Case Report

International Journal of Neurology and Neurosurgery
Volume 10 Number 3, July - September 2018
DOI: http://dx.doi.org/10.21088/ijnns.0975.0223.10318.22

Unilateral Cerebellar Aplasia in Adult–A Rare Incidental Finding

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Abstract

Unilateral cerebellar aplasia is a very rare developmental disorder usually most commonly seen in children after a prenatal insult. It hasmultifarious clinical presentations ranging from development delay to ataxia and cognitive impairment. Here we present a case of a 31-year old adult whopresented with acute confusional state (probable seizure) have incidentally found to haveunilateral left cerebellar aplasia. He had normal uneventful delivery with normal development and cognition.Though the exact cause is unknown, even with unilateral cerebellar aplasia there can still have an uneventful life with normal motor activity and intelligence.

Keywords: Cerebellar Aplasia; Normal Adult; Incidental Finding.

Case Report

A 31 -year old newspaper boy presented with acute onset disorientation for one day. On 18/12/2017 at 6am after distribution of paper, his bystanders noticed him sitting alone in the temple not answering to calls and unable to identify his family members or recall his address. No history of fever, headache, vomiting, drug abuse or alcohol abuse. He was born out of non-consanguineous parents and his past history was unremarkable. On examination, GCS was 14/15(E4M6V4), disoriented to time, place and person. He was not able to name things and kept on repeating words continuously. His motor system was unremarkable except for bilateral Babinski's positive. Following next day, his symptoms resolved completely and was eventually amnestic of the previous day events. His MMSE was 30/30, with normal gait and coordination and bilateral plantar flexor. With clinical diagnosis of acute confusional state he was evaluated. His complete blood counts, urine analysis, urea, creatinine, electrolytes, ammonia, TSH, fasting B12 levels and chest X-ray were normal and showed no evidence of infection. Electroencephalogram (EEG) showed no epileptiform discharges. Magnetic resonance imaging (MRI) of Brain (Figure 1 A, B, C, D) showed asymmetry of the

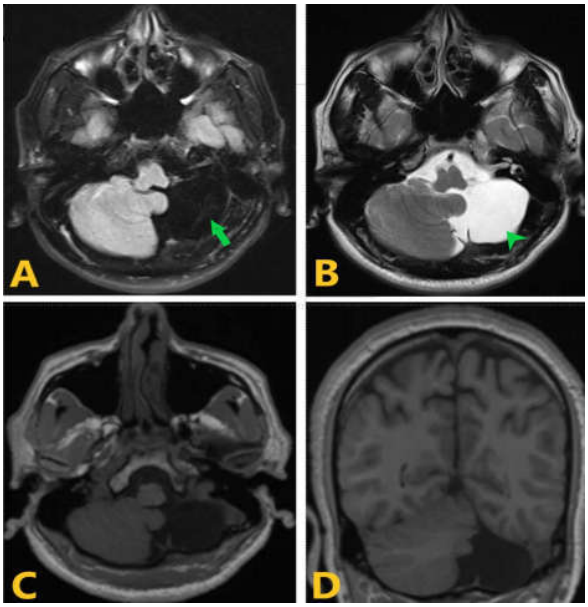

Fig. 1: Magnetic resonance imaging of Brain plain shows hypodensity in fluid attenuated inversion recovery sequence (A) (arrow), T1 weighted axial (C) and coronal (D) sections and hyperdensity in T2 weighted image (arrow head) suggestive of aplasia.

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Received on 18.07.2018, **Accepted on** 31.08.2018

cerebellum with complete unilateral left cerebellar aplasia (an incidental finding) with normal brain stem and cerebrum. Acute confusional state was probably thought secondary due to complex partial seizures with an incidental left cerebellar aplasia. He was treated with anti-epileptics following which his symptoms resolved completely.

Discussion

Unilateral cerebellar aplasia is a sporadic developmental disorder characterized by a decrease in volume of cerebellum secondary to delayed development or insult to the cerebellum. It is most commonly seen in children after a prenatal insult and has various clinical presentations ranging from development delay, ataxia to cognitive impairment. Cerebellum has a diversity of features occupying about 20% of brain volume in the human brain [1]. Its primary function is for maintenance of balance and posture and coordination of fine skilled movements. It also helps in motor learning and has a role in cognitive functions. Any insult causes derangement in cerebellar function causing varied symptoms like delayed development, ataxia to intellectual disability. If cerebellar vermis is involved, it is associated with poorer cognitive outcome, and an intact vermis has a normal result and no truncal ataxia. It is related to a variety of neurological and systemic disorders like congenital malformation syndromes (such as Walker-Warburg syndrome); inherited metabolic disorders, such as Williams syndrome, and some neurodegenerative disorders that begin in early childhood, such as ataxia telangiectasia [2].

Here we have noticed that unilateral cerebellar aplasia in adults with normal motor and cognitive functioning. There is no history of any previous nutritional or metabolic insult. So far only one such case was presented [3]. Though the exact cause of unilateral cerebellar aplasia cannot be identified but can still, affected individuals can have an uneventful life with normal motor activity and intelligence. In our case, there was an incidental finding of left cerebellar aplasia with no other associated neuromuscular disease or developmental delay. As there was no evidence of any focal deficits and cognitive decline, probably considered etiology was secondary to congenital asymptomatic unilateral cerebellar aplasia. He also had no evidence of a history of trauma or anoxic insult.

Conclusion

Hence we conclude that though cerebellum has varied integrative functioning in humans, an incidental finding of unilateral cerebellar aplasia can be still noticed incidentally with normal motor and cognitive functions.

Acknowledgements

Nil. No Source(s) of support in the form of grants, equipment, drugs, or all of these.

Conflict of Interest: Nil

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