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Case Report

Mutism resulting from heterochronic bilateral cerebellar hemorrhages – A case report

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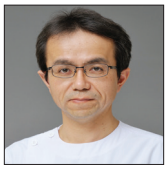

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ABSTRACT

Background: Cerebellar mutism (CM) is a neurological condition characterized by lack of speech due to cerebellar lesions. Interruption of the bilateral dentatothalamocortical (DTC) pathways at midline structure seems the principal cause of CM but not fully understood. We described a rare case of CM due to heterochronic bilateral cerebellar hemorrhages.

Case Description: An 87-year-old woman presented with depression of alertness after sudden vomiting. Neurologically, mild dysmetria and mutism were observed. The head computed tomography (CT) showed both a fresh right cerebellar hemorrhage and an obsolete left one. The patient was diagnosed as CM since both the thalamus and the supplementary motor area were bilaterally intact on both CT and magnetic resonance imaging. Medical treatment and rehabilitation improved her ataxia and ambulation. She became cognitively alert and could communicate by nodding, shaking her head, or facial expression. However, her mutism did not change at 4 months after the stroke.

Conclusion: There are few reports on CM due to direct injuries to the bilateral dentate nuclei. Since our case did not show any injury other than bilateral dentate nuclei, this report can support the hypothesis that the interruptions of the bilateral DTC are the cause of CM.

Keywords: Cerebellar hemorrhage, Cerebellar mutism, Diaschisis, Thalamodentatocortical pathway

INTRODUCTION

Cerebellar mutism (CM) is a neurological condition characterized by lack of speech despite reading and writing with intact comprehension after cerebellar injury,[6,16,18] lasting for a few months on average.[6] CM rarely occurs in children after surgery for posterior fossa midline tumors[2] such as medulloblastoma.[17] CM also occurs following trauma,[9] stroke,[3,5,8,10,11,13,15,16] and infections.[14]

Interruptions of the bilateral dentatothalamocortical pathways (DTC) may be the principal cause of CM.[1,18] CM can result from injuries anywhere along the DTC.[6] However, the anatomical substrate of CM is not fully understood. It has also been under debate whether bilateral, unilateral, or dominant side injury of the DTC is the cause of CM. The previous CM patients generally underwent neurosurgical manipulation to the cerebellum, resulting in difficulty to indicate the responsible region precisely.

We report a CM patient with both a recent right cerebellar hemorrhage and an older left one. The head computed tomography (CT) or magnetic resonance imaging (MRI) showed injuries to the bilateral dentate nuclei. Previous reports are scarce on CM resulting from injuries of the bilateral dentate nuclei.[7,12] This report can contribute to the elucidation of the mechanism of CM and support the hypothesis that bilateral interruption of the DTC is the principal cause of CM.

CASE REPORT

An 87-year-old right-handed female patient presented to our emergency room with depression of alertness after sudden vomiting. She had suffered from the left cerebellar hemorrhage 20 years ago and had been taking antihypertensive medications. Her blood pressure was 235/135 mmHg on admission. She had mild dysmetria and did not speak any words although could obey commands, with the Glasgow coma scale score of 11 (E4V1M6). The head CT showed a high-density area in the right cerebellar hemisphere, presenting 9 mL of a right cerebellar hematoma. It also showed a low-density area in the left cerebellar hemisphere, presenting an older left cerebellar hemorrhage. The head MRI showed perifocal edema surrounding the right cerebellar hematoma. Both the thalamus and the supplementary motor area were bilaterally intact on both CT and MRI [Figure 1].

On the following day, her neurological findings did not change and rehabilitation began. The CT did not show hematoma expansion. On the 3rd day, she was mute but cognitively alert, although she could communicate by nodding, shaking her head, or facial expression enabled communication.

She could not walk independently and was transferred to a rehabilitation hospital 31 days after the onset of the right cerebellar hemorrhage. Her mutism improved a bit and a few short words (e.g. yes and no) were uttered. Four months after the onset, she could walk with a little care. However, her mutism did not change compared to when she was transferred to the rehabilitation hospital. The CT and MRI did not show any additional findings rather than the injuries of the bilateral dentate nuclei. She was diagnosed as CM caused by heterochronic bilateral cerebellar hemorrhages.

DISCUSSION

In general, mutism is defined as the inability to speak despite cognitively alert patient. It can occur due to lesions in several parts of the brain, such as Broca's area, the supplementary motor area, thalamus, mesencephalic reticular formation regions, and bilateral hemispheric lesions.[16]

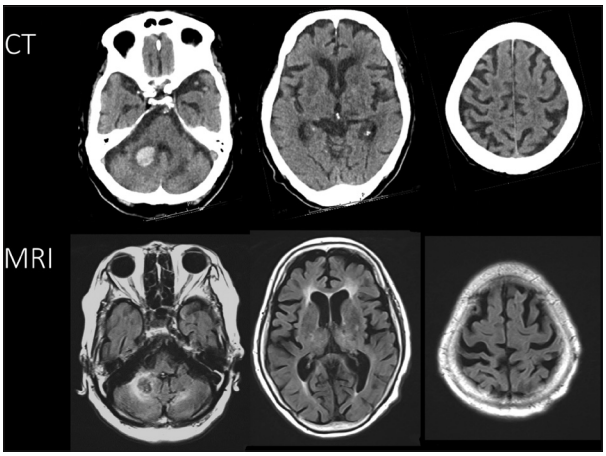

Figure 1: Computed tomography showed a high-density area in the right cerebellar hemisphere and a low-density area in the left cerebellar hemisphere (upper row). The head magnetic resonance imaging showed edema around the right cerebellar hematoma (lower row). Both the thalamus and the supplementary motor area were bilaterally intact on both the computed tomography and magnetic resonance imaging images.

CM is one of the types of transient mutism resulting from interruptions of the bilateral DTC pathways, in general. Posterior fossa tumor surgery is the most common cause of this syndrome, as symptoms usually last for a few months. Surgical excision of the posterior fossa tumor causes CM, occurring in 27.7%[2] and its duration is limited from a few weeks until 120 days.[7] In the present case, surgical treatment was not performed, and the CM lasted for more than 4 months. It seemed different from the typical CM in its clinical characteristics. Thus, the mechanism of CM is discussed below focusing on the bilateral DTC including dentate nuclei.

The anatomical substrate of CM is not fully understood. Vermis, brainstem, and anywhere along the DTC are reported as a candidate for responsible regions of CM after posterior fossa surgery.[18] The vermis has neural connections with the deep cerebellar nucleus and is involved in the coordination of laryngeal and respiratory function. The inferior vermis seems to be associated with speech initiation,[18] suggesting that splitting of the vermis was related to CM.[4] However, either the cerebellar hemisphere or the brain stem could be manipulated to some extent intraoperatively. Therefore, it seems difficult to attribute CM only to the vermis.

CM seems to be also related to brainstem involvement of the tumor.[18] The DTC connects the dentate nucleus to the contralateral cerebral cortex. The nerve passes the decussation of the superior cerebellar peduncle, the red nucleus in the midbrain, and the ventrolateral anterior nucleus of the thalamus[19] [Figure 2a]. Therefore, direct damage or postoperative edema in the brainstem would interrupt the DTC bilaterally, causing CM [Figure 2b].

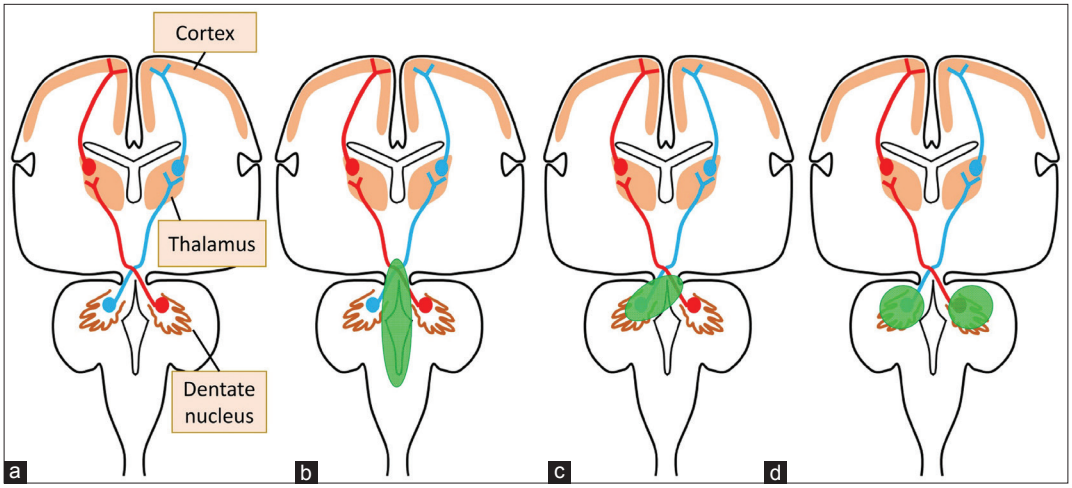

Figure 2: Schematic illustration of the dentatothalamocortical pathway. (a) Surgical excision of posterior fossa tumor would damage the brainstem directly or cause postoperative edema in the brainstem. (b) Massive cerebellar hemorrhage or surgical intervention would injure both the unilateral dentate nucleus and the brainstem or the contralateral superior cerebellar peduncle. (c) Our case presented lesions to the bilateral dentate nuclei which interrupt the dentatothalamocorticals bilaterally. (d) All the lesions interrupt the dentatothalamocortical bilaterally, and cerebellar mutism would be caused.

Table 1: Case series of cerebellar mutism due to cerebellar hemorrhage.[3,5,8,10,13,15]

Author (Year)	Age/Sex	Pathology	Site	Medial or contralateral invasion	Treatment
Coplin (1997)	47/Male	Unknown	Supravermis	+	Hematoma removal
Kawai (2001)	64/Male	HT	R	+	Hematoma removal
Idiaquez (2011)	20/Male	AVM	R	+	Hematoma removal
De Smet (2012)	60/Male	HT	L	+	Hematoma removal
Mariën (2013)	71/Male	HT	R	+	Drainage
Lee (2017)	20/Female	AVM	L	+	Hematoma removal
Our case (2018)	87/Female	HT	R, older L	-	Medical treatment

AVM: Arteriovenous malformation, HT: Hypertension, L: Left, R: Right

There have been six previous case reports of CM after cerebellar hemorrhage [Table 1].[3,5,8,10,13,15] Although the sites of cerebellar hemorrhage were unilateral in five of six reports, all of the hematomas were so massive that they invaded beyond the midline structures, requiring surgical interventions in most cases. Either the massive hematoma and/or the surgical intervention would injure both the unilateral dentate nuclei and the brainstem, including the contralateral superior cerebellar peduncle. Therefore, the bilateral DTC would have been injured even in cases with unilateral hemorrhage [Figure 2c].

In the present case, injuries were limited in the bilateral dentate nuclei due to the recent right cerebellar hemorrhage and the older left cerebellar hemorrhage without surgical treatment. Besides, the CT and MRI did not show any other lesions along the DTC. In previous reports on CM, due to surgical intervention or massive hemorrhage, it has been difficult to indicate the responsible region for CM precisely. The present case seems simple and can support the theory that bilateral interruptions of the DTC are the cause of CM [Figure 2d].

Some cases of CM resulting from injuries of bilateral dentate nuclei have been reported. Kubota *et al.* reported CM due to rotavirus-associated acute cerebellitis.[12] The MRI showed the signal changes in the bilateral dentate nuclei. Guidetti and Fraioli performed stereotactic manipulation to the bilateral dentate nuclei as a treatment for dyskinesia. They observed mutism in two of their 88 patients 1–3 months after the surgery.[7] This is the only report of CM due to direct injuries to the bilateral dentate nuclei. Therefore, our report is the first report on CM due to lesions of the bilateral dentate nuclei without surgical manipulation.

CONCLUSION

We reported the patient with CM due to the recent right cerebellar hemorrhage and the older left one. Both CT and MRI did not show any lesions other than the bilateral dentate

nuclei. This report can support the hypothesis that bilateral interruptions of the DTC are the cause of CM.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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