Cerebellar Mutism after Basilar Artery Occlusion
—Case Report—

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Abstract

A 30-year-old female became comatose due to embolic occlusion of the basilar artery, caused by surgical injury to the origin of the vertebral artery during removal of a neurinoma in the upper thoracic paravertebral region. The basilar artery occlusion was treated by local fibrinolysis through a microcatheter. Two weeks later she recovered her consciousness but suffered mutism. Her speech disturbance was characterized by severe ataxic dysarthria known as "cerebellar mutism" but without cranial nerve paresis. The mutism gradually improved during the following 3 months. This is case of cerebellar mutism was apparently due to ischemic stroke. Disturbance by hypoperfusion of the cerebellum and brain stem may have been involved in the pathogenesis of cerebellar mutism.

Key words: basilar artery, cerebellum, fibrinolysis, mutism, stroke

Introduction

Mutism is defined as the inability of a cognitively alert patient without oral apraxia to produce verbal output, despite being able to read and write with intact comprehension. Transient loss of speech after posterior fossa surgery may occur in children, and is categorized as cerebellar mutism. The post-surgical speech disturbance in children is not accompanied by either nuclear or supranuclear lower cranial nerve paresis or long tract signs. The onset of cerebellar mutism may be linked to degenerative disease, hemorrhage, infection, or trauma. We describe a case of cerebellar mutism following occlusion of the basilar artery, which may have been caused by ischemic stroke.

Case Report

A 30-year-old female underwent upper thoracic (left thoracic 2/3 level) surgery on a dumbbell-shaped neurinoma in the paravertebral region. The left supraclavicular approach was employed, cutting the left clavicle and retracting the proximal portion of the left vertebral artery to successfully remove the tumor. Postoperatively the patient awakened immediately, but 2 hours later became comatose. The patient's neurological status was 3 points on the Glasgow Coma Scale, with flaccid tetraparesis and bilateral non-reactive pinpoint pupils. Computed tomography (CT) of the brain and chest showed no abnormalities. We suspected that the embolus from the injured left vertebral artery had occluded the basilar artery, so cerebral angiography was carried out.

Right vertebral angiography demonstrated possible embolic occlusion of the upper basilar artery (Fig. 1). The right posterior (PICA) and anterior inferior cerebellar arteries (AICA), left superior cerebellar artery (SCA), and bilateral posterior cerebral arteries (PCAs) were not visualized. A microcatheter (Tracker-18 catheter; Target Therapeutics Inc., Fremont, Calif., U.S.A.) was introduced into the basilar artery through the right vertebral artery and local fibrinolysis with urokinase was attempted. After local infusion of 960,000 IU urokinase, antegrade flow was obtained in the basilar artery, right PICA-AICA, left AICA-PICA, right SCA, right PCA, and its perforators. Mild hypothermia therapy (body temperature of about 34°C) and barbiturate coma therapy were then performed to protect the brain. However, 2 days after the ictus, suboccipital decompressive...
Coma therapy was discontinued 5 days after the ictus. Although the patient did not speak for 2 weeks post-ictus, she was alert and seemed to understand speech, and could eat and write. Those symptoms suggested disturbance of the cerebellar hemisphere, and superior and/or middle cerebellar peduncle. At this time she had right oculomotor nerve paresis due to right midbrain lesion as well as slight right extremity and trunk ataxia due to cerebellopontine lesion, but did not develop lower cranial nerve paresis. Three weeks later, magnetic resonance (MR) imaging demonstrated long T1 and T2 areas in the bilateral cerebellar hemispheres, thalamus, right midbrain, and bilateral pons (Fig 2). Follow-up angiography taken during the same week as MR imaging was performed demonstrated that the basilar artery, left AICA-PICA, bilateral SCAs, and bilateral PCAs were all patent (Fig 3). Single photon emission CT (SPECT) with technetium-99m-hexamethylpropyleneammine oxime (99mTc-HMPAO) demonstrated low perfusion in the brain stem and bilateral cerebellum (right cerebellar hemisphere 31.6 ml/100 g/min, left 29.4 ml/100 g/min) [Fig 4 upper left]. The cerebral blood flow (CBF) in the cerebral hemispheres and bilateral thalami was low (right thalamus 58.4 ml/100 g/min, left 55.3 ml/100 g/min) [Fig 4 upper right]. These findings indicated a diagnosis of cerebellar mutism. There is no definitive treatment for cerebellar mutism, so the patient was managed conservatively without medication.

One month later, she gradually began to speak. By 3 months after ictus, she had completely recovered from her loss of speech. Right oculomotor nerve paresis and ataxia disappeared at this time. MR imaging showed the same findings as the previous examination for infarction in the bilateral cerebellar hemispheres, bilateral thalami, right midbrain, and bilateral pons. 99mTc-HMPAO SPECT showed increased CBF in the bilateral cerebellum (right cerebellar hemisphere 38.4 ml/100 g/min, left 33.1 ml/100 g/min) and bilateral thalami (right thalamus 63.4 ml/100 g/min, left 65.0 ml/100 g/min) as well as in the cerebral hemispheres compared with the previous CBF examination (Fig 4 lower row).

Discussion

The term mutism is usually defined as complete loss of speech in a conscious subject with no organic lesion of the neuraxis (functional forms), or more rarely with organic lesions of the neuraxis (organic forms). The functional forms are mostly in the area of the psychiatrist (psychosis, autism). The organic forms are subdivided into six types, according to the area of the nervous system affected: a) lesion of...
Fig. 2  $T_1$-weighted spin-echo magnetic resonance images (A, C) and $T_2$-weighted spin-echo images (D, F) 3 weeks after ictus, showing infarction in the cerebellar hemispheres (arrows in A and the shaded areas in B), the bilateral thalami (arrows in C and D and the shaded areas in E), and the pons (arrows in F) and the bilateral cerebral hemispheres (arrowheads in F and the shaded areas in G).

Broca's area (motor aphasia), b) lesion of the supplementary motor area of the dominant hemisphere, c) lesion of the mesencephalic reticular formation (akinetic mutism), d) stereotactic lesion such as those following post-bilateral thalamotomy in parkinsonian patients, e) diffuse bilateral hemispheric lesions (pseudobulbar palsy), and f) bilateral pharyngeal or vocal cord paralysis (peripheral nerve palsy). Cerebellar mutism is yet another organic form.
The common characteristics of cerebellar mutism are absence of long tract signs, absence of supranuclear or nuclear cranial nerve paresis, and an apparently normal state of consciousness.\textsuperscript{1,7,10-18} Our patient responded to orders but could not speak. She could eat and write, and displayed no swallowing disturbance, indicating no lower cranial nerve paresis and no consciousness disturbance. Therefore, the diagnosis was cerebellar mutism.

The pathogenesis of cerebellar mutism is unclear. The highest incidence of dysarthria was found in patients with paramedian or lateral cerebellar hemispheric lesions.\textsuperscript{2} Transient mutism occurred in two patients after stereotactic lesion of the bilateral dentate nuclei for the treatment of a dyskinetic syndrome.\textsuperscript{10} Cerebellar mutism may result from acute bilateral cerebellar injury,\textsuperscript{17} and bilateral interruption of the dentatothalamocortical pathways may be responsible for the development of postoperative mutism.\textsuperscript{5} Loss of speech may be explained by the transient dysfunction of neurons of the dopaminergic cell group in the mesencephalon, because those neurons play a major role in the mesencephalofrontal activating system.\textsuperscript{4} Involvement of median and/or paramedian structures of the cerebellum may also be involved in the mechanism of cerebellar mutism.\textsuperscript{8}

CT and MR imaging of our patient showed infarction in the bilateral cerebellar hemispheres, bilateral pontis, and bilateral thalami, supporting the hypothesis that disturbance of the cerebellum and brain stem including dentatothalamocortical pathways can produce cerebellar mutism. Postoperative spasm of the arteries supplying the cerebellum may cause ischemia, with disturbed cerebellar perfusion or edema leading to cerebellar mutism.\textsuperscript{9,16} In our patient, SPECT showed increased CBF after recovery from the cerebellar mutism, with increased CBF in the cerebellar hemispheres.

During the period of cerebellar mutism in our patient, angiography showed normal filling of the basilar artery, AICAs, SCAs, and PCAs. Complete recovery of speech nearly coincided with improvement of normal CBF in the posterior fossa, excluding the infarct lesions in the bilateral cerebellar hemispheres, pons, and bilateral thalami. This may indicate that hypoperfusion of thalamus, brain stem, and cerebellum is involved in the pathogenesis of the cerebellar mutism. A transient decrease in CBF in the bilateral cerebellar hemispheres or dentate nuclei may cause crossed cerebellocerebral diaschisis and result in dysfunction of the thalamus and supplementary motor area due to transneuronal deactivation through the dentatothalamocortical pathway.\textsuperscript{3,13}

We think that cerebellar mutism may result from
extensive disorder of the cerebellum and brain stem including the dentatothalamicortical pathway. The present case may support the idea that low CBF in the cerebellum and brain stem can produce cerebellar mutism.

Patients with cerebellar mutism will recover their speech within 1 to 6 months. Our patient required 3 months. Dopamine agonists have been effective for the treatment of akinetic mutism, but this medication is not definitively indicated. As we think that low CBF in the cerebellum, thalamus, and brain stem is the main pathogenetic factor leading to cerebellar mutism, medication may not be required if the circulation in the posterior fossa can be restored.

References


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