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Gliomatosis Cerebri: A Case Report

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Abstract

Background: Gliomatosis cerebri is a rare primary brain tumor that can have extensive infiltration into the brain parenchyma. It can affect any age group and often has poor clinical outcomes. Given its unique pathology, presentation and treatment can be challenging to recognize and efficaciously treat.

Keywords: Gliomatosis Cerebri, Hyperintensities, Toxic Leukoencephalopathy

Introduction

Gliomatosis cerebri is rare with age adjust incidence of 0.01 per 100,000 (95% CI 0.01-0.01) and an age adjust point prevalence of 0.08 per 100,000 (95% CI 0.07-0.09). The differential diagnosis is challenging to make due to its myriad presentations of disease including but not limited to: central venous sinus thrombosis, leptomeningeal gliomatosis, toxic leukoencephalopathy Our aim is to describe a rare presentation of disease and emphasize the importance of broad differentials with optimal pre-operative imaging and brain biopsy to guide appropriate timely oncologic intervention for patients.

Results (Case Report)

A 19-year-old healthy male presented to his local emergency department for rhinorrhea, congestion and body aches. He subsequently tested positive for COVID-19 and was counseled on appropriate quarantine protocol and discharged. He recovered but experienced a similar episode with additional headache and chest pain four months later, but then had a negative PCR test. No imaging had been obtained up to this point. Two months later, new symptoms occurred, headache, nausea/vomiting and body aches and he presented again to his local emergency department and was then transferred to our center. Preliminary labs and imaging were taken at the local hospital while urinalysis and remaining imaging was performed at our center [1].

Complete blood count with differential was drawn which revealed mildly elevated ANC (7.93 k/μL), red blood cells (6.15 k/μL) and hematocrit (51.9%). The complete metabolic panel showed elevated total proteins (8.4 g/dL), albumin (5.1 g/dL), and calcium (10.5 mg/dL). Blood lipase was low (11 U/L) and urinalysis demonstrated slightly cloudy urine with 1+ bilirubin, 3-5 red blood cells/HPF, trace hemoglobin and protein, and 2+ bacteria/HPF. A toxicology screen was negative for all illicit substances and metabolites. Acetaminophen and salicylate blood levels were below the reference ranges (<5 μg/mL, <1.0 mg/dL). Calcium ionized B yielded 1.03 mmol/L for ionized calcium and 1.07 mmol/L for normal calcium. Arterial blood gases revealed elevated readings such as the following: arterial pCO2 to be 50 mmHg, arterial bicarbonate 28 mmol/L and arterial CO2 content 29 mmol/L. Arterial pCO2 temperature corrected was the same as temperature non-corrected. Whole blood sodium was elevated at 146 mmol/L. Initial non-contrast computed tomography (CT) and gadolinium-enhanced magnetic resonance imaging (MRI) of his brain revealed diffuse cerebral edema with effacement of sulci (Figure 1). The edema was attributed to extensive vaping with a diagnosis of vaping induced toxic leukoencephalopathy and the patient was subsequently discharged [2].

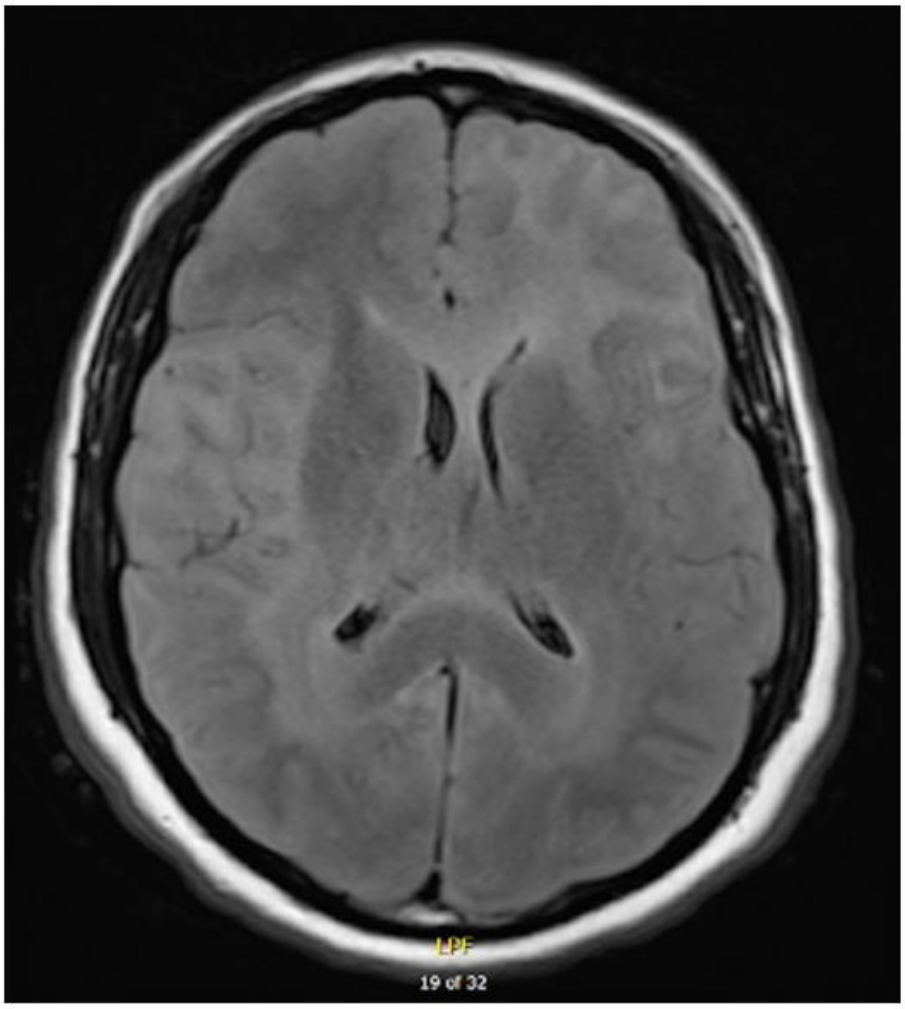

Figure 1: Initial magnetic resonance image (MRI) of the brain with and without contrast showed effacement of the sulci throughout the cerebral hemispheres with effacement of the basal cisterns and partial effacement of the ventricular system. There was loss of the gray-white mater junction in the thalami bilaterally. Initial magnetic resonance imaging with and without contrast showed diffuse cerebral edema with prominent mass effect, and diffusely attenuated intracranial arteries with no evidence of significant large vessel stenosis or focal stenosis or large vessel occlusion

One month after discharge, the patient presented to his local hospital with a seizure and altered mental status. His neurological exam was notable for diffuse motor weakness, lack of desire to participate in the exam, but could move his extremities on command, had poor ability to complete finger-to-nose testing due to lack of participation but had adequate extraocular movement following a finger. Of note was the presence of supra-ventricular cardiac arrhythmia on physical exam and telemetry. Speech was slow but not slurred with no facial droop. He was again transferred to our center where extensive consultation with medical genetics, cardiology and neurosurgery was completed [3]. The patient underwent EEG monitoring with periodic lateralized epileptiform discharges (PLEDs) observed but no seizure activity. Imaging was taken during this stay but did not reveal any new lesions. Upon further review of the aforementioned imaging, multifocal bi-hemispheric supratentorial hyperintensities were seen on T2 FLAIR MRI (Figure 2). Of note, was restricted diffusion on MRI seen with DWI/ADC and attenuation of CT in the right temporal lobe. Due to persistence of neurological deficits, lack of confirmatory testing from consults, and unknown etiology for symptoms, a brain biopsy by neurosurgery was performed. Pathology from frozen section yielded a high-grade glioma with diagnosis of gliomatosis cerebri. The patient was referred to radiation oncology and follow up of treatment planning was assumed by their team (Figure 3) [4].

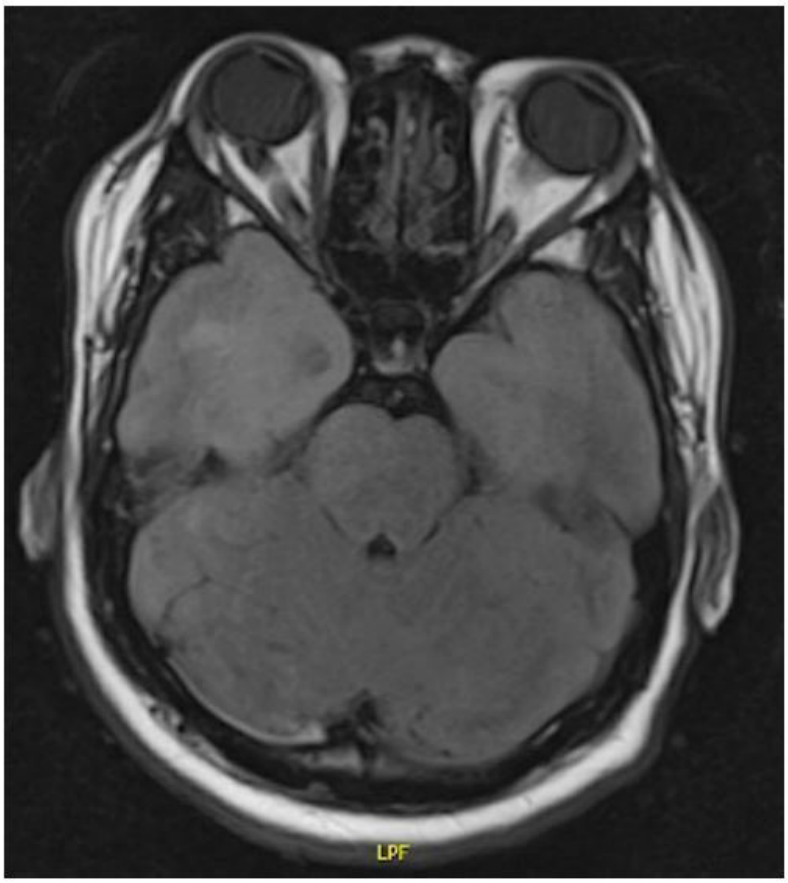

Figure 2: Repeat magnetic resonance imaging (MRI) — (Late Fall) showed multifocal bi-hemispheric supratentorial confluence cortical/ subcortical mass-like T2-FLAIR intensity, and hyperintensity of the brainstem

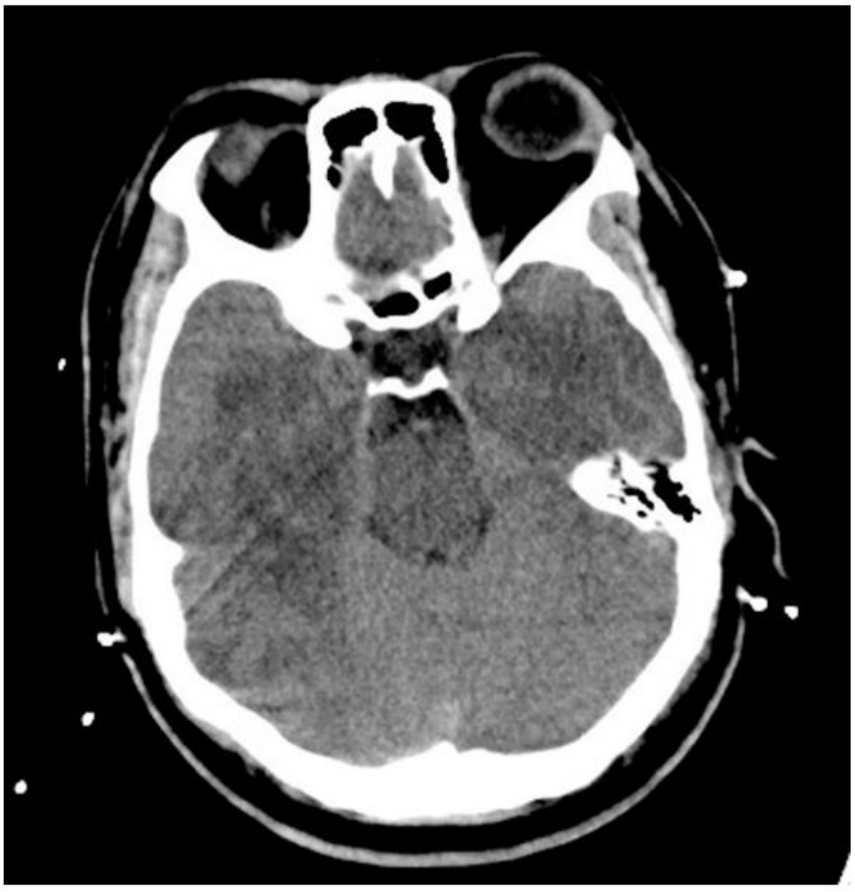

Figure 3: Computed tomography (CT) without IV contrast after previous MRI without contrast. Given the stability of findings in imaging and clinically over the time course, this raises concern for gliomatosis cerebri. Small focal enhancement and restricted diffusion on MRI and increased attenuation on CT in the right temporal lobe would suggest a more aggressive component of the mass

Conclusion

The natural progression of gliomatosis cerebri is bleak with 26-52% of patients surviving past a year from initial symptom onset. Since its initial recognition in the early 1900's, little advancement has been made. As a result of the tumor's heterogeneity, effective treatment has been limited due to its extensive brain tissue involvement. Given its rarity, diagnosis of this is often missed or mis-identified as other pathogenies (as in this case) or identified as idiopathic in nature. In our case, gliomatosis cerebri was under the guise of leukoencephalopathy. The medical community as a whole and clinicians alike should be aware of this disease as a possible diagnosis in cases of atypical neurological symptoms in the absence of clear pathologic mass lesions and in the setting where diffuse vasogenic edema is occluding adequate visualization of neurological structures.

Informed Consent

The patient provided written consent and permission to use patient health information for this publication.

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