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Case Report

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Rare Case of Childhood Gliomatosis Cerebri - A Case Report

Prakash Dhakal[1*], Suraj Sharma[2], Nirmal Prasad[3]

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

[*]Corresponding Author
Prakash Dhakal, Department of Radiology, National Academy of Medical Sciences, Bir Hospital, Kathmandu, Nepal.

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Abstract

Gliomatosis Cerebri (GC) is an uncommon diffuse infiltrating glial neoplasm of astrocytic origin, which is characterized by the involvement of at least three cerebral lobes. It has a very poor prognosis. GC primarily affects adults, it is rare in children and challenging to diagnose. In children, GC frequently manifests as a combination of signs and symptoms that may mimic encephalitis. We presented an 11 year old male with a diagnosis of gliomatosis cerebri who presented to our neuro outpatient department (OPD) with a history of seizure, headache, nausea, vomiting and altered mental status for eight days.

Keywords: Acute Disseminating Encephalomyelitis, Diffuse Astrocytoma, Encephalitis, Gliomatosis Cerebri

List of Abbreviations

ADC- Apparent Diffusion Coefficient
ADEM- Acute Disseminating Encephalomyelitis
Cho/Cr- Choline/Creatine ratio
Cho/NAA- Choline/N-Acetyl Aspartate ratio
CSF- Cerebrospinal Fluid
CT- Computed Tomography
DWI- Diffusion Weighted Imaging
ESR- Erythrocyte Sedimentation Rate
FLAIR- Fluid Attenuation Inversion Recovery
GC- Gliomatosis Cerebri
GRE-Gradient Echo
H&E- Hematoxylin and Eosin Stain
IVIg- Intravenous Immunoglobulins
MRI- Magnetic Resonance Imaging
MRS-Magnetic Resonance Spectroscopy
WHO- World Health Organization

1. Background

Gliomatosis Cerebri (GC) is a rare and widespread type of glioma. According to the World Health Organization (WHO), it is a diffuse glial tumor that infiltrates at least three cerebral lobes, frequently bilateral and preserving anatomic architecture [1,2]. GC primarily affects adults. It is extremely uncommon in children, with only about 60 cases reported in the literature till date [3,4]. In children it presents with nonspecific signs and symptoms like seizures, hemiparesis, ataxia, lethargy, and symptoms of intracranial hypertension [5,6]. GC can be mistaken for more common disorders such as encephalitis and Acute Disseminated Encephalomyelitis (ADEM), causing a delay in diagnosis and treatment.5 Magnetic Resonance Imaging (MRI) is the imaging modality of choice for GC [7]. Although the prognosis is poor and GC is always fatal, there is evidence that treatment improves short-term survival. In cases of widespread infiltrating brain lesions, it is critical to include GC in the differential diagnosis. In order to definitively diagnose GC, stereotactic biopsy to acquire tissue for pathological evaluation is finally required because clinical presentation, Cerebrospinal Fluid (CSF) investigation, and neuroimaging are frequently vague.

2. Case Presentation

Here we present an 11-year-old male who came to the neurology outpatient of Bir Hospital with a history of frequent seizures, nausea, vomiting, and altered mental status for eight days. His parents also noticed he had learning difficulties and decreased cognitive activity since a month. The patient did not have any history of fever, myalgia or flu like symptoms recently. There was no history of surgery, trauma or other illnesses in the past. Family history was negative for similar episode.

On general examination, the patient was irritable with irrelevant talks. Neurological workup revealed 3/5 power in both upper and lower limbs. His blood counts, Erythrocyte Sedimentation Rate (ESR), hepatic and renal function tests were within normal limits.

3. Imaging

Magnetic Resonance Imaging (MRI) demonstrated an ill-defined

lesion in the white matter of left frontoparietal lobe with extension to the white matter of right frontal lobe. The lesion also involved rostrum, genu and body of the corpus callosum without involvement of basal ganglia or brain stem. The lesion showed low signal intensity on T1 weighted image (Fig 1A), and high signal intensity on T2/Fluid Attenuation Inversion Recovery (FLAIR) sequence (Fig 1B&C). There were no blooming artifacts on Gradient Echo (GRE) sequences. (Fig 2A). The lesion showed diffusion restriction in its peripheral aspect (Fig 2 B&C) on Diffusion Weighted Imaging (DWI) and low signal intensity on Apparent Diffusion Coefficient (ADC) mapping. The lesion did not show any contrast enhancement (Fig 3).

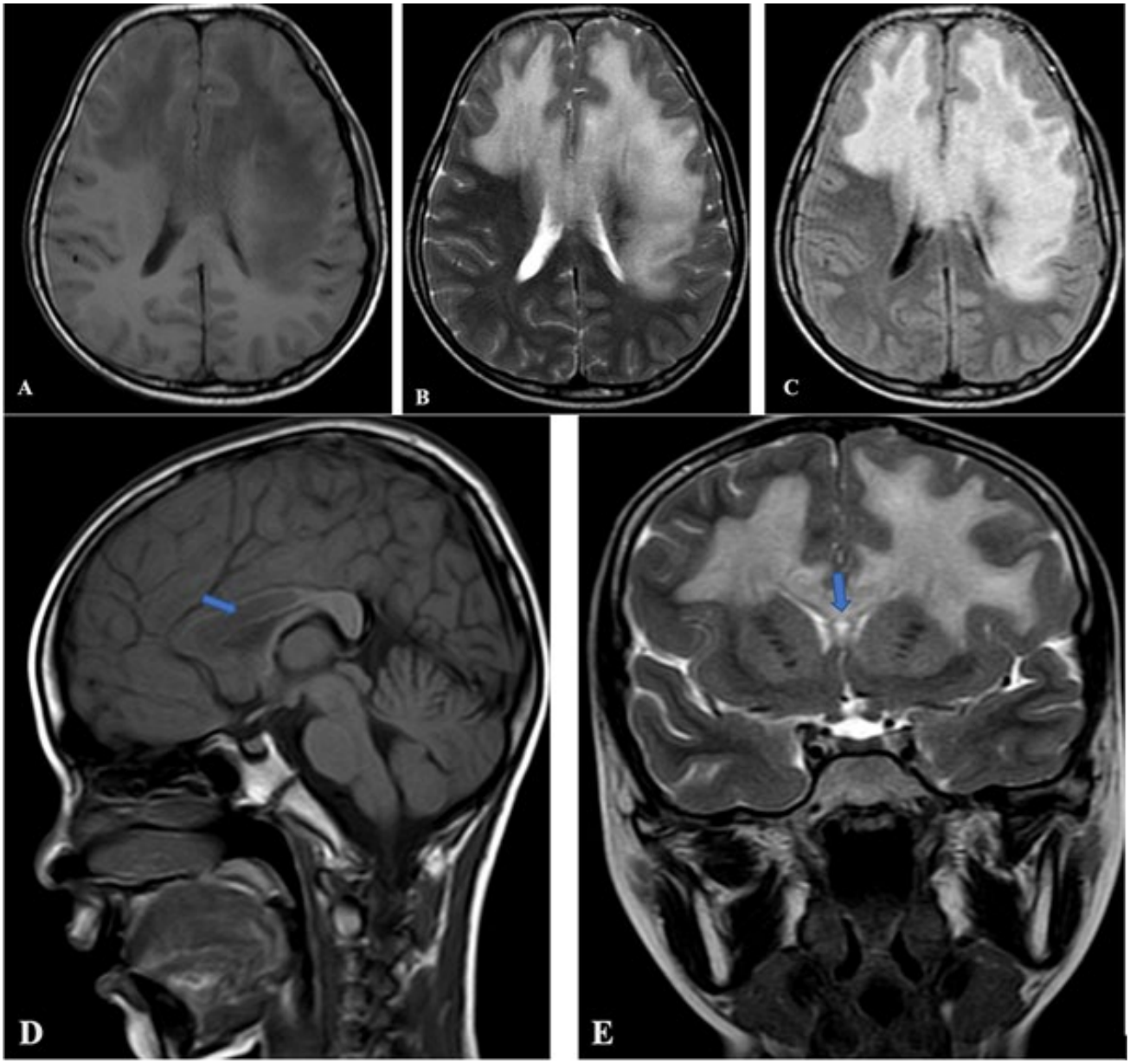

Figure 1: MRI images (A-Axial T1 weighted image, B-Axial T2 weighted image, C-Axial FLAIR image) demonstrating low signal intensity lesion in bilateral cerebral hemisphere, involving white matter of right frontal lobe, left frontal and parietal lobes. The lesion is not suppressed in FLAIR sequence. D-Sagittal T1 weighted image demonstrating involvement of corpus callosum involving rostrum, genu, and body (arrow). E-Coronal T2 weighted image demonstrating T2 high signal intensity lesion involving the white matter of bilateral cerebral hemispheres asymmetrically with involvement of corpus callosum (arrow).

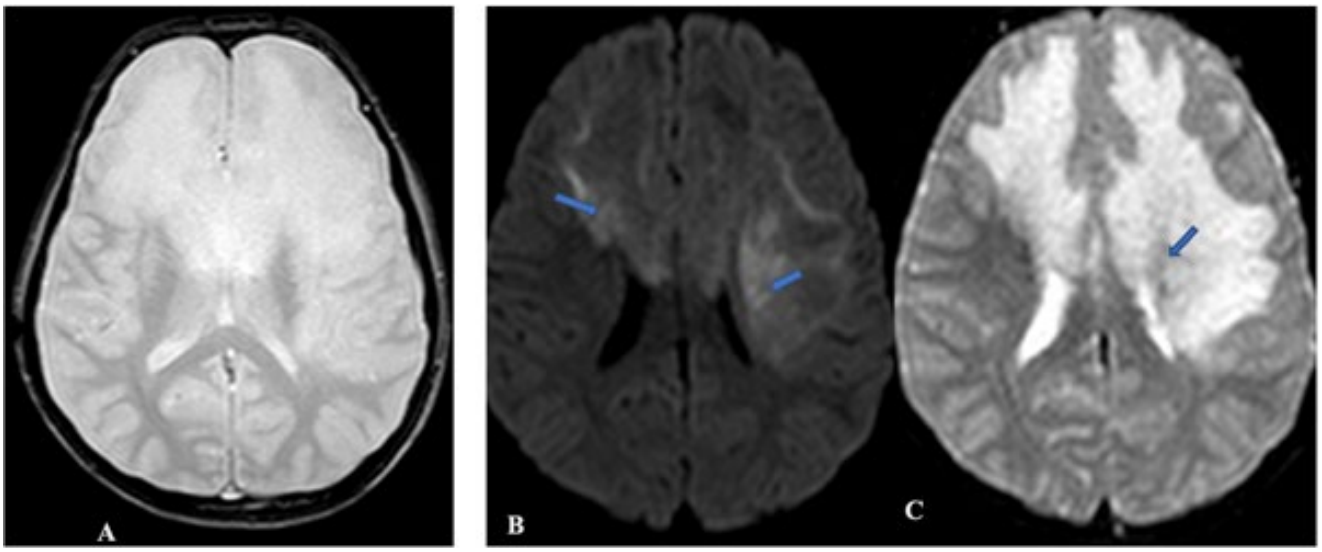

Figure 2: MRI images A-Axial GRE image shows no areas of blooming artifact in the lesion. (B-DWI and C-ADC)-The lesion shows diffusion restriction in its peripheral aspect (arrow) showing high signal intensity on DWI and low signal intensity on ADC mapping.

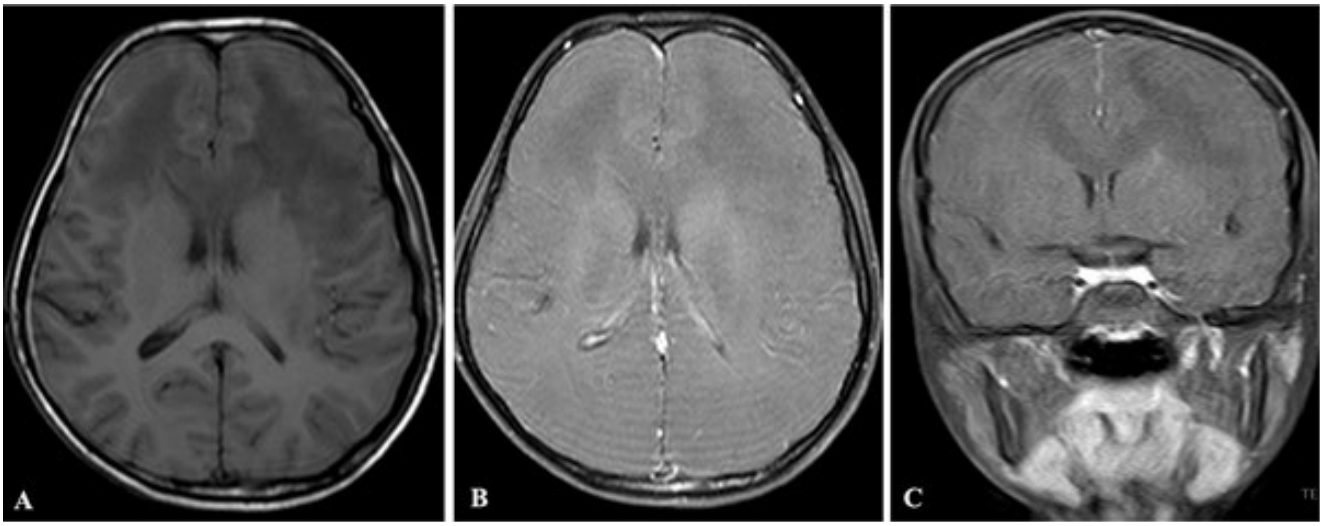

Figure 3: MRI images (A-Axial non-contrast T1 weighted image, B-Axial post contrast T1 weighted image, and C-Coronal post-contrast image) showing no enhancement in the lesion.

4. Magnetic Resonance Spectroscopy (Mrs.) Findings

Voxel taken from the lesion showed elevated Cho/creatine (1.34) and Cho/NAA (2.20) ratios. There was also decreased in NAA concentration (Fig 4).

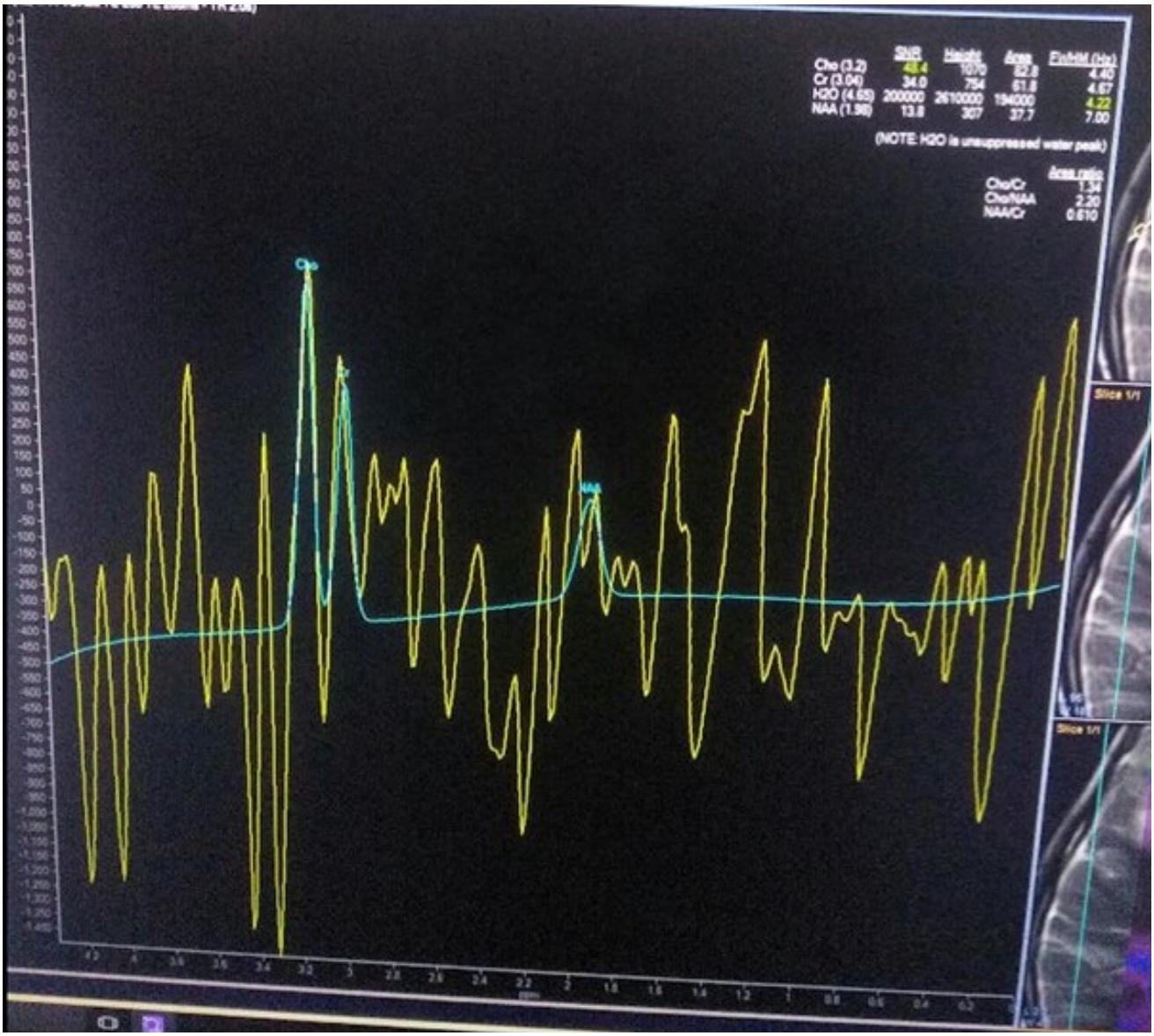

Figure 4: MRS findings taken from voxel containing the lesion demonstrating increased Cho/creatine (1.34) and Cho/NAA (2.20) ratios.

5. Histopathology

Biopsy of the lesion demonstrated an increase in cellularity with diffuse tumor cells in a fibrillary background. The tumor cells showed mild pleomorphism, indistinct cytoplasmic borders and moderate amount of eosinophilic cytoplasm, with their nuclei being round to oval and showing mild hyperchromasia (Fig 5), leading to a histopathologic diagnosis of diffuse astrocytoma (WHO grade II).

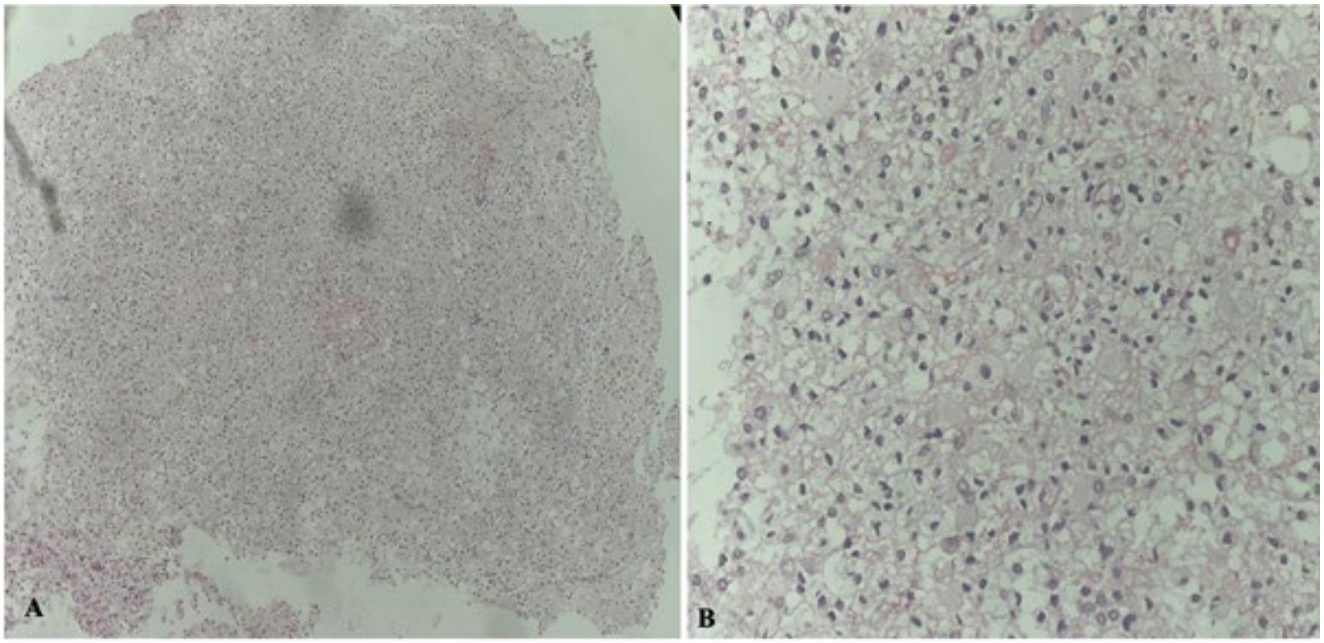

Figure 5: Biopsy of the lesion on Hematoxylin and Eosin (H&E) staining demonstrates increased cellularity with diffusely arranged tumor cells in a fibrillary background. Mildly pleomorphic tumor cells have moderate amount of eosinophilic cytoplasm and indistinct cytoplasmic borders, suggestive of diffuse astrocytoma (WHO grade II).

6. Discussion

Gliomatosis Cerebri is often an aggressive neoplasm, and the majority of cases show substantial bilateral infiltration involving at least three cortical lobel. It is rare and difficult to diagnose in children. It is nearly always fatal with survival length spanning from 6 months to 3 years after initial presentation.

The clinical signs of gliomatosis are non-specific and fairly mild in comparison to the lesion's magnitude. Computed Tomography (CT) results are either normal or exhibit sparsely distributed areas of hypoattenuation or so, MRI is the imaging modality of choice for diagnosis [8]. The lesion shows either isointensity or slightly hypointensity on T1-weighted imaging and shows high signal intensity on T2/FLAIR sequence. The lesion shows minimal or no enhancement on contrast study and no or minimal diffusion restriction on DWI. MR spectroscopy shows increased Choline/Creatine (Cho/Cr) and Choline/N-Acetyl Aspartate (Cho/NAA) ratios. An increase in these ratios is thought to be a result of increased membrane turnover in tumors, causing a decrease in N-Acetyl Aspartate (which reflects the replacement of neurons with neoplastic glial cells) and an increase in Cholin.

Diagnoses of GC can now be made much earlier with the help of MRI and stereo-tactic biopsy, which when combined with modern radiation and chemotherapy techniques, results in an increase in survival. Pathological findings indicate tumoral proliferation of astrocytes, oligodendrocytes, or both, with varying degrees of maturation which helps to differentiate GC from other non-tumoral pathologies [9].

GC is frequently misdiagnosed either as encephalitides, demyelinating diseases, or leukodystrophies. Acute Disseminated Encephalomyelitis (ADEM) is a well-known misdiagnosis in the pediatric population [10].

Both GC and ADEM can cause headaches, vomiting, and seizures, along with other site specific neurologic symptoms. Unlike GC, ADEM may be preceded by a viral illness or vaccination and may be accompanied by fever. MRI is the preferred imaging modality for GC; however, both GC and ADEM appear as hyperintense lesions on T2 weighted and FLAIR images with variable contrast enhancement, making imaging studies rather unspecific for the diagnosis [11].

It was difficult to diagnose this case as either GC or ADEM based on clinical or MRI findings alone and thus, a tissue biopsy was done. The biopsy showed diffuse astrocytoma (WHO grade II), and a final diagnosis of GC was made. Hence, GC should always be considered as a differential in children presenting with an encephalitic presentation, and though not initially recommended, one should not hesitate for proceeding to tissue biopsy in cases of diagnostic dilemma.

Treatment modalities for GC and ADEM are different. most patients with ADEM respond to intravenous corticosteroids. Alternative treatment modalities like plasmapheresis and Intravenous Immunoglobulin (IVIg) are also available if corticosteroids fail. On the other hand, GC is treated by radiotherapy and adjuvant chemotherapy. At the time of writing, the patient is being planned for radiotherapy and chemotherapy. Owing to these therapeutic and prognostic differences among these two, a correct diagnosis is crucial.

7. Conclusion

Gliomatosis Cerebri is rare in children, has misleading clinical picture, and has a dismal prognosis. Hence, it is important to consider it as a differential diagnosis in children with diffuse neurological symptoms and evidence of widespread, infiltrative lesions on MR imaging. A definitive diagnosis necessitates radiological-pathological correlation to differentiate from other nontumoral pathologies like acute disseminating encephalomyelitis. Considering biopsy early in cases of clinical quandary helps in early diagnosis and appropriate treatment.

Declaration

Ethical Approval and Consent to Participate

Not applicable

Consent for Publication

Written consent to publish this information was obtained from the parents.

Availability of Data and Material

The datasets used and/or analyzed during the current study are available from the corresponding author (Dr.Prakash Dhakal) on reasonable request.

Competing Interests

The authors of this manuscript declare that they have no competing interests.

Author's Contribution

Dr.Prakash Dhakal and Dr.Suraj Sharma- Selection of patient, making the diagnosis and image interpretation Dr.Nirmal Prasad Neupane-Provided good guidance for the case report from the beginning Dr.Seema Bhanadari-Prepared the manuscript

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Not Applicable

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Competing Interests

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