Moyamoya Disease Associated with Positive Findings for Rheumatoid Factor and Myeloperoxidase-Anti-Neutrophil Cytoplasmic Antibody

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ABSTRACT

A 39-year old female suddenly fell into a state of unconsciousness. She had no significant past medical history. A computed tomography scan of the head demonstrated a massive left putaminal haemorrhage with a ventricular perforation, low density areas in the right frontal lobe, corona radiata and occipital lobe. A single emergency burr hole drainage of the haematoma was performed. Bilateral common carotid arteriograms showed stenosis of the right internal carotid artery and a complete obstruction of left internal carotid artery which were both accompanied by moyamoya vessels. The biochemical studies indicated serological positive findings for RF and MPO-ANCA. She was transferred to another hospital for nursing care in a vegetative state on the 163rd hospital day. This case indicates that immunological factors, inflammation or vasculitis might have possibly been associated with the development of either an obstruction or stenosis of the intracranial internal carotid arteries.

La Enfermedad de Moyamoya Asociada con Hallazgos Positivos en Relación con el Factor Reumatoideo y los Anticuerpos Citoplasmáticos Antineutrófilos-Mieloperoxidasa

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RESUMEN

Una mujer de 30 años de edad cayó en estado de inconsciencia. No tenía una historia clínica significativa en el pasado. Una tomografía computarizada de la cabeza, reveló una hemorragia puta-minal izquierda masiva con perforación ventricular, áreas de baja densidad en el lóbulo frontal izquierdo, la corona radiada y el lóbulo occipital. Se realizó un drenaje aspirativo de emergencia para tratar el hematoma. Los arteriogramas de las carótidas comunes bilaterales revelaron una estenosis de la arteria carótida interna derecha y obstrucción total de la arteria carótida interna izquierda, ambas acompañadas de vasos moyamoya. Los estudios bioquímicos indicaron hallazgos serológicos positivos de FR y MPO-ANCA. La paciente ya en estado vegetativo, fue trasladada a otro hospital en el día número 163 de su estancia en el hospital, para recibir atención por parte de las enfermeras. Este caso indica que factores inmunológicos, inflamación o vasculitis podrían haber estado asociados con el desarrollo de la obstrucción o de la estenosis de las arterias carótidas internas intracraneales.

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INTRODUCTION

The aetiology of moyamoya disease is still unknown; however, complications with various diseases such as proteins C or S deficiency (1), sickle cell anaemia, essential thrombocythaemia (2) Down's syndrome (3) Noonan syndrome (4) Marfan syndrome (5) Alagille syndrome (6) fibromuscular dysplasia (7) neurofibromatosis (8) Graves disease (9) systemic lupus erythematosus and Sjögren syndrome (10) have been reported. We herein report the first case of moyamoya disease associated with the seropositivities for both rheumatoid factor (RF) and myeloperoxidase (MPO) anti-neutrophil cytoplasmic antibody (ANCA).

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Case report

A 39-year old female suddenly fell into a state of unconsciousness while at work and was transferred to our department. She had no significant past medical history. On arrival, she showed neither eye opening nor a verbal response,

and a decorticate posture upon painful stimulation, corresponding to a sum score of 5 on the Glasgow Coma Scale. She demonstrated anisocoria of the right pupil. A chest X-ray was negative. A computed tomography (CT) scan of the head demonstrated a massive left putaminal haemorrhage with ventricular perforation and an obscure basal cistern, thus suggesting brain herniation. In addition, a CT scan showed low density areas in the right frontal lobe, corona radiata and occipital lobe (Figure 1). No mass or hypertrophy of the

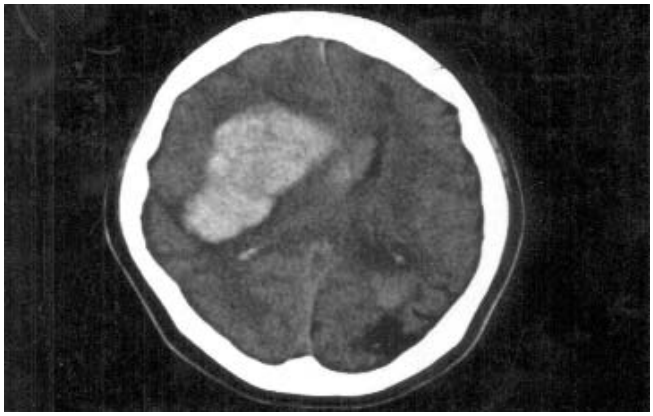

Fig. 1: Head CT on arrival
The CT demonstrates massive left putaminal haemorrhage with ventricular perforation. In addition, the CT scan shows low density areas in the right frontal lobe, corona radiata and occipital lobe, thus suggesting previous ischaemic insults due to haemodynamic insufficiency.

mucosa was identified in the paranasal cavities. Single emergency burr hole drainage of the haematoma under ultrasound guidance via the right frontal lobe was performed. The abnormality of the pupils improved after the operation, however, the patient's unconscious state continued. Bilateral common carotid arteriograms on the 17th hospital day showed stenosis of the right internal carotid artery and a complete obstruction of left internal carotid artery which were both accompanied by moyamoya vessels (Fig. 2). The results of biochemical studies for detecting concomitant diseases with moyamoya disease were as follows: CRP, 0.3 mg/dl; CH50, 66.9 U/ml (normal, 30–50); C3, 176 mg/dL (normal, 65–135); C4, 44 mg/dL (normal, 13–35); rheumatoid factor (RF), 520 IU/ml (normal range: 0–20), antinuclear antibody, < 1:20, anti-dsDNA, < 1 IU/ml, lupus erythematosus factor, negative; cytoplasmic(C)-ANCA, < 10 EU(normal, < 10); MPO-ANCA, 23 EU (normal, < 20); urinary protein, 0.22 g/day. Human leukocyte associated typing showed A24, A26; B46, B62; CW1, CW3. A urinalysis showed a mild grade of proteinuria but the urinary sediment level was within the normal limits. Repeated re-evaluation of RF and MPO-ANCA were positive. A renal biopsy was not performed because her family did not consent to it. Magnetic resonance imaging of the carotid artery in the skull did not show any perivascular infiltrate. She was transferred to

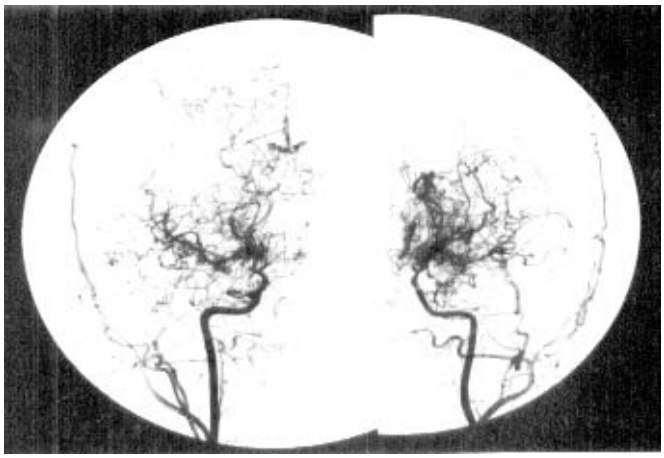

Fig. 2: Bilateral common carotid arteriograms on the seventeenth hospital day. The angiograms show stenosis of right internal carotid artery (2a) and a complete obstruction of the left internal carotid artery (2b) with moyamoya vessels.

another hospital for nursing care in a vegetative state on the 163rd hospital day.

DISCUSSION

There have so far been few reports on moyamoya disease associated with autoimmune diseases and/or status (10, 11). Paciaroni *et al* (11) first reported on moyamoya disease associated with rheumatoid arthritis (RA) (11). They described a 74-year old woman with active RA and atherosclerosis who had moyamoya vessels on angiography. They hypothesized that an inflammatory process of RA and atherosclerosis might have contributed to the formation of an obstruction of the internal carotid artery. We previously re-ported a 30-year old woman who had unilateral moyamoya disease associated with systemic lupus erythematosus and Sjögren's syndrome (10). Since the deposition of immunoglobulins in the intima of the internal carotid artery and subendothelial infiltration of T-cells were revealed in immunohistochemical studies of the vascular lesion in some cases of moyamoya disease, immunological factors are therefore considered to possibly play a role in the pathogenesis of this disease (10).

ANCA are sensitive and specific markers for ANCA-associated systemic vasculitides. C-ANCA is preferentially associated with Wegener's granulomatosis. In contrast, MPO-ANCA is associated with several vasculitides, such as microscopic polyangiitis, idiopathic necrotizing crescentic glomerulonephritis or Churg-Strauss syndrome (12). ANCA with different and unknown antigen specificities are also found in a variety of conditions other than ANCA-associated systemic vasculitides, including inflammatory bowel diseases, autoimmune diseases and infections 12. In addition, ANCA has also been reported to occur incidentally in large vessel vasculitis 13–15 but the diagnostic significance of these conditions is so far not clear. In this case, a mild grade of proteinuria indicated that she might have had glomeru-

lonephritis. Nevertheless, no immunosuppressive therapy had been necessary in this case since the proteinuria and/or renal dysfunction were not progressive. Other underlying vasculitides were ruled out based on the physiological findings, the blood analyses and radiological studies for the chest, head and face.

To our knowledge, the association of moyamoya disease with serologically positive findings for RF and MPO-ANCA has not been previously described. This case did not have a past history of RA and her joints were all physiologically normal. However, we hypothesized that immunological factors, inflammation or vasculitis might have possibly been associated with the development of either an obstruction or stenosis of the intracranial internal carotid arteries in this case.

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