CASE REPORT

https://doi.org/10.3126/njr.v13i1.57832

Hemiatrophy of The Cerebral Hemisphere with Contralateral Cerebellar Atrophy: A Case Report

Dhakal P, Khadka H, Sharma S

Department of Radiology and Imaging, National Academy of Medical Sciences Bir Hospital, Kathmandu, Nepal

Received: September 18, 2022 **Accepted:** April 25, 2023 **Published:** June 30, 2023

Cite this paper:
Dhakal P, Khadka H, Sharma S. Hemiatrophy of The Cerebral Hemisphere with Contralateral Cerebellar Atrophy: A Case Report. *Nepalese Journal of Radiology* 2023;13(1):28-31. **https://doi.org/10.3126/njr.v13i1.57832**

ABSTRACT

Childhood cerebral hemiatrophy is a rare clinical condition. Its causes are numerous, but they can be divided into two categories: congenital and acquired. The congenital type develops intrauterine, whereas the acquired type develops early in life, usually before the age of two. Cerebral hemiatrophy syndromes are characterized by a wide range of neurological symptoms. Childhood epilepsy, mental retardation,and neuropsychiatric disorders are common, whereas movement disorders such as highly asymmetric parkinsonism or hemidystonia, as well as neuropsychiatric problems, have been reported in adults. Here we present a 22-year-old patient who presented with refractory seizure. Imaging findings showed hemiatrophy of the right cerebral hemisphere with left cerebellar atrophy.

Keywords: *Adult; Dystonic Disorders; Movement Disorders; Dystonia*

INTRODUCTION

In pediatric clinical practice, cerebral hemiatrophy (CHA) is uncommon. It exists, however, and it could be primary or secondary. As a result of the de-novo lack of cerebral development, the primary (congenital) CHA may be intertwined or aptly called cerebral hemi-hypoplasia or unilateral cerebral hypoplasia. The insult occurs in-utero, resulting in a shift of midline structures to the disease's side and the absence of sulcal prominence. These characteristics distinguish it from secondary CHA, which may be caused by a cerebrovascular lesion, an inflammatory process, or cranial trauma. The characteristic imaging findings of a decreased cortical volume and an enlarged ipsilateral lateral ventricle on an MRI are used to make the diagnosis of cerebral hemiatrophy. Mild contralateral cerebellar atrophy may also be present. Congenital hemiatrophy patients exhibited better seizure control than those with acquired disease. Hemidystonia is another symptom of cerebral hemiatrophy disease. Hemidystonia hemiatrophy may be brought on by a static event, such as a stroke, hypoxia, or trauma that affects the putamen and thalamus in the basal ganglia.[1,2,3,4,5]

CASE REPORT

Here we presented a case of 22 years old male patient, who presented to our neuro outpatient department with a history of weakness in the left half of his body since the age of 14 years. He had also an abnormal movement of the left side of his limbs (both upper and lower). His father reported that he had bee

Correspondence to: Dr. Prakash Dhakal
Department of Radiology and Imaging
National Academy of Medical Sciences
Bir Hospital, Kathmandu, Nepal
Email: *dhakalprakashmed@gmail.com*

experiencing episodes of generalized tonic-clonic seizures with brief periods of loss of consciousness for 8 years. He also reported developmental delay and intellectual impairment since childhood. No similar illness was noted in family members. No significance and medical and surgical illness was noted in the past. On general examination, vitals were within normal limits. The patient was irritable with abnormal body movement. On neurological workup, power was 3/5 on the left upper and lower limbs. The sensation was decreased in the left half of the body as compared to the right half of the body. A mild facial tilt towards the left side was also noted. Biochemical parameters were within normal limits.

MRI images demonstrate atrophic changes in the right cerebral hemisphere, right side of the midbrain and contralateral (left) cerebellar hemisphere with ex-vacuo dilatation of the right lateral ventricle. In addition, there is calvarial thickening in the frontal region of the skull. These features are likely representative of crossed cerebellar diaschisis. (Figures 1 & 2)

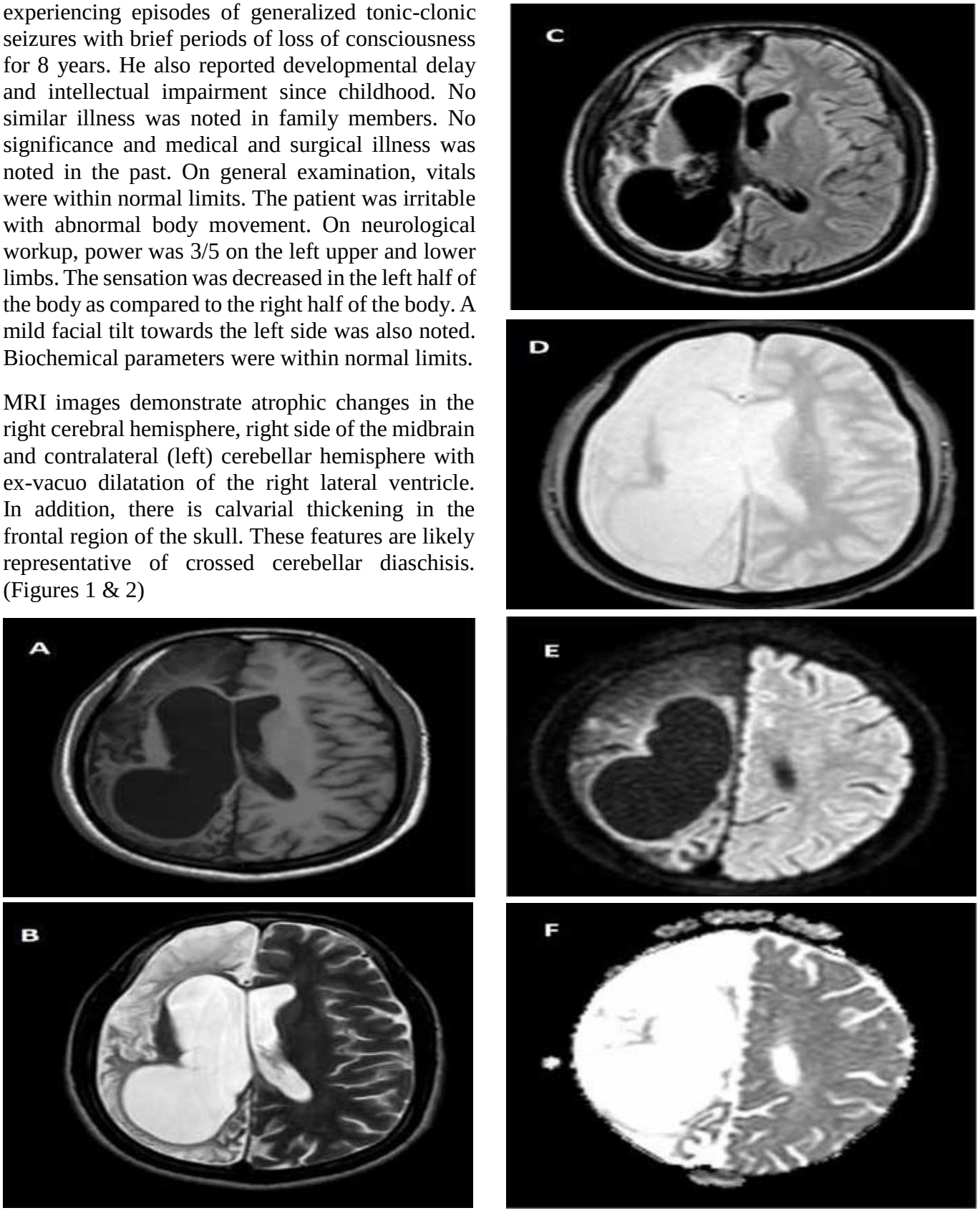

Figure 1: *T1 weighted (A), T2 weighted (B) and FLAIR image (C) of MRI brain in axial plane shows atrophy of right cerebral hemisphere with ex-vacuo dilatation of ipsilateral lateral ventricle. GRE sequence (D) do not show foci of blooming artefacts. DWI/ADC sequences (E and F) do not show areas of diffusion restriction.*

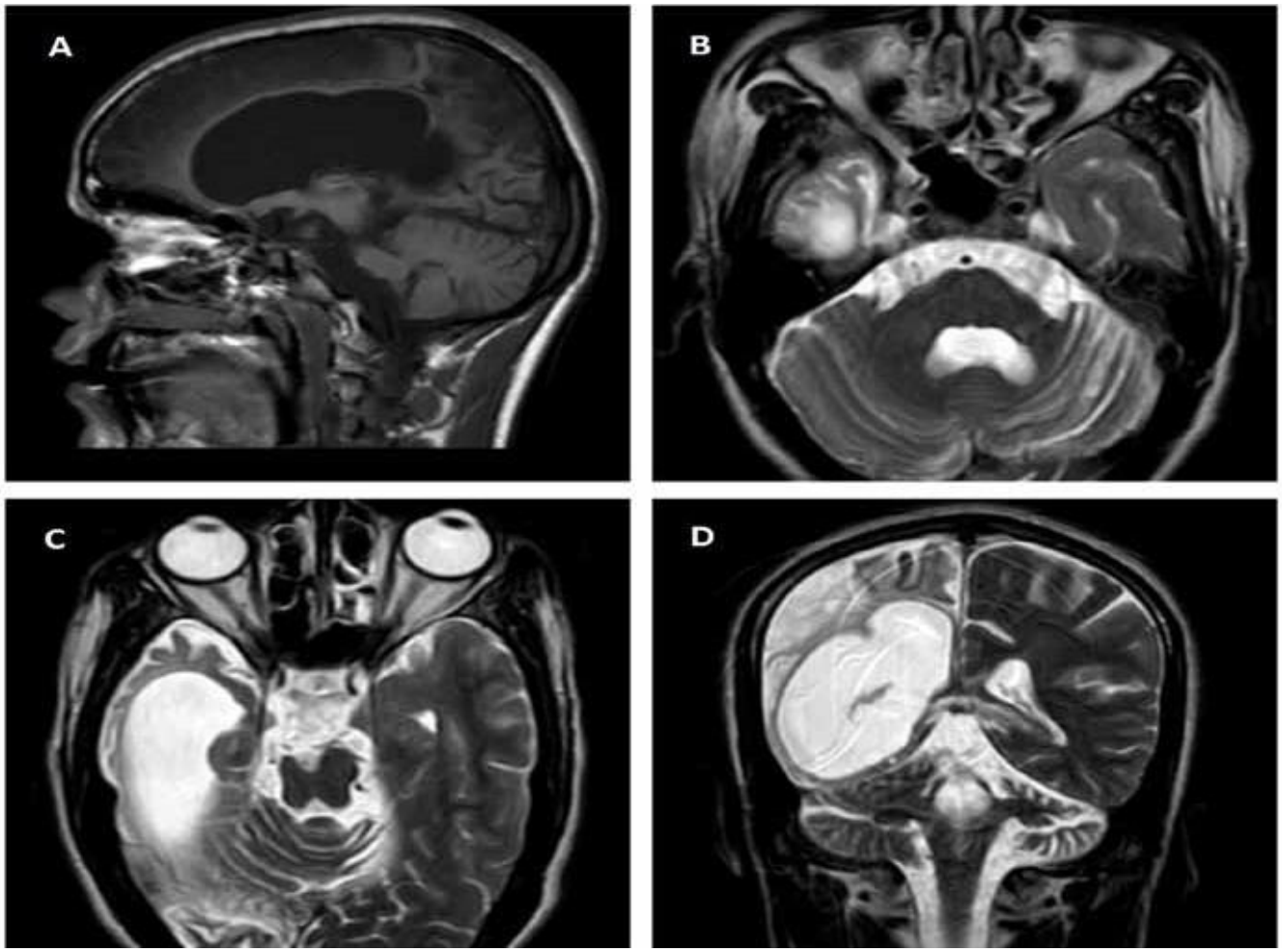

Figure 2: A).*T1 weighted image in the sagittal plane shows calvarial thickening in the right frontal region of the skull.* ***B).****T2 weighted image in the axial plane shows an atrophic left cerebellar hemisphere with prominent cerebellar folia.* ***C).****T2 weighted MRI image in the axial plane demonstrates atrophy of the midbrain on the right side. Dilated temporal horn of the right lateral ventricle is also noted.* ***D).****T2 weighted image in the coronal plane demonstrates atrophy of the right cerebral hemisphere and contralateral (left) cerebellar hemisphere*

DISCUSSION

The condition known as cerebral hemiatrophy (CHA) has a variety of etiologies and a clear asymmetry of the cerebral hemispheres. It could come from a single occurrence or a spectrum that develops over time. As the brain grows, it pulls outward on the calvarial tables, emphasizing the adult head's steady increase and general form. Finding compensatory calvarial changes assumes that cerebral abnormalities are the result of atrophic or hypoplastic changes brought on by childhood brain trauma (usually before 2 years). The condition known as crossed cerebellar diaschisis (CCD) is characterized by a decrease in the cerebellum's oxidative metabolism, glucose metabolism, and blood flow as a result of a supratentorial lesion in the opposing cerebral hemisphere. In our case also there was atrophy of the right cerebral hemisphere with atrophy and functional impairment of the left side of the cerebellum indicating crossed cerebellar diaschisis. Neurological symptoms that are present in cerebral hemiatrophy disorders can vary. Contralateral hemiplegia, refractory seizures, cognitive deficits, as well as emotional and behavioural problems, are frequent clinical outcomes of cerebral hemiatrophy. For effective therapy and prognostication of cerebral hemiatrophy, determining the etiology is crucial. Compared to children with atrophy or gliosis alone, children with cystic encephalomalacia and atrophy had a worse prognosis. Morphometric factors such as lesion size, white matter integrity, and cortical grey matter volumes are predictors of intellectual functions. Common differentials for hemiatrophy of the brain are Sturge-Weber syndrome, Parry-Romberg syndrome, Rasmussen's encephalitis and

Dyke-Davidoff-Masson syndrome (DDMS).[2,6,7]

Sturge Weber syndrome (SWS) is characterized by choroidal angioma, leptomeningeal angiomatosis, and face port wine stain (PWS). One of the key characteristics of SWS is leptomeningeal angiomatosis, which is evident as aberrant leptomeningeal enhancement on contrast-enhanced magnetic resonance imaging (MRI), often in the parieto-occipital lobe distribution, ipsilateral to the facial PWS. Unicerebral atrophy or hypoplasia in children with DDMS, also known as hemispheric infarction, is accompanied by compensatory ipsilateral calvarial enlargement. This uncommon illness results from brain injury during infancy or the early years of childhood.The injury could be in the form of trauma, inflammation, vascular, congenital, or acquired ischemic disease. In our case, lack of port wine stain, leptomeningeal angiomatosis, and calvarial thickening rules out SWS and DDMS. Another differential is Rasmussen encephalitis which is an uncommon, unihemispheric neurological condition that progresses over time and is characterized by late-stage progressive cognitive deterioration,intractable focal seizures, and hemiparesis in up to 85% of instances, the disease starts within the first decade of life. Treatment modalities include anti-epileptic medications and hemidecortication. Our patient was under antiepileptic medications for 4 years with minimal improvement of symptoms anddecreased frequency of seizure episodes.[2,4,7,8]

CONCLUSION

Cerebral hemiatrophy is an uncommonly encountered clinical entity. It has different aetiology and clinical features. Among them, refractory seizure is one of the common presentations of the patient. For the diagnosis, MRI is the imaging modality of choice. Thus early imaging is helpful to diagnose the disease which helps in timely and appropriate treatment. In the case of developing countries like Nepal, because of the lack of MRI in rural areas diagnosis of hemiatrophy is delayed leading to late diagnosis of the disease and treatment. Early referral to higher centres is required for patients who presented with seizures in rural areas.

CONFLICT OF INTEREST

None

SOURCES OF FUNDING

None

REFERENCES

1. Dix JE, Cail WS. Cerebral hemiatrophy: classification on the basis of MR imaging findings of mesial temporal sclerosis and childhood febrile seizures. R*adiology* 1997;203(1):269-74. https://doi.org/10.1148/radiology.203.1.9122406
2. Mathieu M, CN OP, Ongolo PC, Muna W. Differential diagnoses of cerebral hemiatrophy in childhood: a review of literature with illustrative report of two cases. *Glob J Health Sci* 2013;5(3):195-207. https://doi.org/10.5539/gjhs.v5n3p195
3. Trivedi A, Radadiya K, Kalola J, Jethva, Chawla A, Pankhania M. A case of cerebral hemi atrophy with associated mesial temporal sclerosis in a child. Available from; http://dx.doi.org/10.1594/EURORAD/CASE.14015 [Accessed 15th March 2023]
4. Jaiswal A, Garg RK, Malhotra HS, Verma R, Singh MK. Seizures in patients with cerebral hemiatrophy: A prognostic evaluation. *Ann Indian Acad Neurol* 2015;18(1):39-44. https://doi.org/10.4103/0972-2327.144296
5. Reiter E, Heim B, Scherfler C et al. Clinical heterogeneity in cerebral hemiatrophy syndromes. *Mov Disord Clin Pract* 2016;3(4):382-8. https://doi.org/10.1002/mdc3.12301
6. Han S, Wang X, Xu K, Hu C. Crossed cerebellar diaschisis: Three case reports imaging using a tri-modality PET/CT–MR system. *Medicine* 2016;95(2):e2526.https://doi.org/10.1097/MD.0000000000002526
7. Tan AP, Wong YL, Lin BJ, Yong HR,Mankad K. Clinico-radiological approachto cerebral hemiatrophy. *Childs Nerv Syst* 2018;34(12):2377-90. https://doi.org/10.1007/s00381-018-3934-3
8. Ignelzi RJ, Bucy PC. Cerebral hemidecortication in the treatment of infantile cerebral hemiatrophy. *J Nerv Ment Dis* 1968;147(1):14-30. https://doi.org/10.1097/00005053-196807000-00002