Letter to Editor

Transient mutism due to posterior circulation infarction

Sir,

Verbal mutism is described as the speechless state in a cognitively alert patient, without buccolingual apraxia. It has been a well-documented complication of resective surgery for posterior fossa tumors, especially in children. However, there are only few reports of posterior fossa mutism that are not due to neuro-trauma or neuro-surgical trauma. We report the case of transient mutism in the setting of non-traumatic posterior circulation infarction.

An 11-year-old boy, who was right handed, developed recurrent bouts of vomiting, giddiness, right hemiparesis, left upper limb incoordination and loss of speech, one day prior to presentation. There was no preceding history of fever, headache, neck pain or trauma. On admission, his blood pressure was normal. He was drowsy; moving left upper and lower limbs spontaneously, but could not vocalize any phoneme. He had evidence of unilateral pyramidal sign in the form of right extensor plantar response. The following day, he was more alert. He had left hemiataxia on testing for limb co-ordination with repeated coaxing, but no nystagmus. Though mute and withdrawn, he exhibited inconsolable whining for several hours. Swallowing, chewing, facial, tongue and neck movements were normal. Diagnostic investigation revealed normal findings on serum biochemistry, hemogram, erythrocyte sedimentation rate, sickling cell test, lipid profile and anti-phospholipid antibody work-up. Electro-cardiogram, echocardiography, carotid and vertebral doppler scan and X ray cervical spine were normal. Plain and contrast enhanced brain computed tomography imaging (CT scan) obtained on the day of admission, revealed multiple hypodensities involving part of left cerebellar hemisphere, right paravermal region, and left rostral pons. Brain magnetic resonance imaging (MRI) performed subsequently revealed hyperintense areas involving left superior cerebellar hemisphere, right paravermal region, left middle cerebellar peduncle and baso-tegmental region of left rostral pons on T2WI and FLAIR sequences (Figures a, b, c). Magnetic resonance angiogram (arterial and venous study) of the intracranial vasculature was normal. He was treated with aspirin and was placed on regular rehabilitation programme. There was gradual improvement of right hemiparesis in the first week. He could verbalize monosyllables in the third week and ill-articulated single words in the fourth week of illness. The subsequent slurred speech was slow and the words were uttered laboriously with strained voice and audible respiration. Gait was ataxic and broad based requiring one-person assistance. When examined two months later, his speech was near normal, the behavioral alteration had subsided and he could walk without assistance. Repeat cranial MRI scan revealed marked regression in the size of the right paravermal lesion (probably resolution of edema) and persistence of other infarcts, which became well delineated.

Our patient presented with transient mutism, behavioral alteration and cerebellar and right pyramidal tract signs in the setting of multiple infarcts involving the cerebellum, left middle cerebellar peduncle and left rostral pons. The evolution of speech disorder from mutism to dysarthria and the characteristic behavioral disturbance with inconsolable whining, in the absence of dysphagia or oral apraxia, anytime during the course of illness, indicated that his speech disturbance was due to transient posterior fossa mutism, also known as, cerebellar mutism. The onset and clinical course of our patient’s illness and the topography of cerebellar and brainstem...
lesions (infarcts), in the absence of supratentorial involvement or antecedent illness indicated posterior circulation stroke as the aetiology. The distribution of these ischaemic lesions was multifocal, in the areas supplied by left superior cerebellar, left anterior inferior cerebellar and the penetrating left pontine branches of basilar arteries, thus involving the middle and distal posterior circulation territories (apart from the non-territorial right paravermal infarct). Miyakita et al.\(^ 6\) reported the first case of transient cerebellar mutism after brainstem infarction following traumatic injury of right vertebral artery. However, our patient had involvement of both brainstem and cerebellar structures in the non-traumatic context. It is noteworthy that there was transient disturbance in the level of consciousness at the onset along with transient right hemiplegia and gait ataxia in both cases. This finding was in agreement with the reported association of cerebellar mutism with a spectrum of neurological deficits including pyramidal, cerebellar and eye movement signs.\(^ 1\)

We believe that a complex interaction of cerebellar and brainstem involvement might be the cause of mutism in our patient. One of the suggested mechanisms for postoperative cerebellar mutism includes brainstem ischemia and subsequent edema, to account for the transient nature of the syndrome.\(^ 1\) The vascular basis of this syndrome in the nonsurgical setting is better exemplified by our report. The good outcome of the speech disorder (including the improvement of dysarthria with the return of near normal speech during the subsequent period of observation), despite the presence of established infarcts, could be attributed to unilateral brainstem infarction, resolution of edema or neuronal plasticity. The behavioral disturbance also normalized with the resolution of mutism. Hence recognizing this syndrome is important in informing the parents about the prognosis and rehabilitation of speech and behavioral disorder, as most of these cases were encountered in the pediatric age group.

R. Nandagopal, S. G. Krishnamoorthy
Department of Neurology, Sri Venkateswara Institute of Medical Sciences, Tirupati, Andhra Pradesh, India.
E-mail: mandagopal@yahoo.com

References


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Diagnostic approach for adult mitochonrdiopathy with limited resources

Sir,

We appreciated reading the paper by Challa et al.\(^ 1\) on the findings in 60 adult patients with respiratory-chain-disorder (RCD), diagnosed during 12y according to Bernard’s criteria.\(^ 2\) The study raises the following concerns:

What was the indication for muscle-biopsy in those patients without evidence for myopathy? Only 18/60 patients presented with proximal weakness, only 5/35 had elevated CK, and only 12/31 myopathic EMG.

Why was the left vastus lateralis muscle chosen in all cases? It appears unlikely that this muscle was affected in all 60 patients, given the facts that 26 had only chronic external ophthalmoplegia (CPEO), that only 3 of those with CPEO and other signs had myopathy, and that only 30% had proximal weakness. Thus, RRFs >2% in 25/60 investigated muscles suggests subclinical involvement of certain limb muscles in MCP.

Why did only 8% of the patients present with additional non-neurological manifestations? Multisystem involvement is a dominant feature in the majority of MCP patients.\(^ 3\) Could the low number of patients with cardiac involvement result from the small number of patients undergoing comprehensive cardiological examination? Only 35 had an ECG and only 13 underwent echocardiography. Which specific abnormalities were found in the 13 who underwent echocardiography? Did the investigators also look for left-ventricular hypertrabeculation?

Also noteworthy are the low prevalence of diabetes, hyper-CK-emia, and polynuropathy. In a retrospective study on 130 MCP-patients, the prevalence of diabetes was 12%,\(^ 4\) that of hyper-CK-emia 42%, and that of polynuropathy 35%.\(^ 5\) Latency between onset and diagnosis was relatively short. The number of 6.7y remains questionable given the fact that mean age at onset was 19.7y and mean age at diagnosis 29.3y.

We don’t agree with the statement that MCP is mostly associated with various central-nervous-system (CNS) symptoms. Although subclinical abnormalities, like cortical or cerebellar atrophy, homogenous parieto-temporal hyperintensities, non-specific white matter lesions, or uni- or bilateral basal ganglia calcifications are often found in adult MCP patients, overt CNS-affection, including dementia, migraine, stroke, Parkinson syndrome, spasticity, ataxia, or dystonia, is rather rare among adults. The authors themselves report only 5/60 patients presenting with encephalomyopathy.

Overall, there is subclinical muscle-affection in MCP long before any clinical manifestation; MCP can be diagnosed even without sophisticated and cost-expensive methods; there is