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Post-Infarct Cerebellar Cognitive Affective Syndrome: A Case Report

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Abstract

Post Infarct cerebellar cognitive affective syndrome is a rare disorder, characterized by cognitive impairment in the domains of memory, language, visuo-spatial functioning and affect after cerebellar stroke. We report a case of young female who developed mood alteration and cognitive disturbance following isolated cerebellar infarct. We, therefore, advocate a potential role of cerebellum in regulation of cognition and behaviour in humans

Introduction

'Cerebellar Cognitive Affective Syndrome' is the term, used to describe cognitive impairment in the domains of memory, language, visuo-spatial functioning and affect in the presence of an isolated cerebellar pathology. Lesions in the posterior cerebellar lobe and vermis may result in clinically prominent behavioural and cognitive impairments whereas the cognitive changes associated with the lesions of the anterior cerebellar lobe are minimal.¹ The evidence is based mainly on case series of children with cerebellar tumours, who underwent surgical resection and adults with multiple etiologic processes.^{1,2,3} There are very few reports of this syndrome as a result of isolated cerebellar infarctions and to our knowledge none is reported from Pakistan.⁴⁻⁶

Case Report

A 30 years old married lady presented to emergency room (ER) with progressive drowsiness over a period of 24 hours, preceded by nausea, vomiting and vertigo. There was no prior history of such symptoms. There was no history of fever, trauma or intake of any medication, preceding this illness. She did not have any history of diabetes mellitus, hypertension, smoking or alcohol intake. In ER her blood pressure was 130/80 mmHg and radial pulse was regular at 82 beats per minute. Her neurologic examination was significant for disorientation to time and place, mild drowsiness, dysarthria, gaze evoked horizontal nystagmus, quadriparesis with power of 4/5 on MRC grading system of power, pathologically brisk deep tendon reflexes and extensor planters. She was unable to walk unaided. There was no sign of meningeal irritation. Systemic examination was unremarkable. Complete blood count, serum electrolytes, creatinine, prothrombin time and activated partial thrombin time were normal.



Figure 1. MRI brain coronal FLAIR image showing acute infarction of left cerebellar hemisphere (arrow) and vermis, slightly towards right (arrow head).

MRI brain revealed acute cerebellar infarction involving vermis and superior part of left cerebellar hemisphere (Figure 1). There was no cerebral and brain stem infarction. The patient was admitted in stroke unit and started on standard stroke management i.e. antiplatelet agents, statins. Over 24 hours of admission she deteriorated and became comatose (GCS 7/15). Pupils were equally reactive to light at that time and fundi were also normal. An urgent CT scan of head was done which revealed the infarction and mass effect leading to compression of cisterns around the brainstem, on left side. She was given intravenous mannitol and was intubated for mechanical ventilation. She was kept on ventilator support for 8 days. One day after extubation, she was able to follow one step command and was moving all four limbs with power of 2/5 on MRC scale. However, she was unable to talk.

During the proceeding course of the hospital stay, an alteration of behaviour and cognition was observed. This consisted of frequent spontaneous and intractable bouts of laughter and crying spells. The patient was also unable to recognize her family members, including her husband. An alteration of sleep pattern, drooling and excessive salivation was seen. An in-depth exploration into personal, past medical, psychosocial and family history did not reveal any clues pointing towards a psychiatric pathology.

The patient was initially started on fluoxetine and was later given amitriptyline. Within a period of two weeks she started to improve. The frequency of the laughter and crying spells decreased and she started recognizing her family members. A marked improvement in the memory function and sleep patterns was also noted.

A detailed workup to determine the cause of stroke was performed. The tests for hypercoagulopathy were negative and echocardiogram was normal. MRA of head and neck revealed atherosclerotic disease affecting both the vertebral arteries and the basilar artery.

Patient's hospital course was complicated by infections as ventilator associated pneumonia and line sepsis. She was discharged from the hospital after 25 days. At the time of discharge, she was afebrile, could speak small phrases, following three steps command and displaying appropriate behavior. However, she continued to have spontaneous laughter spells.

She was followed up in outpatient neurology clinics up to 6 months after her discharge and except occasional inappropriate laughter spells she was making satisfactory progress.

Discussion

Role of cerebellum in motor functioning is well established. However, in the last couple of decades researchers have noted cognitive, behavioural and affective abnormalities as a result of cerebellar dysfunction, suggesting possible role of cerebellum in higher mental function processing.

A potential role of cerebellum in cognitive functioning was first suggested by Botez et al as early as 1985.⁷ This was based on the observation that cerebellar ataxia due to Phenytoin toxicity in two epileptic patients was accompanied by cognitive deficits and behavioural dysregulation. Since then the notion has been reinforced by case reports and series. Levisohn et al² reported a series of 19 children, who underwent cerebellar tumour resection in childhood, in whom formal in depth neuropsychological assessment revealed impairment of executive functioning, language expression, visuo-spatial functioning, alteration of affect and behaviour.¹ The studied population did not undergo any cranial irradiation and chemotherapy, which could have potentially altered the results of the investigation. Riva et al³ in their series of 26 children observed problems in memory and language functions. They also noted that left hemispheric lesions were associated with visuo-spatial problems while right hemispheric lesions with auditory memory and language processing. Similar hemispheric topographic effects are reported with isolated cerebellar stroke.⁶ Impairment in

similar domains have been found in patients with isolated cerebellar degeneration.⁸ Abel et al⁸ published a series of 12 adult patients with remote cerebellar degeneration and demonstrated substantial cognitive deficits in the spheres of executive, memory and visuo-spatial functioning, on standardized battery of neuropsychological tests.

A few case series and some case reports of post stroke cerebellar affective cognitive syndrome suggest the possible role of cerebellum in higher mental functioning.⁴⁻⁶ Cognitive deficits ranging from subtle emotional disturbances, impairment of executive functioning, deficits in spatial cognition, language disturbances and behavioural modifications have all been reported with isolated cerebellar infarctions. Our patient exhibited most of the above mentioned behavioural disturbances, following an isolated cerebellar infarction.

This syndrome may mimic post stroke depression, especially if affective symptoms like crying spells are predominant but laughter spells, disorientation and significant cognitive, behavioural manifestations and lack of insight are important features to differentiate this syndrome from post stroke depression. Subtle form of this syndrome will probably require formal neuropsychologic testing to differentiate the two syndromes.

The constellation of these clinical observations has resulted in the birth of the term 'Cerebellar Cognitive Affective Syndrome' which has been observed to be associated with lesions of vermis and posterior cerebellum.¹ Our patient also had infarction of the vermis. This has been further supported by the observation that lesions in the territory of posterior inferior cerebellar artery (PICA) resulted in cognitive and affective deficits whereas cognition and affect were unaltered in the lesions involving territory of superior cerebellar artery (SCA).⁵ Our patient had stroke involving left SCA region and vermis. The clinical syndrome in our patient can be explained on the basis of infarction of vermis but strokes of SCA region have been reported to be associated with a similar syndrome.¹ Moreover, a prospective analysis of 26 patients with cognitive impairment after isolated cerebellar infarction demonstrated a transient course of the cognitive impairments.⁶ Our patient also made a remarkable recovery.

A scientific basis for the role of cerebellum in cognitive and behavioural control has also been provided by anatomical and functional studies both in humans and other primates. It has been explicated that cerebellum is connected to the pre-frontal, occipito-parietal and temporal cortical association areas as well as the limbic system through complex circuitries. Studies employing functional imaging have also revealed the activation of cerebellum in performance of tasks which are purely cognitive and are not

related to movement.⁹ It is imperative to keep under consideration that although much convincing evidence supporting a modulatory role of cerebellum in cognition and affect has amassed, a few clinical observations fail to reiterate this concept. Most notable of such studies has been the work of Richter et al on patients with chronic cerebellar infarction.¹⁰ They demonstrated an absence of cognitive deficits in 21 adult patients with chronic cerebellar infarctions both in territory of PICA and SCA based on an objective bed-side mental examination.¹⁰ However, this simply may imply that such deficits with cerebellar stroke are transient and likely to improve over a course of weeks to months.

Though we did not subject our patient to formal neuropsychological testing, clinically prominent symptoms/signs and a clear temporal relation to the stroke suggest this being post stroke cerebellar cognitive affective syndrome. We, therefore, advocate a potential role of cerebellum in regulation of cognition and behaviour in humans, based upon our clinical observation of transient cognitive and affective impairments in a 30 year old female after isolated cerebellar infarction. To the best of our knowledge this is the first reported case from Pakistan and the neighboring countries. This case substantially adds to

the repertoire of clinical evidence favouring the notion.

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