

Transient Mutism Resolving into Cerebellar Speech after Brain Stem Infarction Following a Traumatic Injury of the Vertebral Artery in a Child

Y. Miyakita¹, Y. Taguchi¹, Y. Sakakibara¹, M. Matsuzawa¹, and H. Kitagawa*

¹ Divisions of Neurosurgery, St. Marianna University Yokohama City Seibu Hospital, Yokohama, Japan

² Paediatric Surgery, St. Marianna University Yokohama City Seibu Hospital, Yokohama, Japan

Summary

A 3.7-year-old girl presented with an anterior neck injury followed by progressive subcutaneous emphysema and loss of consciousness. After resuscitation, a laceration on the first tracheal cartilage was closed surgically. As she was extubated one week later, she was found to have right hemiplegia and muteness. MRI showed a T2-bright lesion on the tegmentum of the left midbrain down to the upper pons. Right vertebral angiography disclosed an intimal flap with stenosis at the C3 vertebral level presumably caused by a fracture of the right C3 transverse process later confirmed in a cervical 3D-CT scan. Her muteness lasted for 10 days, after which she began to utter some comprehensible words in a dysarthric fashion. Her neurological deficits showed improvement within 3 months of her admission. Transient mutism after brain stem infarction has not been reported previously. We discuss the anatomical bases for this unusual reversible disorder in the light of previous observations and conclude that bilateral damage to the dentatohalamocortical fibers at the decussation of the superior cerebellar peduncle may have been responsible for her transient mutism.

Keywords: Mutism; child; trauma; midbrain; infarction.

Introduction

Mutism is defined in Stedman's Medical Dictionary as "the state of being silent" and "organic or functional absence of the faculty of speech." The aetiologies of mutism include trauma, epilepsy, tumours, stroke, psychoses, and surgery [1, 2, 10, 14, 15, 19]. Transient mutism is most commonly seen following extensive callosotomy [14] and infrequently after posterior fossa surgery, particularly in paediatric patients [1, 2, 5, 19]. The anatomic basis for this postoperative functional change is unclear but may reside in the supplementary motor area [10, 15], the medial deep cerebellar nuclei [3], and their fiber connections [1, 2, 5, 19]. We report a case of a 3.7-year-old girl who developed transient mutism resolving into cerebellar speech

after a brain stem infarction following a traumatic injury of the vertebral artery. To our knowledge, this is the first reported case showing "cerebellar mutism" after brain stem infarction. We will discuss the anatomical basis responsible for this speech disturbance.

Case Report

A 3.7-year-old girl presented to an outside hospital with an anterior neck injury. The patient suffered a laceration on the front of the neck when she fell forward. No details were known, but her friends witnessed a classmate making fun of her and jumping upon her back. The patient complained of no particular abnormalities except for neck pain. While waiting at the registration area of that hospital, she began to experience progressive swelling on her face, neck, and upper trunk, and lost consciousness. The patient was then transferred to our Emergency and Critical Care Center for further evaluation and treatment. The medical history of the patient, including speech development, was unremarkable.

Immediately after her arrival, the patient was intubated because of respiratory failure which appeared to be caused by marked subcutaneous emphysema following a tracheal injury. No laterality in the spontaneous movement of her extremities was revealed, though neurological findings were not fully obtained. A chest X-ray showed bilateral pneumothorax and subcutaneous emphysema. In the operating room, chest tubes were inserted bilaterally to release the bilateral tension pneumothorax. A lacerated wound measuring 1 cm in horizontal length just below the level of the cricoid cartilage was extended to explore the cause of this serious condition. This procedure allowed us to find the first tracheal cartilage to be lacerated vertically approximately 5 mm in length and to suture it simply. The patient was mechanically ventilated under deep sedation for the following week. After endoscopic confirmation of the healing of the tracheal wound, sedatives were discontinued and the patient was extubated on the 8th day in hospital.

The next morning, the patient was still drowsy but was noticed to have a right hemiplegia, upward gaze palsy, and no use of speech despite being able to obey simple verbal commands. A computerized tomography (CT) scan showed an equivocal low density in the brain stem. Magnetic resonance (MR) imaging performed on the 10th hospital day revealed an irregularly shaped high intensity area in the

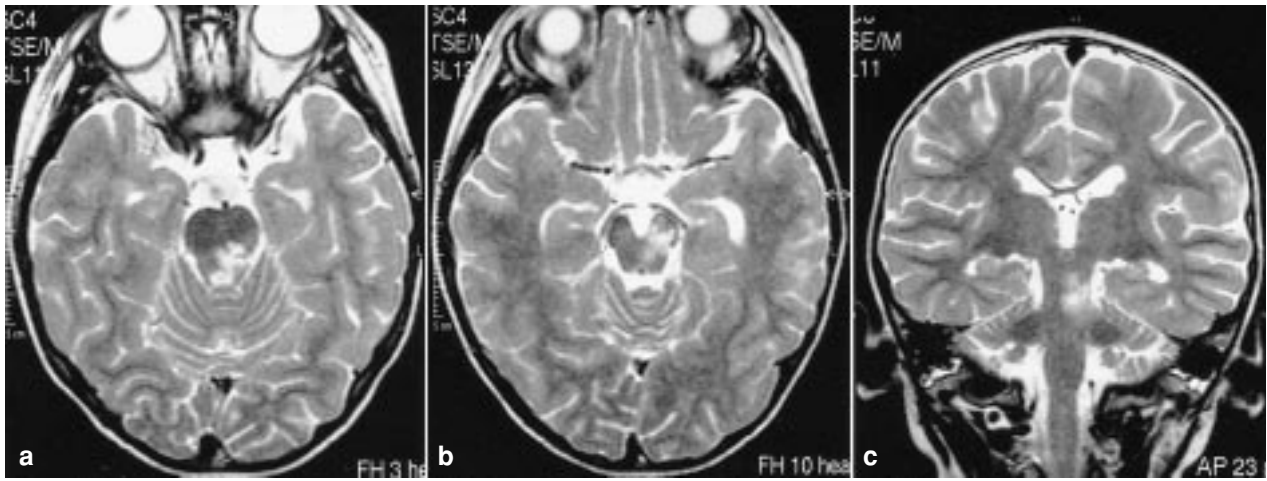


Fig. 1. Axial (a, b) and coronal (c) T2-weighted magnetic resonance images showing an irregularly shaped high intensity area in the tegmentum of the lower left midbrain down to the upper pons, including the left cerebral peduncle and extending contralaterally



Fig. 2. Selective right vertebral angiogram disclosing a focal stenosis with indentation of the arterial wall at the upper limit of the C3 vertebral level

tegmentum of the lower left midbrain down to the upper pons including the left cerebral peduncle and extending contralaterally (Fig. 1). No abnormalities were found in the cerebrum or the cerebellum. MR angiography showed a segmental signal void on the upper cervical portion of the right vertebral artery. Selective right vertebral angiography disclosed an intimal flap with stenosis at the C3 vertebral level (Fig. 2). A cervical three-dimensional (3D) CT reconstruction carried out later disclosed a fracture in the anterior aspect of the right C3 transverse process bending posteriorly to compromise the vertebral foramen (Fig. 3). This could not be detected in the initial cervical plain X-ray films. The patient was diagnosed to have a

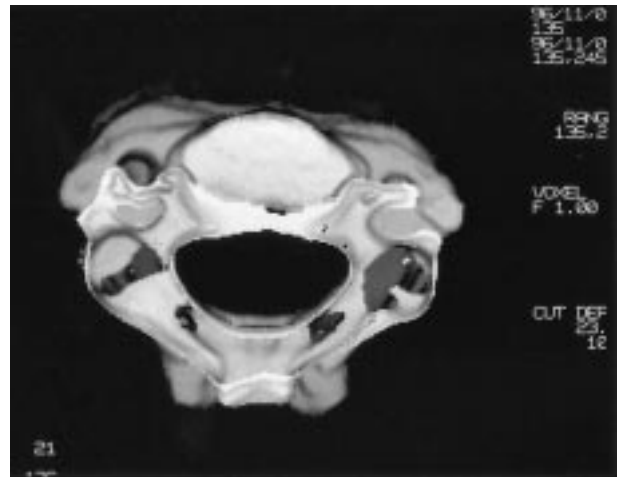


Fig. 3. A cervical 3-dimensional computed tomography reconstruction showing a fracture in the anterior aspect of the right C3 transverse process bending posteriorly to narrow the vertebral foramen

brain stem infarction following a traumatic injury of the right vertebral artery, and treated conservatively with oral use of antiplatelets and the application of a cervical collar.

The patient was mute from the 8th to the 18th hospital day. During this period, she became able to take her meals orally without difficulty in swallowing and to communicate by signs or by pointing at a communication board. By the 19th hospital day, her upward gaze palsy disappeared. The patient was walking with motor weakness on her right side and gait ataxia. She began whispering a few words. Over the next several days she could express comprehensible speech in a dysarthric fashion consisting of monotonous and somewhat explosive speech. Her gait disturbance improved markedly, as did her speech. She was discharged home on the 33rd hospital day. Within 2 months of her discharge, her speech returned near to her previous baseline. She had only minimal right hand weakness without any problems in her daily life.

Discussion

Occlusive cerebrovascular disease in children is uncommon and that in the posterior cerebral circulation is quite rare [4]. Trauma is the commonest most likely predisposing factor in young patients with ischaemic stroke [7]. In our patient, a traumatic injury occurred to the right vertebral artery at the C3 vertebral level associated with a C3 transverse process fracture, which was thought to be the cause of the brain stem infarction. The anterior aspect of the right transverse process was disclosed in a 3D-CT scan to be fractured and bent dorsally. At the time of injury, the vertebral artery may have been jammed causing the intimal damage, which was most likely a small dissection. The stroke resulted after thrombi formed at the site of dissection and subsequently embolized after a period of time [7]. This type of fracture is not yet known but may be likely to occur in children, because in younger patients, significant hyperextension may be accompanied by unusual spinal injuries such as small anterior body chip avulsion fractures or disruption of the anterior longitudinal ligament [13]. The rarity and mechanism of this type of transverse process fracture as well as its relationship with the anterior neck injuries call for further discussion, but are beyond the scope of this report. The focus is the patient's transient mutism after brain stem infarction resolving into cerebellar speech.

Mutism is defined as the state in which a patient is conscious but unwilling to speak [6]. The aetiology of mutism is variable, but mutism is an infrequent and usually transient complication of brain surgery. Mutism following removal of posterior fossa tumours in children has been described. This condition has recently been called cerebellar mutism [2, 19]. Ersahin *et al.* [2] reviewed a total of 46 cases of cerebellar mutism including those of their seven patients and analysed the characteristics. Most of the patients were children. All mass lesions were considered to be large or very large. The latency for the development of mutism ranged from 0 to 6 days and the mutism itself lasted from 4 days to 4 months. Dysarthric speech ensued after resolution of the mutism in 35 of 46 patients. Since the cerebellar vermis was the tumour site in the majority of cases, surgical removal could potentially lead to damage of midline cerebellar tissue including the cerebellar nuclei bilaterally. The delay in the development mutism may be explained by oedema or disturbed perfusion in the midline cerebellar structures [2, 19]. This may explain in part the transient nature of

cerebellar mutism. The anatomical basis for this syndrome remains conjectural. Considering the occurrence of complete loss of speech after bilateral stereotactic lesioning of the dentate nucleus in patients with dyskinesia [3], bilateral damage of the dentate nucleus may be a strong candidate for causing cerebellar mutism [3]. The dysarthria that marks the process of speech recovery suggests that the cerebellar damage is undoubtedly the most important of the possible organic factors. As for the speech function of the human cerebellum, Lechtenberg and Gilman [8] analysed 162 dysarthric patients with nondegenerative cerebellar disease and concluded that involvement of the left cerebellar hemisphere was most likely to correlate with a speech dysfunction and, within the left cerebellar hemisphere, a correlation was found between dysarthria and superior hemispheric injury. As the dentate nucleus represents input widely from the superior cerebellar hemispheres, it is reasonable that the cerebellar mutism seen in patients with posterior fossa surgery is accompanied with cerebellar dysarthria in the process of speech recovery.

Postoperative mutism is rather commonly seen among patients who have undergone corpus callosotomy for medically intractable seizures. Ross, *et al.* [14] noted mutism in 35 out of 40 patients who underwent corpus callosotomy, then reported that retraction in the region of the supplementary motor area may have played a role in the occurrence of mutism. In a series by Rostomily *et al.* [15], five of six patients with a primary glial tumour of the supplementary motor area became mute within 24 hours of resection without disturbance of sensorium or comprehension. These reports suggested that the supplementary motor area is responsible for the production of mutism. The patterns of speech recovery from mutism seen in the patients with lesions of the supplementary motor area are similar to those of patients with cerebellar mutism, though they differ in regard to the dysarthria and the time frame.

To explain concurrently, several investigators have postulated the dentatohalamocortical pathway as the anatomical substrate responsible for the development of mutism [1, 2, 5, 19]. Cruchfield *et al.* [1] reported that bilateral interruption of the dentatohalamocortical pathway appears to be a plausible cause of postoperative mutism. Frim and Ogilvy [5] reported on an 8-year-old girl who developed mutism for 12 days after resection of a pontine cavernous malformation. The mutism of their patient resolved into cerebellar dys-

arthria similar to many patients who had cerebellar mutism after posterior fossa surgery. They hypothesized that the operative procedure injured speech-related fibers somewhere along the tracts of the cerebellar corticodentate connections or the dentatohalamocortical tract. However, with superficial lesions in the pons, the cerebellar efferent fibers from the dentate nucleus are unlikely to be injured [11].

Stroke may cause a state of mutism. In most patients exhibiting mutism after occlusive cerebrovascular disease, the lesion has involved the supplementary motor area [10]. Although vascular lesions involving the diencephalon or the mesencephalon may produce akinetic mutism [17], this condition is apparently different from the transient mutism discussed above. To our knowledge, no patient showing transient mutism after brain stem infarction has been reported previously.

In our patient, T2-weighted MR imaging showed an irregular high intensity area in the tegmentum of the lower left midbrain down to the upper pons extending contralaterally with no abnormalities anywhere in the cerebrum or the cerebellum. This lesion was diagnosed as a brain stem infarction following the traumatic injury of the right vertebral artery, which included the dentatorubrothalamic tract bilaterally between the decussation of the superior cerebellar peduncle and the red nuclei. Based on our previous clinical observations, we concluded that bilateral damage to the dentatorubrothalamic tract may have been responsible on an anatomical basis for the patient's transient mutism resolving with cerebellar dysarthria.

Although it appears plausible that bilateral interruption of the dentatohalamocortical pathways may be responsible for the development of postoperative mutism, the reason why mutism occurs remains uncertain. Recent advances in functional imaging techniques have given clues to clarify the mechanism of mutism development. Traditionally, the human cerebellum has been recognized as a motor mechanism, but the images obtained by positron emission tomography scan have provided us new facts suggesting that the cerebellum is involved in higher functions, such as cognition and language [12, 16]. The cerebellum is activated when humans perform certain cognitive and language tasks even in the complete absence of any motor activity [16]. Rather than subserving the motor function of word-articulation, the frontal association area including Broca's language area may be involved in processes of word-finding, which are regarded as cognitive processes [18]. Thus, there may be an inti-

mate connection between the cerebellum and the frontal association area. Leiner *et al.* [9] reported that the phylogenetically new cerebellum consisting of the lateral cerebellum, the macrogyric part of the dentate nucleus and their connections may play a major role in carrying out cognitive and language tasks, and that signals from the newly-evolved structures of the cerebellum can reach the association area of the cerebral cortex via ascending connections. Lesions in the structures described above may impair word-finding processes and consequently lead to the development of mutism. This may also be true in the anatomical substrate conversely connecting between the association area and the lateral cerebellum (descending connections). Presumably because of immaturity, children may be susceptible to having trouble with cognitive and language functions. Functional neuro-imaging studies on patients with mutism may offer more valuable information in order to clarify the mechanism of the speechless state.

References

1. Crutchfield JS, Sawaya R, Meyers CA, Moore III BD (1994) Postoperative mutism in neurosurgery. Report of two cases. *J Neurosurg* 81: 115–121
2. Ersahin Y, Mutluer S, Cagli S, Duman Y (1996) Cerebellar mutism: Report of seven cases and review of the literature. *Neurosurgery* 38: 60–66
3. Fraioli B, Guidetti B (1975) Effects of stereotactic lesions of the dentate nucleus of the cerebellum in man. *Appl Neurophysiol* 38: 81–90
4. Fraser RAR, Zimble SM (1975) Hindbrain stroke in children caused by extracranial vertebral artery trauma. *Stroke* 6: 153–159
5. Frim DM, Ogilvy CS (1995) Mutism and cerebellar dysarthria after brain surgery: case report. *Neurosurgery* 36: 854–857
6. Geschwind N (1971) Aphasia. *N Engl J Med* 284: 654–656
7. Hilton-Jones D, Warlow CP (1985) Non-penetrating arterial trauma and cerebral infarction in the young. *Lancet* 2: 1435–1438
8. Lechtenberg R, Gilman S (1978) Speech disorders in cerebellar disease. *Ann Neurol* 3: 285–290
9. Leiner HC, Leiner AL, Dow RS (1993) Cognitive and language functions of the human cerebellum. *TINS* 16: 444–447
10. Masdeu JC, Schoene WC, Funkenstein H (1978) Aphasia following infarction of the left supplementary motor area: a clinicopathologic study. *Neurology* 28: 1220–1223
11. Mickle JP. Comment on reference 5
12. Petersen SE, Fiez JA (1993) The processing of single words studied with positron emission tomography. *Annu Rev Neurosci* 16: 509–530
13. Rea GL (1996) Subaxial injuries of the cervical spine. In: Menzies AH, Sonntag VKH (eds) *Principles of spinal surgery*. McGraw-Hill, New York, pp 885–898
14. Ross MK, Reeves AG, Rovers DW (1984) Postcommissurotomy mutism. *Ann Neurol* 16, p 114 (abstract)

15. Rostomily RC, Berger MS, Ojemann GA, Lettich E (1991) Postoperative deficits and functional recovery following removal of tumors involving the dominant hemisphere supplementary motor area. *J Neurosurg* 75: 62–68
16. Ryding E, Decety J, Sjöholm H, Stenberg G, Ingvar DH (1993) Motor imagery activates the cerebellum regionally. A SPECT rCBF study with 99mTc-HMPAO. *Cogn Brain Res* 1: 94–99
17. Segarra JM (1970) Cerebral vascular disease and behavior. I. The syndrome of the mesencephalic artery (basilar artery bifurcation). *Arch Neurol* 22: 408–418
18. Tonkonogy J, Goodglass H (1981) Language function, foot of the third frontal gyrus, and rolandic operculum. *Arch Neurol* 38: 486–490
19. Van Dongen HR, Catsman-Berrevoets CE, Van Mourik M (1994) The syndrome of ‘cerebellar’ mutism and subsequent dysarthria. *Neurology* 44: 2040–2046

Comments

The authors describe a case of transient mutism after brain stem infarction in a child. The case report is distinct with good figures. The

discussion of mutism is relevant and gives a good and accurate overview of the question of mutism in general and cerebellar mutism in particular.

L. Rabow

This case is well investigated and documented, especially the subtle diagnostics using good MRI brain stem imaging, DSA and 3-D-CT reconstruction of C3. The pathophysiology of this patient was clarified in detail allowing discussion of the anatomical reasons for her mutism. This case is therefore a good illustration of today’s standards in the evaluation of cervical injury.

Based on the MRI imaging of the brain stem damage, this paper gives a complete overview of the actual literature on mutism, especially after trauma/infarction. The discussion of this topic is factual and comprehensive. As such consequences after cervical injury are not so rare, but often not well diagnosed, Reading of this case is recommended.

G. M. Gaab

Correspondence: Yoshio Taguchi, M.D., Division of Neurosurgery, St. Marianna University Yokohama City Seibu Hospital, 1197-1 Yasashi-cho Asahi-ku, Yokohama, Kanagawa, Japan 241-0811.