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SYSTEMIC LUPUS ERYTHEMATOSUS INITIALLY PRESENTED WITH MOYAMOYA DISEASE

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The term moyamoya was used to describe characteristic patterns of collateral blood vessels in the cerebral circulation that occurs in response to occlusion of the arteries at the base of the brain. The etiology of moyamoya disease has not been fully elucidated, but hereditary and immunogenic factors have been suggested. Some reported a case of recurrent TIA and unilateral moyamoya vessels with lupus anticoagulant, but there has never been reported a case with bilateral moyamoya disease followed by systemic lupus erythematosus (SLE).

A 17-year-old girl was admitted for right hemiplegia and visual disturbance, which developed seven months earlier. Neurological examination showed deep tendon reflex (grade III), and right hemiparesis (grade V). Ophthalmologic examination showed left homonimous hemianopsia. On the third day of admission, malar rash was seen, and multiple joint pain and swelling developed on both proximal interphalangeal, wrist, knee, and ankle joints. Laboratory examination revealed that white blood cell count was 2770/mm^3 and hemoglobin was 9.7g/dl. IgG was 1520mg/dL and complements were decreased (C3, 57.0mg/dL; C4, 10.0mg/dL). Cryoglobuline was positive and rheumatoid factor was negative. ANA were detected at a titer of 1: 2560 with nucleolar and cytoplasmic pattern. Anti-dsDNA was 17.6IU/ml and anti-ribosomal P was positive. VDRL test, anticardiolipin antibody, and lupus anticoagulant were negative. Skin biopsy showed perivascular lymphocytic infiltration consistent with lupus erythematosus. CSF study was negative. Brain MRI and 4 vessel-angiography revealed the obstruction of both internal carotid arteries and bilateral collateral vessels which were characteristics of moyamoya disease. Steroid pulse therapy was done with much improvement on arthritis, speech and walking.

We report here a case of SLE initially presented with bilateral vascular lesions compatible with moyamoya disease.

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Myocardial infartion associated with antiphospholipid syndrome in a young man who developed systemic lupus erythematosus later

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Myocardial infartion(MI) is an unusual complication of systemic lupus erythematosus(SLE). Although MI may supervene late in the disease, usually as a result of atherosclerosis or coronary arteritis, coronary occlusion with MI is infrequent early in the course of SLE. We describe a case of MI associated with antiphospholipid antibody syndrome in a young man who developed SLE 9 months after MI.

Case report: A 23-year-old male was admitted because of acute anterior chest pain in August, 1997. He denied risk factors for ischemic heart disease. At the time echocardiography showed anteroseptal ischemia and coronary angiography revealed 30% narrowing of left anterior descending artery. Anti-cardiolipin, antiphospholipid antibodies, and lupus anticoagulant were all positive. After percutaneous transluminal coronary angioplasty was taken, he was being treated with angiotensin converting enzyme inhibitor and 100 mg of aspirin. Nine months later he was admitted to our hospital because of fever, myalgia, arthralgia and chest pain of 7 days. Examination showed malar rash and both of PIP joints swelling. Results of laboratory studies included pancytopenia, prolonged aPTT, C3 59.6 mg/dl(normal 80-155), C4 10 mg/dl(normal 13-37), anti-dsDNA >100 u/ml(normal < 7), ANA 1:320 titer in homogenous pattern, positive anti-RNP, anti-cardiolipin IgG 94.9 GPL u/ml(normal < 15), anti-phospholipid IgG 15.5 GPL u/ml(normal < 5). However lupus anticoagulant, VDRL, anti-Sm, Ro, La antibodies were all negative. Kidney biopsy revealed focal proliferative glomerulonephritis(type III) and skin biopsy of malar rash showed positive lupus band test. A diagnosis of SLE was made based on the diagnostic criteria. He was treated with high dose corticosteroid and warfarin and then he was discharged in good condition. He is receiving prednisolone and warfarin daily.