Bilateral crossed cerebello-cerebral diaschisis and mutism after surgery for cerebellar medulloblastoma

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A 7-year-old boy developed mutism after surgery for cerebellar medulloblastoma. Postoperative magnetic resonance imaging (MRI) showed atrophy of the cerebellar vermis and both cerebellar hemispheres, predominantly on the right side. Single photon emission computed tomography (SPECT) with technetium-99m-ethyl cysteinate dimer (Tc-99m ECD) revealed decreased cerebral blood flow (CBF) in the bilateral thalami, bilateral medial frontal lobes, and left temporal lobe in addition to the cerebellar vermis and both cerebellar hemispheres when mutism was manifest, indicating the existence of bilateral crossed cerebello-cerebral diaschisis (BCCCD). Circulatory disturbance in both cerebellar hemispheres secondary to tumor resection probably caused BCCCD in both cerebellar hemispheres, predominantly in the left, via the dentatothalamicortical pathway (DTCP). With recovery of his mutism, CBF increased in the right thalamus, bilateral medial frontal lobes and left temporal lobe. Thus BCCCD was improved, with only a slight decrease in CBF still persisting in the left thalamus. The mechanism of mutism may have involved damage to the cerebellar vermis (the site of incision at operation), the left dentate nucleus (heavily infiltrated by the tumor) and the right dentate nucleus of the cerebellum (affected by circulatory disturbance secondary to acute postoperative edema). The SPECT findings suggested that mutism was associated with BCCCD-induced cerebral circulatory and metabolic hypofunction in the supplementary motor area mediated via the DTCP.

Key words: crossed cerebello-cerebral diaschisis, cerebellar mutism, medulloblastoma, single photon emission computed tomography, technetium-99m-ethyl cysteinate dimer

INTRODUCTION

Mutism is a state of complete inhibition of speech despite clear consciousness. The causes of mutism may be divided into functional and organic categories. Among the known organic pathologies, Benson et al.1 listed acute lesions involving Broca’s speech area,2 lesions in the supplementary motor area (SMA) of the dominant hemisphere,3-7 lesions involving the reticular formation in the midbrain, bilateral thalamotomy for Parkinson’s disease,8 and pseudobulbar palsy in patients with diffuse bilateral cerebral hemispheric lesions.9 In addition, cerebellar mutism associated with lesions of the posterior cranial fossa has attracted attention. Since Sakai et al.10 described three cases occurring after surgery on posterior fossa tumors in 1980, there have been a number of such case reports.11-19 Cerebellar mutism commonly occurs in children after resection of posterior fossa tumors, with an incidence of 8.2%,11 and 8.5%.12 Most of the tumors involved medulloblastomas, although astrocytomas and ependymomas have also been involved. Cerebellar mutism is presumed to be related to involvement of the superior vermis10 and parasyuperior vermis10 or the inferior vermis13 of the cerebellum, the base of the fourth ventricle (dentate nucleus of the cerebellum and superior cerebellar peduncle),14 the bilateral dentate nuclei, cerebellar impairment caused by operative manipulations,15 and disturbance of the dentatothalamicortical pathway (DTCP) communicating with the SMA.16 In a patient with
Neurological examination showed bilateral choked discs, bilateral horizontal gaze nystagmus and gait ataxia, and computerized tomography (CT) revealed a brain tumor in the posterior fossa (Fig. 1), so the patient was referred to the department of neurosurgery for emergency admission. Neurological examination on admission showed clear consciousness, bilateral choked discs, bilateral horizontal gaze nystagmus, and gait ataxia. His speech was normal. CT and magnetic resonance imaging (MRI) revealed a lesion located in the cerebellar vermis with a cystic component and uniform enhancement. The tumor was adjacent to the pons and medulla oblongata, and had a poorly defined edge that suggested infiltration into the base of the 4th ventricle. The 4th ventricle was compressed, whereas the lateral ventricles, 3rd ventricle and aqueduct were dilated, indicating acute hydrocephalus. From the imaging findings, a cerebellar medulloblastoma was suspected.

On January 6, 1998, emergency surgery was performed. The operation was done in the prone position by a midline suboccipital approach. After the dura mater was incised, the pyramid of the cerebellar vermis in the posterior fossa was seen to be noticeably enlarged. The arachnoid and the vermis were incised to expose the tumor. The margin of the tumor was unclear and the lesion was highly vascular. The dentate nucleus of the cerebellum and the left side of the base of the 4th ventricle were partially invaded, so the tumor and these areas were resected as completely as possible. The histologic diagnosis was medulloblastoma.

Immediately after surgery, the patient had clear consciousness, and responded well to commands by blinking, rotating the head, and shaking hands, but he had no speech and mutism was evident. There were no otorhinolaryngological abnormalities of the vocal cords or larynx. Postoperative edema caused mild left abducens and left facial palsy, but these symptoms improved gradually. CT obtained immediately after surgery showed that the tumor was completely removed and that the hydrocephalus had improved. MRI was done on day 16 postoperatively because of persistent mutism, and revealed enhancement of the cerebellar vermis and the bilateral cerebellar hemispheres, induced by postoperative circulatory disturbance. To clarify the cause of mutism, cerebral perfusion SPECT was performed 5 minutes after intravenous injection of 370 MBq of technetium-99m-ethyl cysteinate dimer (Tc-99m ECD). We used a gamma camera (Starcam 400AC/ T, GE-Yokogawa Medical Co., Tokyo, Japan) equipped with a low-energy all-purpose collimator, and collected data under the following conditions: 128 × 128 matrix, 360°, 64 directions and 35 seconds per direction. During reconstruction of SPECT images, a Butterworth filter and a Ramp filter were used for preprocessing and post-processing, respectively. SPECT (Fig. 2) on postoperative day 23 showed a decrease in cerebral blood flow (CBF) mainly in the bilateral thalami, bilateral medial

**CASE REPORT**

A seven-year-old right-handed boy who had been healthy since birth developed headache, nausea and several episodes of vomiting with a temperature of 37°C in late December 1997. On January 3, 1998, he suffered from worsening headache and dizziness on walking. Two days later, he was brought to Kitasato University Hospital.
therapy from April 27 to June 26, 1998. At present, his speech is normal and he has had no evidence of local recurrence or meningeal dissemination. He remains under observation at the outpatient clinic.

**DISCUSSION**

Circulatory and metabolic hypofunction secondary to the remote effect of a unilateral cerebellar hemispheric lesion on the contralateral cerebral hemisphere can sometimes be observed on radionuclide imaging studies of the brain and is known as crossed cerebello-cerebral diaschisis (CCCD). There have been reports of posterior fossa lesions inducing CCCD in patients with cerebellar infarction, and cerebellar hemorrhage. CBF is reduced by a DTCP-mediated remote effect of posterior fossa lesions on the contralateral cerebellar hemisphere. Output from the cerebral cortex via the corticopontocerebellar pathway passes through the internal capsule to reach the pontine nuclei. From the pontine nuclei, the pathway runs through the middle cerebellar peduncle and then terminates in the contralateral cerebellar cortex. The impulses from this pathway are input into the dentate nucleus, which receives the output from the DTCP, and then ascend crosswise through the midbrain from the superior cerebellar peduncle, to be relayed to the SMA of the cerebral cortex via the contralateral ventrolateral nucleus of the thalamus. This suggests that mutism involves a nerve tract running through the DTCP that communicates with the dentate nucleus of the cerebellum, the contralateral ventrolateral nucleus of the thalamus, and the SMA.

SPECT studies have rarely been done in patients with mutism after surgery for posterior fossa tumors, and only seven patients (including the present one) have been described to date. Among the previous patients, one had normal perfusion on SPECT, one had transiently reduced CBF in the left cerebellar hemisphere, and four had transiently reduced CBF in one cerebellar hemisphere and in the contralateral cerebral hemisphere. The present patient had BCCCD with a transient decrease in CBF in both cerebellar hemispheres and both cerebral hemispheres. In five of the seven patients (including the present one), CCCD appeared to be responsible for the development of cerebellar mutism. The association of CCCD with mutism suggests the role of the SMA, which is the terminus of the DTCP in the production of speech. Many researchers, including Penfield et al., have discussed the relationship between the SMA and speech, as well as the association between lesions in this area and speech disorders. Larsen et al. confirmed an increase in CBF in the bilateral SMAs (predominantly on the left) during conversation, and reported a correlation between speech function and the SMA. In our patient, among the sites showing reduced CBF in the presence of mutism, supratentorial CBF increased predominantly in the left cerebral hemisphere along with the improvement in speech.

**Fig. 3** Tc-99m ECD-SPECT on postoperative day 68 shows cerebral blood flow (CBF) increased in the right thalamus, bilateral medial frontal lobes and left temporal lobe. Bilateral crossed cerebello-cerebral diaschisis is improved, with only a slight decrease of CBF still persisting in the left thalamus.

**Fig. 4** Enhanced MRI shows mild atrophy of both cerebellar hemispheres (mainly on the right) and slight enlargement of the 4th ventricle, but no supratentorial lesions.
The mechanism of mutism in the present patient may have involved damage to the cerebellar vermis, which was the site of the surgical incision; the dentate nucleus of the left cerebellum, which was heavily infiltrated by the tumor; and the dentate nucleus of the right cerebellum, which was affected by circulatory disturbance secondary to postoperative edema. SPECT findings suggested that BCCCD caused by a remote effect via the DTCP led to a decrease in CBF and metabolism that resulted in hypofunction of the SMA, probably playing a role in the development of cerebellar mutism. In the future, it will be necessary to compare the changes in CBF and metabolism in cerebellar mutism by means of positron emission tomography (PET) and SPECT with a quantitative flow-mapping image. Statistical images such as statistical parametric mapping (SPM) and three-dimensional stereotactic surface projections (3D-SSP) are expected to assist in clarifying the mechanism of cerebellar mutism. Assessment of changes in CBF by performing SPECT before and after surgery for brain tumors may be useful in clarifying obscure neurological symptoms that appear after surgery, as in our patient. Accordingly, radionuclide studies are important as well as CT and MRI.

REFERENCES