.5 μg/L and was not suppressed during an oral glucose tolerance test. She was given 100 μg octreotide subcutaneously 3 times daily, but her blood glucose, ACTH, TSH, LH, FSH and cortisol levels did not change. An echocardiogram showed marked dilatation of the left ventricle with severe septal hypokinesia and 20% ejection fraction (EF). A strongly enhanced, round mass in the pituitary region was demonstrated on coronal brain computed tomography (CT) scan. The right internal carotid artery was occluded abruptly at the proximal portion, and...
its area was supplied by collateral flow from the right external carotid artery and the vertebral artery via extensively developed dural, leptomeningeal and pial collaterals. The left internal carotid artery was also occluded, with a tapering pattern of the proximal portion, and its area was supplied by the external carotid artery and vertebral artery via inumerable small collaterals (Fig 5A–D). These findings were consistent with Moyamoya disease. Stress single photon emission computed tomography (SPECT) revealed extensive fixed perfusion defects with a partially reversible myocardium in the territory of the left anterior descending (LAD) artery (Fig 6A). Critical eccentric stenosis was demonstrated in the middle LAD artery on diagnostic coronary angiogram (Fig 7A). We performed percutaneous transluminal coronary angioplasty and placed a stent at 12 atm. No significant residual stenosis remained after stenting (Fig 7B). The patient’s subjective symptoms were relieved after coronary intervention and the perfusion defect area was decreased on follow-up SPECT (Fig 6B).

Discussion

Coronary artery disease is very rare in young female patients without risk factors for atherosclerosis. A few young female patients with ischemic heart diseases have been diagnosed with associated Kawasaki’s disease and Takayasu’s arteritis, especially in Eastern countries.7–16 However, there has been no previous report of myocardial infarction associated with Moyamoya disease. The present patient had occlusion of both internal carotid arteries, and critical stenosis in the coronary artery, but previously reported cases of Moyamoya disease have not had clinical symptoms of ischemic heart disease.4–6 In the present case, no coronary risk factors were evident, not even hypertension despite a long history of pituitary gigantism. The etiology of the myocardial infarction might be Moyamoya disease because there were no risk factors for coronary
Atherosclerosis. The patient's blood glucose did not elevate even after a stimulation test and all of the coagulation tests were within normal limits. A viable myocardium was documented on stress SPECT with improvement of the EF seen on the 3-dimensional image. Because of the severe left ventricular dysfunction, we infused low-dose dopamine and prepared a provisional intra-aortic balloon pump for the prevention of acute heart failure during coronary intervention. The patient's EF was below 20% before coronary intervention and improved to 30% immediately after intervention. Her chest pain was relieved and the perfusion defect area was seen to be decreased on stress SPECT after coronary intervention, with the pattern of reserved perfusion defect. The patient did not have angina or dyspnea at the 2-month clinical follow-up.
Fig 6. (a) Stress SPECT revealed extensive fixed perfusion defects with a partially reversible myocardium in the area of the left anterior descending artery. (b) The perfusion defect area was decreased on follow-up SPECT after coronary intervention.

Fig 7. (a) Critical eccentric stenosis was demonstrated in the middle left anterior descending artery on diagnostic coronary angiogram. (b) No significant residual stenosis was evident after coronary stenting.
References