Cukurova Medical Journal
ÇUKUROVA ÜNİVERSİTESİ TIP FAKÜLTESİ

Cukurova Med J 2024;49(1):232-234
DOI: 10.17826/cumj.1356140

LETTER TO THE EDITOR

A case of cerebellar mutism initially diagnosed as adjustment disorder with anxiety

Başlangıçta anksiyete ile giden uyum bozukluğu olarak değerlendirilen bir serebellar mutizm olgusu

Gülşah Çomak[1], Şeyma Selcen Macit[1], Ali Evren Tufan[1], Yusuf Öztürk[2]

[1]Bolu Abant Izzet Baysal University, Bolu, Turkey
[2]University of Health Sciences, Ankara, Turkey

To The Editor

Cerebellum, along with balance and motor function, also plays a role in higher cognitive processes and affect regulation[1]. Left cerebellar hemisphere may be responsible for executive tasks while right cerebellar hemisphere may play roles in language perception, speech timing, motor planning, transient storage of verbal information, verbal fluency and processing grammar[2].

"Cerebellar mutism" (CM) defines absence or severe reduction of speech along with irritability, emotional lability and social withdrawal[4]. CM may develop in 11.0-29.0 % of children after surgeries involving the posterior fossa and is especially seen among patients with medulloblastomas and/ or brainstem lesions[4]. However, cases developing after ischemic strokes involving the cerebellum have also been described[5]. In CM patients who were operated on for tumors, typically, the speech is normal for a brief period after surgery and deteriorates thereafter. In the majority of patients, mutism resolves after weeks to months although it may persist longer in some[5]. The exact pathophysiology of CM is unclear and features of the syndrome may be interpreted as psychological in origin. Here, we report diagnosis of CM in a three-years old toddler.

Psychiatric evaluation of a three-years old girl hospitalized at the pediatric ICU was asked for "periodical crying, agitation and mutism" and to rule out "anxiety due to problems in adjusting to treatment". Her complaints started after hospitalization for convulsions. She developed a febrile seizure ten days ago after an intestinal infection. Thereafter she developed status epilepticus and lost consciousness. She regained consciousness on the third day and displayed periodical agitation and crying, receptive language was preserved for basic commands while speech was limited to one word only (i.e. "mum"). Laboratory examinations at intake revealed leukocytosis (17.770) and neutrophilia (77.0 %). Lumbar puncture was negative. Cranial MRI and CT displayed reduced blood flow at right anterior internal carotid artery and ischemia in the left middle cerebellar peduncle; respectively. Electroencephalography demonstrated changes in base rhythm. Urine and blood cultures as well as an auto-antibody panel (including Anti-GAD, NMDA, AMPA1-2, GABA-B and CASPR2) was negative. No antibodies against VZV, HSV, Rubella, influenza and M. Pneumoniae could be detected. At the time of the psychiatric evaluation she was receiving 300 mg/ day levatiracetam, 800 mg/ day ceftriaxone and 1200 mg/ day acyclovir as well as adequate hydration and supportive therapy.

She was evaluated in her bed with help of her mother. Orientation was intact in all axes, communication was limited to simple wovels ("ı-ıh"), working and recent memory was reduced, remote memory was intact, spontaneous and voluntary attention and concentration was reduced, mood was anxious, affect was labile. No perceptual abnormality was noted.

Address for Correspondence: Gülşah Çomak, Bolu Abant Izzet Baysal University Faculty of Medicine Hospital, Department of Child and Adolescent Mental Health and Diseases, Bolu, Turkey E-mail: gulsahcomak3@gmail.com
Received: 31.08.2023 Accepted: 11.11.2023

Psychomotor activity was reduced and limited (i.e. left sided palsy was noted), sleep and appetite were normal. Past medical and family history were normal.

Evaluations with Delirium Rating Scale and Clinical Global Impression- Severity Scales revealed scores of 7 (no risk for delirium) and 5 (markedly ill); respectively[6,7]. According to the history and evaluations, she was diagnosed with Major Cognitive Disorder (Due to multiple etiologies with behavioral disturbance/ agitation, severe) as per DSM-5 criteria ("Cerebellar Mutism", [8]). Psychoeducation was provided to the parent and risperidone 0.25 mg/ day was started. The patient gained ambulation on the twentieth day and was discharged. Expressive language improved at discharge although it was limited to a few two-word sentences. Receptive language was intact. Risperidone was gradually tapered and stopped a week after discharge. She was still being followed by the Department of Pediatric Neurology on the third month and was receiving physiotherapy, speech and language therapy and rehabilitation. Expressive language on the second month consisted of 20-40 words and two word sentences while receptive language was intact.

Here we report a case of CM in a toddler which developed after an infection, status epilepticus and ischemia of the left middle cerebellar peduncle. Treatment of the infection, convulsions and restoration of blood flow to the CNS helped improve expressive language, although deficits persisted at the second month of follow-up. Despite extensive investigations, the offending agent could not be elucidated although the history and evaluations suggest a combination of GIS infection and CNS viral invasion led to a complicated febrile convulsion and ischemia of the CNS.

Adjustment disorder, acute stress disorder and selective mutism may be listed among the differential diagnoses of our patient although the symptom presentation was not typical of adjustment disorder and there were signs of neuropsychiatric impairment. Acute stress disorder was ruled out due to lack of avoidance, numbing and re-experiencing symptoms. Contrary to what would be expected in patients with selective mutism, her development including language and social interaction was normal prior to infection and she clearly attempted to interact and speak with both the examiner and her parent but could not due to her disability [8]. Preservation of orientation and sleep, lack of perceptual abnormalities, limited variability in symptoms ruled out delirium. In accordance with history, symptoms and laboratory evaluations the patient was though to develop CM. Reduced blood flow of the right anterior internal carotid artery probably led to left sided palsy in our patient while ischemia of the left middle cerebellar peduncle and the resulting changes at the brain stem led to the CM. As an alternative, acute cerebellitis may have been responsible for the CM in our patient, although the protracted course suggests otherwise[5].

Our case may be important in reminding the clinicians that acute cases of mutism, irritability and anxiety along with neurological signs and symptoms may have CM while highlighting the importance of middle peduncle lesions leading to cerebellar dysfunction resulting with cognitive problems.

Author Contributions: Concept/Design : GÇ, ŞSM, AET; Data acquisition: GÇ, ŞSM, AET; Data analysis and interpretation: GÇ, ŞSM, AET; Drafting manuscript: GÇ, ŞSM, AET; Critical revision of manuscript: GÇ, ŞSM, AET; Final approval and accountability: GÇ, ŞSM, AET; Technical or material support: GÇ, ŞSM, AET; Supervision: GÇ, ŞSM, AET; Securing funding (if available): n/a.
Ethical Approval: An informed consent form was taken from the patient.
Peer-review: Editorial review.
Conflict of Interest: Authors declared no conflict of interest.
Financial Disclosure: Authors declared no financial support

REFERENCES

1. Stoodley CJ, Schmahmann JD. Evidence for topographic organization in the cerebellum of motor control versus cognitive and affective processing. Cortex. 2010;46:831-44.
2. Marien P, Ackermann H, Adamaszek M, Barwood CH, Beaton A. Desmond J, et al. Consensus Paper: Language and the cerebellum: an ongoing enigma. Cerebellum. 2014;13:386–410.
3. De Witte E, Wilssens I, De Surgeloose D, Dua G, Moens M, Verhoeven J, Manto M, Marien P. Apraxia of speech and cerebellar mutism syndrome: a case report. Cerebellum Ataxias. 2017;4:1-9.
4. Gudrunardottir T, Sehested A, Juhler M, Schmiegelow K. Cerebellar mutism: review of the literature. Childs Nerv Syst. 2011;27:355-63.
5. Moscote- Salazar LR, Alvis-Miranda H, Lee A, Escorcia HF, Castellar-Leones SM. Transient Mutism and cerebellar ischemic stroke: case report. Rom Neurosurg. 2013;20:271-76.
6. Aydemir O, Noyan A, Gulseren S, Kayahan B, Bodur Z, Elbi E et al. Development of delirium rating scale, reliability and validity. Journal of Psychiatry Psychology Psychopharmacology. 1998;6:21-7.
7. Guy W. ECDEU Assessment Manual For Psychopharmachology. Rockville, MD, U.S. Department of Health, Education and Welfare. 1976.

8. American Psychiatric Association Diagnostic and Statistical Manual of Mental Disorders -5th Edition (DSM-5). Washington DC, American Psychiatric Association, 2013.