Child's Nerv Syst (1995) 11: 306–310

CASE REPORT

Paulo Henrique Aguiar
José Pindaro P. Plese
Orildo Ciquini
Raul Marino

Transient mutism following a posterior fossa approach to cerebellar tumors in children: a critical review of the literature

Received: 22 February 1993

P.H. Aguiar (✉)[1] · J.P.P. Plese
O. Ciquini · R. Marino
Division of Neurosurgery,
Department of Neurology,
Hospital das Clínicas,
University of São Paulo,
São Paulo, Brazil

Present address:
[1] Rua Raul Pompéia 1050, Apto 111,
Perdizes, São Paulo CEP 05025-010, Brazil
Fax: 0055 11 289 0661

Abstract Transient mutism has been known as a rare complication following a posterior fossa approach to cerebellar tumors and its cause has not been clearly elucidated. The cerebellar mutism is not accompanied by cranial nerve deficits and disorders of consciousness. Since 1985 only 23 cases of mutism following removal of a cerebellar tumor in children have been reported in the literature. Two additional cases have been operated upon in our department. Extensive injury to the vermian and paravermian cerebellar area, involving the hemispheric cortex, cerebellar peduncles, fibers from the dentato-thalamocortical pathway, and dentate and interpositum nuclei may be the most important anatomical substrate of mutism. The mechanism of such transient mutism seems to be a complex of two or more factors (vascular disturbances due to manipulation or retraction of the cerebellar region around the IV ventricle and emotional factors). On the basis of these 25 cases the major features of the cerebellar mutism are discussed.

Key words Cerebellar neoplasm · Posterior fossa tumor · Mutism · Aphasia

Introduction

Mutism is generally defined as complete absence of speech in a conscious patient with intact comprehension and no evidence of oral apraxia [1, 5]. Mutism can be subdivided in motor aphasia, akinetic mutism, mutism following thalamotomy, pseudobulbar palsy due to diffuse, bilateral hemispheric lesion, phonatory system lesion [4, 6, 17], and transient mutism after transcallosal approach to the ventricles, which has recently been reported [15].

Only in 1985, Rekate et al. [17] were the first to describe cerebellar mutism. Cerebellar mutism may be defined as a transient mutism occurring after a posterior fossa tumor approach in children with unimpaired consciousness, unimpaired symbolic functions, no detectable deficit of cranial nerves or peripheral organs of speech, and no lesions of long pathways in the course of the cranial nerves at the level of the brain stem [3, 17].

Twenty-three reported cases of transient mutism in children since 1985 were found in the literature [3, 5, 6, 10, 14, 15, 17, 21]. Salvati et al. [18] reported one case of transitory mutism after removal of cerebellar medulloblastoma in an adult. A further two children with posterior fossa tumors treated in our department developed cerebellar mutism. In the present study, these 25 cases of cerebellar mutism in children were reviewed with regard to the age and sex of the patients, location of the tumor, the period of latency, the duration of the mutism, and the size and histological type of the tumor.

Personal cases

Case 1

A 9-year-old boy was admitted to our department with a 1-month history of headache, vomiting, truncal ataxia, and diplopia. Neurological examination disclosed bilateral papilledema and a cerebellar

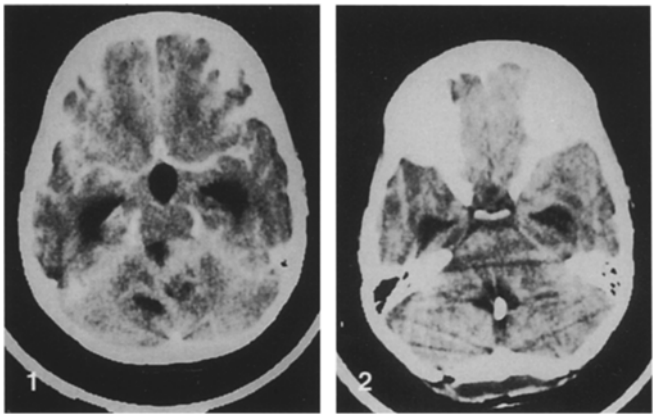


Fig. 1 Axial brain computed tomography (CT) demonstrating a large midline vermian tumor filling the IV ventricle and triventricular hydrocephalus (case 1 or case 24 in Table 1)

Fig. 2 Axial brain CT demonstrating radical tumor removal, improvement of ventricular size, and bilateral hypodense areas at approximate level of the dentate nuclei with a catheter placed in the IV ventricle (case 1 or case 24 in Table 1)

syndrome. Brain computed tomography (CT) demonstrated a large midline vermian tumor with patchy and ill-defined limits filling the IV ventricle and marked triventricular hydrocephalus (Fig. 1).

A posterior fossa approach (suboccipital craniotomy) was performed with the patient in the prone position. A large medulloblastoma with IV ventricle adhesion was totally removed using a Cavitron Ultrasonic Surgical Aspirator (CUSA) and microsurgical technique. A IV ventriculostomy was performed with a catheter connecting the III ventricle to the cisterna magna.

The patient awoke in the recovery room without cranial nerve palsy and was neurologically intact. On the 2nd postoperative day, he abruptly developed dysarthria progressing to muteness, and marked emotional lability. He was able to follow commands, but when asked to speak he cried. Control brain CT demonstrated complete tumor removal, improvement of ventricular size, and bilateral hypodense areas at the approximate level of the dentate nuclei with a catheter placed in the IV ventricle (Fig. 2). On the 5th postoperative day he underwent placement of a definitive ventriculoperitoneal shunt because of progression of hydrocephalus. He gradually improved over the next 5 weeks, and 2 months after the operation he was neurologically intact. The histopathological diagnosis was medulloblastoma. The patient underwent radiation therapy.

Case 2

A 5-year-old boy with a few months' history of unsteady gait, headache, and vomiting was admitted to our department in coma, after progressive loss of consciousness in the previous 24 h. Brain CT revealed a hyperdense vermian tumor partially occupying the IV ventricle with advanced hydrocephalus (Fig. 3). Magnetic resonance imaging (MRI) studies demonstrated a midline vermian mass of mixed signal intensity of T1- and T2-weighted images. After intravenous administration of gadolinium-DTPA, the lesion increased the signal on T1, although it was heterogenous and suggestive of intratumoral bleeding (Fig. 4). A ventriculoperitoneal shunt was placed and the child awoke a few hours after surgery. A right sixth cranial nerve palsy was detected and neurological function was intact.

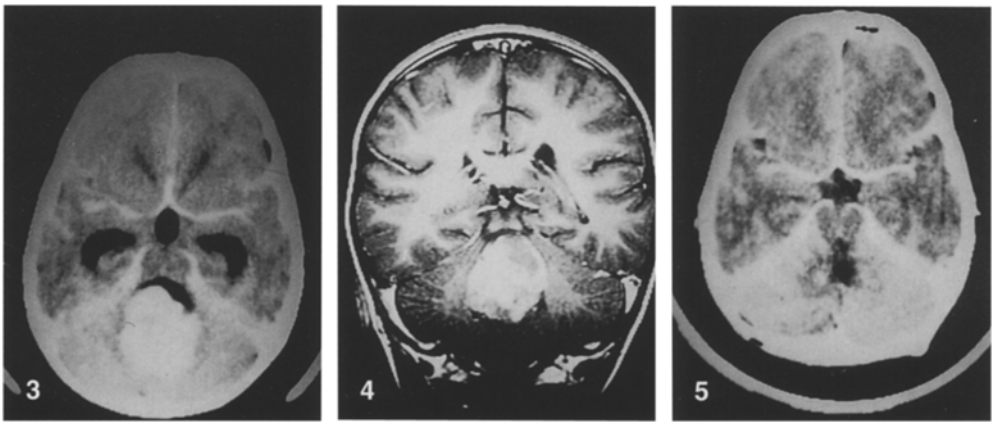
After 1 week the tumor was approached through a midline suboccipital craniotomy (with the patient in the prone position) using a microsurgical technique and the tumor was completely removed with help of a CUSA. The tumor was attached to the lateral recess of the IV ventricle. The histopathological diagnosis was medulloblastoma. Immediately after surgery the patient was awake and fully oriented. There was no additional neurological deficit.

On the 2nd postoperative day, the patient was mute. He had normal cranial nerve function with the exception of the previous right sixth nerve palsy and truncal ataxia. CT scanning demonstrated decreased attenuation in the area around the surgical site (Fig. 5). The patient was extremely irritable, but he could understand commands. He continued to make a gradual recovery and 6 weeks after the operation was markedly improved, with only mild dysarthria and minimal ataxia.

Fig. 3 Axial brain CT demonstrating a hyperdense vermian tumor partially occupying the fourth ventricle with advanced hydrocephalus (case 2 or case 25 in Table 1)

Fig. 4 Coronal T1-weighted magnetic resonance image demonstrating a midline vermian mass of mixed signal after intravenously administration of gadolinium-DTPA (case 2 or case 25 in Table 1)

Fig. 5 Axial brain CT demonstrating reduced attenuation in the area of the surgical site (case 2 or case 25 in Table 1)



Literature review

Clinical data

Twenty-three cases of mutism after posterior fossa surgery in children have been previously reported in the literature. The patient age and sex, the location of tumor, the period of latency, the duration of mutism, and the size and histological type of these tumors are summarized in Table 1. The average age was 7.6 years (range 2–15 years). There was a significant sex difference; 75% male to 25% female in the 16 cases in which sex was specified. In 13 of 21 cases vermian lesions occupied the IV ventricle. Posterior fossa tumors were large in all 18 cases. The most common histological findings were medulloblastoma ($n=14$), astrocytomas ($n=6$), ependymomas ($n=4$), and vascular malformation with hemorrhage ($n=1$). The average time to the appearance of mutism after the posterior fossa surgery (latency period) was 42.5 h (range 0–72 h). The average duration of mutism was 8 weeks (range 2–19.5 weeks).

One case of mutism in an adult was added to this world literature series. This patient was female, 20 years old, with large vermian medulloblastoma occupying the IV ventricle (case 26).

Operative data

The operative position of the patient was described in seven cases. Two of these patients were operated on in the semi-setting position (patients 1 and 12), two in the park bench position (patients 21 and 22), and three in prone position (patients 11, 24, and 25). Only patients 24 and 25 underwent a suboccipital craniotomy; another nine underwent a suboccipital craniectomy. The use of CUSA was reported in six cases (patients 19, 20, 23, 24, 25, and 26).

In 7 of 8 cases the tumor had IV ventricle adhesion. "Radical" removal was performed in all cases out of 11 analyzed. The tumor was removed except for a small attachment to the floor of the IV ventricle in three cases.

Table 1 Summary of 26 cases of mutism after posterior fossa approach to cerebellar tumors (NA, data not available)

Case	Reference	Age (years)	Sex	Tumor location	Tumor size	Histology	Latency (h)	Duration (weeks)
1	Rekate et al. [17]	8	F	Vermis	NA	Medulloblastoma	48	6
2	Rekate et al. [17]	6	M	Vermis, hemisphere	Large	Astrocytoma	72	4
3	Rekate et al. [17]	2	NA	NA	NA	Ependymoma	NA	8
4	Rekate et al. [17]	10	NA	NA	NA	Medulloblastoma	NA	8
5	Rekate et al. [17]	9	NA	NA	NA	Medulloblastoma	NA	12
6	Rekate et al. [17]	11	NA	NA	NA	Medulloblastoma	NA	3
7	Yonemasu [23]	NA	NA	Bilateral	Large	Ependymoma	18–72	4–12
8	Yonemasu [23]	NA	NA	Bilateral	Large	Ependymoma	18–72	3
9	Yonemasu [23]	NA	NA	Bilateral	Large	Medulloblastoma	18–72	4–12
10	Yonemasu [23]	NA	NA	Bilateral	Large	Medulloblastoma	18–72	4–12
11	Volcan et al. [21]	8	F	IV Ventricle	Large	Medulloblastoma	0	2
12	Ammirati et al. [3]	14	M	Vermis, IV ventricle	NA	Astrocytoma	48	6
13	Humphreys [10]	7	M	Vermis, hemisphere	Very large	Medulloblastoma	24	16
14	Humphreys [10]	3	M	IV Ventricle	Large	Medulloblastoma	NA	7
15	Humphreys [10]	7	M	IV Ventricle	Large	Medulloblastoma	NA	10
16	Humphreys [10]	4.5	M	Vermis, IV ventricle	Large	Astrocytoma	72	7
17	Humphreys [10]	10	M	IV Ventricle	NA	Ependymoma	24	10
18	Ferrante et al. [6]	9	M	Vermis, IV ventricle, medial cerebellar hemisphere	Large	Astrocytoma	48	4
19	Ferrante et al. [6]	5.5	F	Vermis, IV ventricle, medial cerebellar hemisphere	Large	Astrocytoma	48	8
20	Ferrante et al. [6]	6	M	Vermis, IV ventricle, hemisphere	Large	Astrocytoma	36	8
21	Dietze and Mickle [5]	7	NA	Vermis, bilateral IV ventricle	Large	Medulloblastoma	NA	12
22	Dietze and Mickle [5]	15	NA	Vermis, medial cerebellar hemisphere	NA	Arteriovenous Malformation	NA	12
23	Nagatani et al. [14]	4	F	Vermis, IV ventricle	Large	Medulloblastoma	24	19.5
24	Present study	9	M	Vermis, IV ventricle	Large	Medulloblastoma	48	5
25	Present study	5	M	Vermis, IV ventricle	Large	Medulloblastoma	48	6
26	Salvati et al. [18]	20	F	Vermis, IV ventricle	NA	Medulloblastoma	46	4

[a] Adult patient

Discussion

Cerebellar mutism is a rare complication after surgery on the posterior cranial fossa [12] and it is a relatively benign phenomenon, characterized by a well-defined latency period and short duration. This well-defined syndrome must be distinguished from pseudobulbar palsy after posterior fossa operation in children, reported in 1984 by Wisoff and Epstein [22], which is characterized by postoperative onset of supranuclear cranial nerves palsies and swallowing difficulties associated with inability to speak and emotional lability. However, in some cases of cerebellar mutism psychic changes were also observed.

The cerebellar mutism syndrome has been described in only one adult case [18]. An aggressive or radical resection of large tumors in the vermian region is associated with this syndrome. Medulloblastoma and astrocytoma were the most frequent histological diagnoses. This finding may explain the predominance of male patients.

Ferrante et al. [6] suggest that secondary insult such as meningitis or hydrocephalus during the postoperative period can cause the loss of speech in children. In our two cases we did not find any parallel complication (infection, hydrocephalus, or postoperative hematoma).

Fraioli and Guidetti [7, 9] reported two cases of mutism after infliction of stereotactic bilateral lesions to the dentate nuclei of the cerebellum to treat spasticity. Mutism has also occurred following bilateral thalamotomy for Parkinson's disease, which may indicate the influence of superior cerebellar peduncles connecting the dentate nucleus. An interpositum nucleus lesion my also be a contributory factor [19]. The dentate nucleus has nucleocortical projections over large areas of the ipsilateral cerebellar cortex, including the vermis, and is larger than the nucleocortical projections from the interpositum and fastigial nuclei [20].

The hypothesis that mutism may occur after a lesion to the dentatothalamic tract is reinforced by the similarity between transient mutism after callosotomy [14] and cerebellar mutism. Probably the connection of the limbic system to the thalamus and basal ganglia is also disrupted, showing that the thalamus and basal ganglia are important structures of coordination and modulation of speech in both kinds of mutism, directly or indirectly through the dentato-thalamo-cortical connection. There are close relationships between the superior medullary velum, the cerebellar peduncles, and midportion of the vermis (declive folium, and tuber) [13]. The midportion of the cerebellum is often involved in IV ventricle tumors. Acute injury to this portion with or without dentate nuclei lesion can also cause speech disturbances [5]. There is a high incidence of dysarthria before surgery in patients with cerebellar tumors in paravermal and the lateral elements of the hemispheres [2]. Neurophysiological studies localize the speech modulation center to the midportion of the left paravermal area [11]. The essential anatomical substrate of mutism is probably extensive lesion of the vermian and/or paravermian cerebellar region, involving the hemispheric cortex, cerebellar peduncles, fibers from the dentato-thalamo-cortical pathway, and part of deep nuclei. A vascular ischemic disturbance such as postoperative spasm of arteries supplying the cerebellum and brainstem could be a cause of the disconnection of the dentato-thalamic pathway [6]. Cerebellar speech disturbance, like severe dysarthria, is also found in patients with transitory ischemic disturbances and hemorrhage in the posterior fossa [8]. Retraction of the cerebellum and excessive manipulation in surgery on large tumors can cause vascular spasm and edema, as well as direct compression of the cerebellar peduncle.

Patients often show a concomitant postoperative emotional lability [3, 15, 21]. Our two patients also showed this psychic symptom. The organic factors combined with the psychic factor might account for the overall mechanism. Perhaps the higher incidence in children than in adults is to be explained by the influence of the additional psychic factor.

There is no relation between this complication and the position of the patient during surgery.

We performed craniotomy in our reported cases instead of craniectomy. The other patients with cerebellar mutism reported in the literature were subjected to craniectomy [3, 5, 6, 14, 17].

Raimondi recommends that is would be better to take the tumor out piecemeal, instead of en bloc. This avoids traction on the cerebellar peduncles and infarction or edema of the brain stem [16]. He also suggests the use of laser vaporization, with continuous wave, which helps one to remove the tumor easily and precisely, without the need to dissect around the superior or lateral surfaces [16].

The use of CUSA did not avoid this complication in our cases and others [6, 14, 18].

Only one case has been reported of a child in whom magnetic resonance imaging (MRI) was performed postoperatively. Six months after operation, MRI showed encephalomalacia around the IV ventricle [5]. The possibility of this complication should always be remembered and we suggest earlier MRI postoperative studies in future cases, in order to detect and confirm lesioning of the dentato-thalamo-cortical pathways.

Acknowledgements The authors are grateful to Dr. M. Tatagiba of the Department of Neurosurgery, Nordstadt Hospital, Hannover, Germany, for his helpful suggestions and Mrs. Fiona Weber for revision of the English.

References

1. Altshuler LL, Cummings JL, Mills MJ (1986) Mutism: review, differential diagnosis and report of 22 cases. Am J Psychiatry 143:1409–1414
2. Amici R, Avanzini G, Palini L (1976) Cerebellar tumours. Clinical analysis and physiopathologic correlations. (Monographs in neural sciences, vol 4) Karger, Basel
3. Ammirati M, Mirzai S, Samii M (1989) Transient mutism following removal of a cerebellar tumor. A case report and review of the literature. Child's Nerv Syst 5:12–14
4. Benson DF (1979) Aphasia, alexia, and agraphia. Churchill Livingstone, New York
5. Dietze DD, Mickle JP (1990–1991) Cerebellar mutism after posterior fossa surgery. Pediatr Neurosurg 16:25–31
6. Ferrante L, Mastrornardi L, Acqui M, Fortuna A (1990) Mutism after posterior fossa surgery in children: report of three cases. J Neurosurg 72:959–963
7. Fraioli B, Guidetti B (1975) Effects of stereotactic lesions of the dentate nucleus of cerebellum in man. Appl Neurophysiol 38:81–90
8. Gilman S, Lechtenberg R (1981) Cerebellar dysarthria. In: Gilman S (ed) Disorders of the cerebellum. Davis, Philadelphia, pp 223–230
9. Guidetti B, Fraioli B (1977) Neurosurgical treatment of spasticity and dyskinesias. Acta Neurochir (Wien) 24 [Suppl]:27–39
10. Humphreys RP (1989) Mutism after posterior fossa tumor surgery. In: Marlin AE (ed) Concepts in pediatric neurosurgery, vol 9. Karger, Basel, pp 57–64
11. Lechtenberg R, Gilman S (1978) Speech disorders in cerebellar disease. Ann Neurol 3:285–290
12. Maire JP, Guérin J, Rivel J, San Galli F, Bernard C, Dautheribes M, Caudry M (1992) Le médulloblastome de l'enfant: incidence prognostique de l'hyperplasie vasculaire, de la nécrose, de coagulation, et de l'état clinique post-opératoire sur la survie. Neurochirurgie 38:80–88
13. Matsushima T, Rothon AL Jr, Lenkey C (1982) Microsurgery of the fourth ventricle. Neurosurgery 11:631–667
14. Nagatani K, Waga S, Nakagawa Y (1991) Mutism after removal of a vermian medulloblastoma: cerebellar mutism. Surg Neurol 36:307–309
15. Nakasu Y, Isgumi T, Nioka H, Handa J (1991) Mechanism of mutism following the transcallosal approach to the ventricles. Acta Neurochir (Wien) 110:146–156
16. Raimondi AJ (1987) Pediatric neurosurgery. Springer, New York Berlin Heidelberg
17. Rekate HL, Grubb RL, Aram DM, Hahn JF, Ratcheson RA (1985) Muteness of cerebellar origin. Arch Neurol 42:697–698
18. Salvati M, Missori P, Lunardi, Orlando ER (1991) Transient cerebellar mutism after posterior cranial fossa surgery in an adult. Clin Neurol Neurosurg 93–94:313–316
19. Siegfried J, Esslen E, Gretener U, et al (1970) Functional anatomy of the dentate nucleus in the light of stereotaxic operations. Confin Neurol 32:1–10
20. Tolbert DL, Bantli H, Bloedel JR (1978) Organizational features of the cat and monkey cerebellar nucleocortical projections. J Comp Neurol 182:39–56
21. Volcan I, Cole GP, Johnston K (1986) A case of muteness of cerebellar origin (letter). Arch Neurol 48:313–314
22. Wisoff JH, Epstein FJ (1984) Pseudobulbar palsy after posterior fossa operation in children. Neurosurgery 15:707–709
23. Yonemasu Y (1985) "Cerebellar mutism" and speech disturbance as a complication of posterior fossa surgery in children. Presented at the 13th Annual Meeting of the Japanese Society for Pediatric Neurosurgery, Tsukuba, Japan