Dilated Fourth Ventricle on Brain Computed Tomography Scan in the Diagnosis of Chiari I Malformation with Syringomyelia

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A 31-year-old woman noticed weakness of the right upper and lower limbs with numbness in the right arm on waking up one morning. The medical history included surgery for patent ductus arteriosus at the age of 3 and atopic dermatitis at the age of 24. On admission, blood pressure was 122/70 mmHg, pulse rate was 64/min, and body temperature was 36.4°C. Neurological examination revealed distal weakness of the right upper and lower limbs but no muscle atrophy, and the presence of hypesthesia to touch, pain in the right upper limb and hyperesthesia below the right T6 segmental areas, but the deep sensation was intact. The tendon reflexes were generally hyperactive but absent in the right upper limb, with positive Babinski’s sign on the right. Cerebrovascular disease was suspected based on the sudden onset of the focal signs. However, brain computed tomography showed no abnormalities except for a dilated fourth ventricle and lateral ventricles (Picture 1). Chiari I malformation with syringomyelia was diagnosed by spinal magnetic resonance images (Picture 2).

Syringomyelia occurs in 25% to 65% of patients with Chiari I malformations (1). The clinical course is characterized by slowly progressive onset with spontaneous resolution (2), although acute progressive onset triggered by neck...
flexion (3) or cough (4) has been reported also. However, to our knowledge, Chiari I malformation with syringomyelia diagnosed by acute onset without any preceding neurological symptoms have not been reported. In the present case, the patient could not identify any precipitating factor that triggered the onset of neurological disturbance. A single finding; dilatation of the fourth ventricle, preceded the clinical manifestations including neurological disturbances and cerebellar tonsillar herniation in Chiari I malformation with syringomyelia (5), suggesting this finding may be useful for the diagnosis of the onset of Chiari I malformation with syringomyelia. In young patients who develop acute neurological disturbances, Chiari I malformation with syringomyelia should be included in the differential diagnosis of stroke, particularly in a patient showing dilatation of the fourth ventricle.

References