

Ipsilateral hemiparesis and spontaneous horizontal nystagmus caused by middle cerebral artery territory infarct in a patient with agenesis of the corpus callosum

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Abstract Ipsilateral hemiparesis and spontaneous nystagmus have rarely been reported after a cerebral lesion. A 35-year-old man with agenesis of the corpus callosum developed ipsilateral hemiparesis and spontaneous horizontal nystagmus after an infarct in the right middle cerebral artery territory. Magnetic resonance imaging revealed the presence of an acute infarct in the right middle and inferior frontal gyrus, supramarginal gyrus, insular gyrus, internal capsule, head of caudate nucleus, putamen, and globus pallidus. Transcranial stimulation of the hand area of the cerebral motor cortex produced motor-evoked potentials in the abductor pollicis brevis muscle exclusively on the ipsilateral side. No motor-evoked potentials were evoked in the abductor pollicis brevis muscle contralateral to the stimulation. The motor-evoked potentials and magnetic resonance imaging findings in our case suggest that anomalies of the decussation of the corticospinal tracts can be found in agenesis of the corpus callosum.

Keywords Transcranial magnetic stimulation · Motor-evoked potentials · Agenesis of the corpus callosum · Ipsilateral hemiparesis · Nystagmus · Cerebral infarction

Introduction

Previous neuroanatomic studies have firmly supported the notion that the primary motor cortex predominantly innervates the contralateral half of the body. Conversely, some case reports supported the existence of the uncrossed corticospinal tract in exceptional patients [1]. Anomalies of the decussation of the corticospinal tract are mostly non-specific and coincidental, but are frequently found in posterior fossa malformations, such as occipital encephaloceles, the Dandy–Walker malformation, Joubert syndrome, and in cases with extensive malformations of the brain stem such as Möbius syndrome [1]. Furthermore, ipsilateral hemiparesis has rarely been reported after a cerebral lesion [2–5]. We used magnetic resonance imaging, including diffusion-weighted imaging, motor evoked potentials and somatosensory-evoked potentials to investigate a 35-year-old man who developed right-sided hemiparesis and spontaneous nystagmus after right middle cerebral artery territory infarct.

Case report

A 35-year-old man, who had finger deformities and mild mental retardation from a very young age, suddenly developed vertigo, nausea, and mild right-sided hemiparesis. When he tried to walk, he staggered and fell toward his left side. There was no family history of mental retardation or congenital abnormality.

Upon neurological examination, he was alert and had spontaneous right-beating nystagmus, mild right-sided hemiparesis (grade 4 on the Medical Research Council scale), and axial lateropulsion toward the left. The pupils were symmetric and reactive to light. His speech was not

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hoarse. The pinprick sensation was normal. Limb ataxia was absent. The little fingers were clawed. The patient was unable to flex the joints of the index fingers completely when making a fist. Spontaneous right-beating nystagmus and axial lateropulsion disappeared within a couple of days after admission. His blood pressure, serum glucose concentration, and serum cholesterol concentration were normal. Prothrombin time, partial thromboplastin time, protein C and protein S activity, homocysteine level, anticardiolipin and antiphospholipid antibody levels, antithrombin III level, and lactate: pyruvate ratio were normal. Results of tests for antinuclear antibodies and factor V Leiden mutation were negative. An electrocardiogram and a transthoracic echocardiogram revealed no sources of cardiac embolism.

Brain magnetic resonance imaging (MRI), including diffusion-weighted imaging, was performed using a 1.5 T superconducting MAGNETOM Sonata (Siemens) MRI system. The patient had agenesis of the corpus callosum associated with a large interhemispheric cyst. The shape of medulla was unusually flat, and the anterior median fissure was not prominent. Axial diffusion-weighted imaging revealed hyperintensity in the right middle and inferior frontal gyrus, supramarginal gyrus, insular gyrus, internal capsule, head of caudate nucleus, putamen, and globus pallidus. Brain magnetic resonance angiography revealed occlusion of the right middle cerebral artery (Fig. 1).

Transcranial magnetic stimulation was performed using a magnetic stimulator (MagPro Compact, Medtronic) with a circular electromagnetic coil with a diameter of 12 cm. We placed the center of the coil over the scalp at 2 cm posterior to Cz to stimulate the finger area. We used counterclockwise coil current for preferential activation of the left motor cortex and clockwise coil current for preferential activation of the right motor cortex. Electromyographic electrodes were placed on the belly tendons of the abductor pollicis brevis muscle on each side. Stimulation of the left motor cortex evoked responses in the left abductor pollicis brevis muscle with a latency of 23.4 ms; the responses were ipsilateral to the side of the stimuli. No response was evoked in the muscles on the right side. Stimulation of the right motor cortex evoked responses in the right abductor pollicis brevis muscle with a latency of 21.8 ms; the responses were ipsilateral to the side of the stimuli. No response was evoked in the muscles on the left side (Fig. 2). Magnetic stimulations of the posterior neck produced bilateral symmetric motor evoked potentials in the muscles of the hands.

Sensory-evoked potentials were recorded with a standardized electrodiagnostic system (Medelect Synergy, VIASYS HealthCare). Electrical stimulation with square-wave pulses was delivered by a bipolar surface electrode to the right and left median nerve at the wrist, producing a minimal thumb twitch. A total of 450 responses were

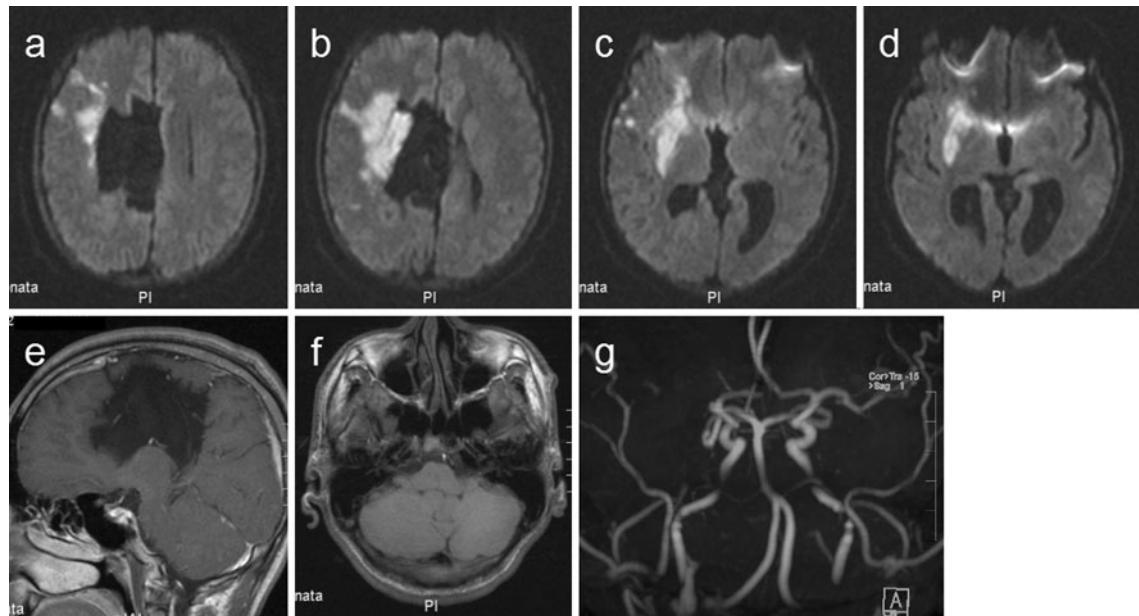
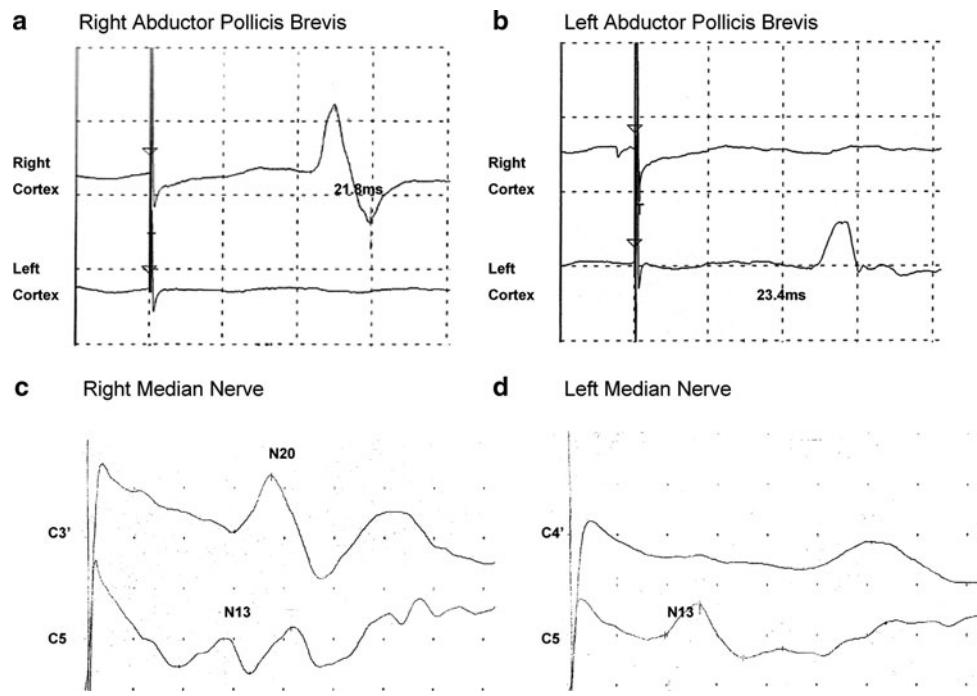


Fig. 1 **a–d** Axial diffusion-weighted imaging showed hyperintensity in the right middle and inferior frontal gyrus, supramarginal gyrus, insular gyrus, internal capsule, head of caudate nucleus, putamen, and globus pallidus. Magnetic resonance imaging revealed agenesis of the corpus callosum associated with a large interhemispheric cyst on

a, b diffusion-weighted images and on **e** a T1-weighted image. **f** The shape of medulla was unusually flat, and the anterior median fissure was not prominent on a T1-weighted image. **g** Brain magnetic resonance angiography revealed occlusion of the right middle cerebral artery

Fig. 2 **a, b** Motor evoked potentials following transcranial magnetic stimulation of the hand area of the cerebral motor cortices and **c, d** sensory-evoked potentials after stimulation of the median nerves. Stimulation of the right motor cortex produced motor evoked potentials **a** in the right abductor pollicis brevis muscle, but **b** not in the left abductor pollicis brevis muscle. Stimulation of the left motor cortex produced motor evoked potentials **b** in the left abductor pollicis brevis muscle, but **a** not in the right abductor pollicis brevis muscle. **c** A cortical potential (N20) was evoked in the left cerebral cortex (C3') after stimulation of the right median nerve. **d** A cortical potential (N20) was not evoked in the right cerebral cortex (C4') after stimulation of the left median nerve



summed and averaged. The recording electrodes were placed according to the international 10–20 system at 3 cm behind C3 and C4 (cortical N20 peak). The active electrodes were referenced to a mid-frontal Fz electrode. After right median nerve stimulation, the primary cortical response N20 was elicited in the left parietal region, which was contralateral to the side of stimulation. After left median nerve stimulation, the primary cortical response N20 was not elicited in the right parietal region (Fig. 2).

His right-sided hemiparesis gradually improved and he was discharged 10 days after admission without hemiparesis. Atherothrombosis was considered an etiologic mechanism of right middle cerebral artery occlusion, because many other rare causes of young stroke could not be identified despite the detailed and comprehensive diagnostic work-up. A diagnosis of Moyamoya disease was unlikely because the steno-occlusive change was confined to the unilateral middle cerebral artery [6]. He was placed on aspirin to prevent another stroke.

Discussion

Our patient is of interest for two reasons. First, he developed ipsilateral hemiparesis after middle cerebral artery territory infarction, and motor-evoked potentials indicate a predominantly ipsilateral innervation of the primary motor cortex. Secondly, the middle cerebral artery territory infarct resulted in vertigo and spontaneous horizontal nystagmus.

Agenesis of the corpus callosum can be accompanied by diminutive corticospinal tracts within the brainstem, causing spastic paraparesis [1, 7]. In men, mutations in the L1CAM gene are responsible for a clinically variable X-linked disorder that has been described as X-linked hydrocephalus, MASA (mental retardation, adducted thumbs, spastic paraparesis, agenesis of the corpus callosum) syndrome, X-linked agenesis of the corpus callosum, or spastic paraparesis type 1 [1, 7]. The most consistent features in affected males are lower limb spasticity, mental retardation, hydrocephalus, and flexion deformity of the thumbs. It is now recognized that together these syndromes represent the phenotypic variability of L1CAM mutations [7]. Perhaps the most striking pathological observation is hypoplasia or apparent absence of two long axonal tracts: the corticospinal tract and corpus callosum [7]. However, there is no anatomical or neurophysiological evidence to support significant failure of decussation of corticospinal axons. Investigators studied males hemizygous for the L1CAM mutation and found that stimulation of the cerebral motor cortices produced motor-evoked potentials in the contralateral biceps brachii muscles in all subjects [7]. Therefore, although our patient did not undergo genetic analysis, we presume that his abnormalities of corticospinal tract decussation are not associated with L1CAM mutations.

The motor-evoked potentials in our patient showed that his cerebral motor cortex predominantly innervates the ipsilateral half of his body. However, motor-evoked potentials cannot visualize the fiber bundles of the corticospinal tract. Diffusion-tensor MRI is a non-invasive,

quantitative imaging modality that is used for tracking white matter fibers and examining anatomical connectivity in the brain [8]. Diffusion-tensor MRI has broadened our knowledge of the brain and of the interrelationships between disruption of white matter integrity and neurological function and its susceptibility to aging and disease [8, 9]. Some investigators reported a patient with congenital left-sided hemiparesis secondary to right closed-lip schizencephaly [10]. In the detailed analysis of diffusion-tensor imaging-based three-dimensional tractography, they observed three different corticospinal tracts at the level of the brainstem. One branch decussated at the medulla, another branch descended ipsilaterally in the cervical cord, and the third branch decussated twice: once at the pons and then again at the medulla. If our patient had undergone magnetic resonance diffusion-tensor imaging-based three-dimensional tractography of the corticospinal tract, we might have confirmed the existence of the uncrossed corticospinal tract in our patient.

Vertigo and nystagmus are most often associated with problems in the semicircular canals, vestibular nerve, or brainstem, although it has also been reported to occur in patients with cerebral cortical lesions caused by infarction or hemorrhage [11–14]. The vestibular cortical system seems to be distributed among several multisensory areas in the parietal and temporal cortices, and is integrated in a larger network for spatial attention and sensory-motor control. The parieto-insular cortex is postulated to be the core region within the vestibular cortical system [11]. Our patient's brain infarction was located in the right insular and parietal cortices. Owing to the abnormal shape of the medulla, we suspect that this patient may have anomalies of the central vestibular pathway as well as the corticospinal tract and corpus callosum. It is not clear as to whether he was more likely to develop vestibular symptoms after middle cerebral artery territory infarct because of the suspected anomalies of the central vestibular pathway.

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