Case Report
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Gyratory Seizures in Hypothalamic Hamartoma

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Gyratory seizures (GS) are a rare seizure type characterized by body rotation of ≥180° around its vertical axis. While GS have been documented in various epileptic syndromes, their occurrence in association with hypothalamic hamartomas (HH) has not been reported previously. This case report introduces the first documented instance of GS in a patient with a HH, a non-neoplastic tumor originating from the tuber cinereum. The patient, a 25-year-old female, with a history of recurrent seizures since childhood, initially presented with gelastic seizures, marked by inappropriate laughter, and subsequent evolution of symptoms including right oculocephalic version and gyratory seizures to the right side. Despite multiple antiepileptic medications, seizures persisted. Neuroimaging revealed a HH in the right hypothalamic region. The presence of polydactyly prompted consideration of Pallister Hall syndrome (PHS). PHS is an autosomal dominant condition linked to GLI3 gene mutations. While some features of PHS were absent in this case, the presence of both gelastic and gyratory seizures indicated the hypothalamus as the lesion site, despite inconclusive electroencephalogram findings. This report underscores the novel association of GS with HH and highlights the importance of considering PHS in patients with HH and polydactyly presenting with gelastic and gyratory seizures. Understanding GS in HH may offer insights into broader hypothalamic lesion-related epileptic phenomena. **(2024;14:47-49)**

Key words: Seizures, Hypothalamic hamartoma, Pallister hall syndrome

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Introduction

Gyratory seizures (GS) are defined as seizures causing the body to rotate around its vertical axis by more than or equal to 180 degrees.[1] These seizures have been described in primary generalized as well as focal epilepsies.[2,3] Symptomatic focal epilepsies with GS can be due to structural pathology involving the frontal, parietal, or thalamic regions. However, GS in hypothalamic hamartomas have never been reported previously in the literature. We describe the case of a young woman who had a hypothalamic hamartoma (HH) and manifested both the characteristic gelastic as well as the phenotype of gyratory seizures.

Case Report

A 25-year-old female, born to a non-consanguineously married couple with normal growth and development, presented with recurrent seizures since the age of 11 years. She had bouts of inappropriate laughter to start with, which were not associated with impaired awareness or any involuntary movements. There was no aura or postictal confusion. She had poor recollection of the events. Her symptoms were poorly controlled and progressed over time, as evidenced by increased frequency, prolonged duration, and a change in the semiology of attacks. After a few years, she had seizures with a right oculocephalic version, followed by circular movements of her trunk. Seizures occurred two to three times a week and lasted around 3 minutes. All her seizures were characterized by gyrations to the right side. She continued to have seizures despite taking four antiseizure medications. She was found to have polydactyly during the physical examination. Neurological examination was within normal limits. Interictal electroencephalogram (EEG) showed multifocal interictal discharges with complex dipoles and generalized epileptiform abnormalities with generalized paroxysmal fast activity. Ictal EEG had no localizing or lateralizing features. Her recent brain magnetic resonance imaging showed a hamartoma in the right hypothalamic region (Fig. 1). The diagnosis of Pallister Hall syndrome (PHS) was also entertained in the setting of HH with polydactyly. Dysplastic nails, bifid epiglottis, imperforate anus, renal anomalies,

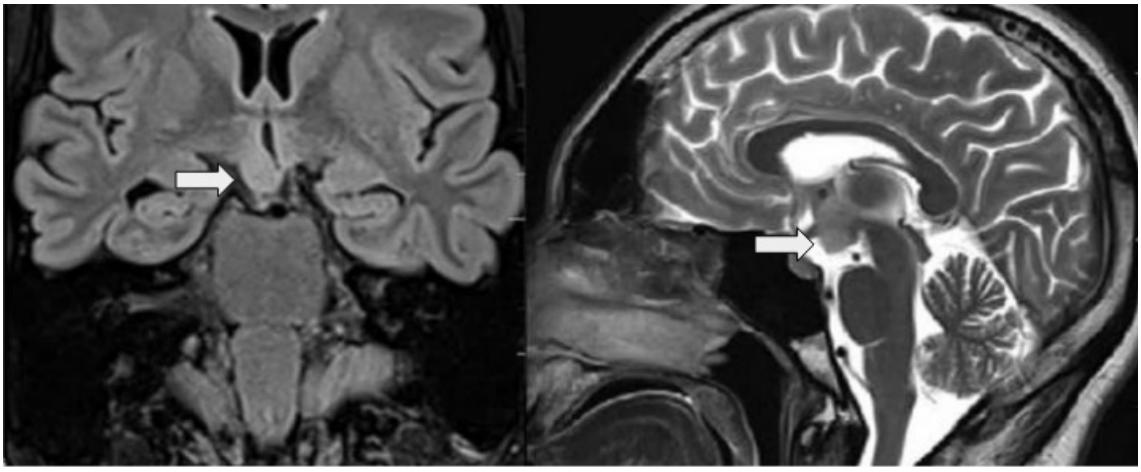

Figure 1. Coronal (FLAIR) and sagittal (T2W) sections of brain showing right hypothalamic hamartoma (marked in arrows). FLAIR, fluid-attenuated inversion recovery; T2W, T2 weighted.

pituitary dysplasia, and hypopituitarism are the other features seen in PHS that were not observed in our case.[4]

Discussion

Recurrent or drug-refractory epilepsy is the most common presentation of HH.[5] HH are non-neoplastic tumors arising from the tuber cinereum. Gelastic seizures characterized by inappropriate bouts of laughter are the hallmarks of HH.[5]

GS are typically seen in focal epilepsies and have also been reported in cases of generalized epilepsy like juvenile myoclonic epilepsy.[6] In a study by Dobesberger et al,[7] out of 277 patients who underwent video EEG monitoring, 12 patients manifested GS. In their series, the origin of seizures was either frontal or temporal.[7] Certain uncommon etiologies have been found to be associated with GS, like temporal encephalocele and N-methyl-D-aspartate receptor encephalitis.[8,9] A case report from the early nineties described a 66-year-old woman who turned a complete 360° with the ease of a ballet dancer before she fell and appeared fearful. She was later discovered to be harbouring a thalamic neoplasm, a biopsy of which revealed a differentiated oligodendroglioma.[10] Previous pre-clinical studies have demonstrated rotational behaviour to be evoked by electrical stimulation of deep thalamic nuclei.[11]

This is the first report of GS in HH. The phenomenon could be explained by the propagation of ictal activity to the thalamus and subcortical motor pathways, or to the frontotemporal regions. The propagation to other regions of the brain, such as the frontal eye fields or supplementary motor cortices, resulted in contralateral deviation of eyes and head, and sometimes the whole body in cases of prolonged cortical activation. In the study by Dobesberger et al,[7] the authors concluded that lateralization of seizures was contralateral to the direction of gyration when it was preceded by an oculocephalic version and ipsilateral when it was not. The occurrence of gelastic seizures along with GS with a demonstrated lesion in the hypothalamus, led us to localise the hypothalmus as the focus, despite the fact that EEG was ineffective in localisation.

The presence of polydactyly prompted us to consider PHS.[12] PHS is an inherited autosomal dominant disease that occurs due to pathogenic mutations in the GLI3 gene. About 25% of patients with PHS can have *de novo* mutations.[13] So, the absence of family history does not preclude the diagnosis in our case.

Our case highlights the unique presentation of GS in HH. In patients presenting with gelastic and GS, the lesion can be localized to the hypothalamic region. PHS should be considered in all patients with HH with polydactyly.

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