Available online at www.sciencerepository.org

Science Repository

Case Report

Atypical Localization and Clinical Presentation of a Hypothalamic Hamartoma in an Adult with Epilepsy

Marone Abril M[1*], Sanchez Melanie[2], Osa Sanz Emilia[3], Yañez Paulina[3] and D´Giano Carlos H[1]

[1]*Comprehensive Epilepsy Center, Fleni, Ciudad Autónoma de Buenos Aires, Argentina*
[2]*Department of Neurology, Fleni, Ciudad Autónoma de Buenos Aires, Argentina*
[3]*Department of Radiology, Fleni, Ciudad Autónoma de Buenos Aires, Argentina*

ARTICLE INFO

Article history:
Received: 17 January, 2022
Accepted: 29 January, 2022
Published: 21 February, 2022

Keywords:
Hypothalamic hamartoma
focal epilepsy
temporal lobe epilepsy
secondary epileptogenesis

ABSTRACT

Epilepsy in patients with Hypothalamic hamartomas (HH) have a typical semiology (gelastic or dacrystic seizures), but there is also secondary epileptogenesis that can explain why some of them have "pseudotemporal" seizures. We present a case report of an atypical localization and clinical presentation of HH in an adult with a rare presentation of secondary epileptogenesis.

Introduction

Hypothalamic hamartomas (HH) are non-neoplastic heterotopic tissues which contains normal neurons and glia including oligodendrocytes and fibrillary astrocytes but in an abnormal distribution. Even though they are rare and don't have a neoplastic evolution, they grow as fast as the tissue around them [1]. Munari *et al*., have already demonstrated with stereoEEG studies the role of the HH in generating seizure activity [2]. "Epileptic" HH are usually sessile and closely connected to the mammillary bodies [3]. The epilepsy syndrome noted in adult patients with HH appears to be different from the catastrophic epilepsy noted in children. The epilepsy syndrome is usually restricted to one or two focal seizure types, commonly focal onset with impaired consciousness ("pseudotemporal") seizures, or tonic seizures, with a variable expression of gelastic seizures (less frequent than in children), and occasional generalized seizures. Also, learning difficulties and behavioural problems are less frequent [4]. In this case report, we present a patient with focal onset and impaired consciousness seizures and a HH originating in the left mammillary body.

Case Report

A 45-year-old, right handed, male patient, with a history of thyroid dysfunction, initiates at the age of 36, with focal with impaired consciousness seizures characterized as unresponsive period with oral and bilateral hand automatisms, especially during sleep. His seizure frequency was variable, from daily to weekly. Interictal EEG showed left temporal anterior spikes and sharp waves. Ictal EEG showed late left anterior temporal slow activity in alfa/theta range (Figure 1). In the magnetic resonance imaging (MRI) we observed a sub-centimetric nodular expansion of the left mamillary body, with isointense signal in respect to the contralateral, suggestive of hamartoma. It was isointense to the gray matter on T1- and T2-weighted (W) imaging and showed no enhancement on post gadolinium T1W series (Figure 2). DTI sequences revealed an asymmetric anisotropy fraction in both temporal lobes, being smaller in the left side. The patient did not develop cognitive dysfunction, behavioural problems or gelastic seizures. He received medical treatment with LEV, BRV, VPA, LCM without clinical response.

[*]*Correspondence to: Marone Abril M, Comprehensive Epilepsy Center, Fleni, Ciudad Autónoma de Buenos Aires, Montañeses 2325, C1428AQK, Argentina; E-mail: mlmarone@fleni.org.ar*

http://dx.doi.org/10.31487/j.SCR.2022.01.06

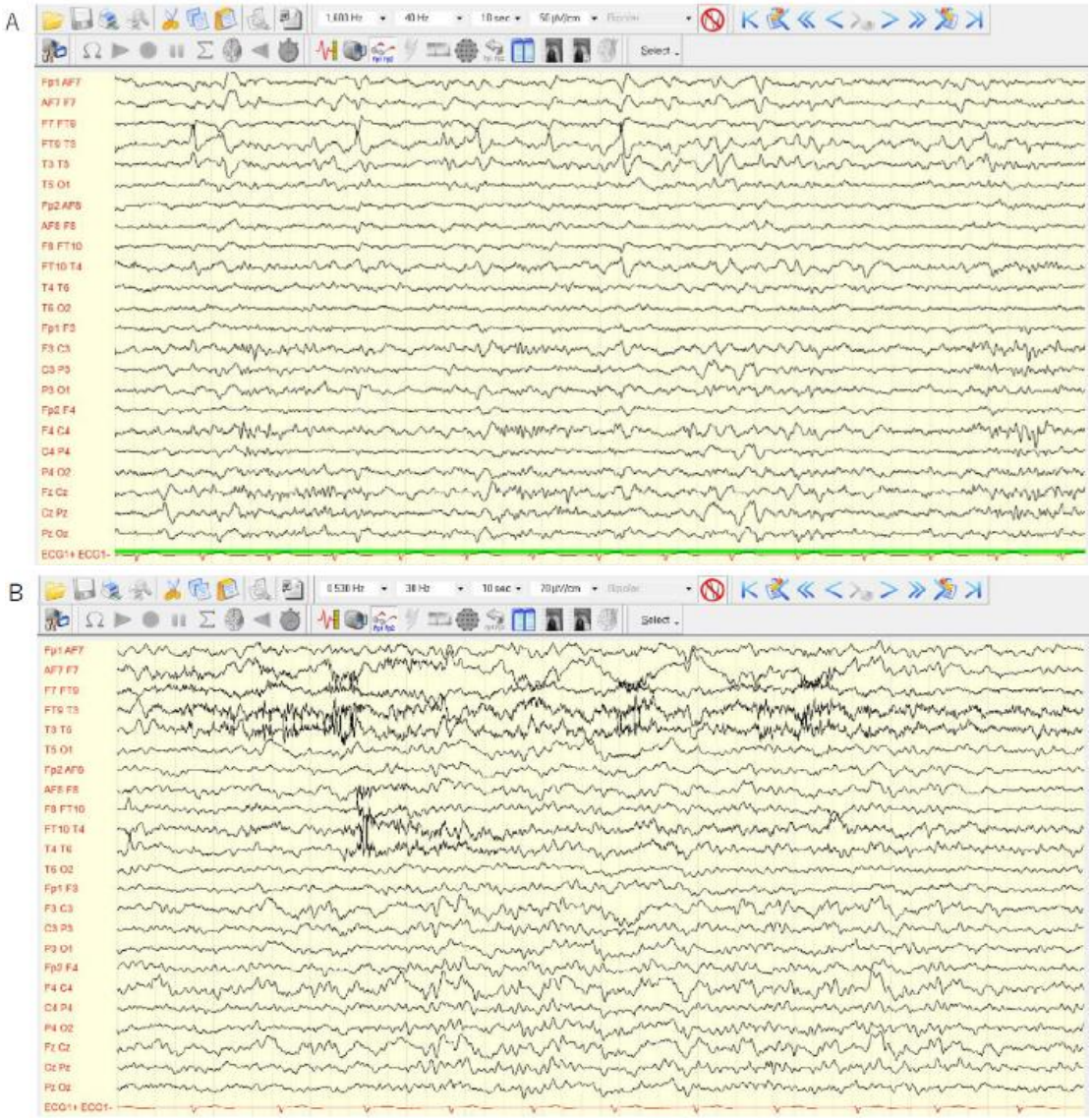

Figure 1: A) Interictal EEG: Left anterior temporal spikes and sharp waves. **B)** Ictal EEG: Alfa/theta range slowing in left anterior temporal area.

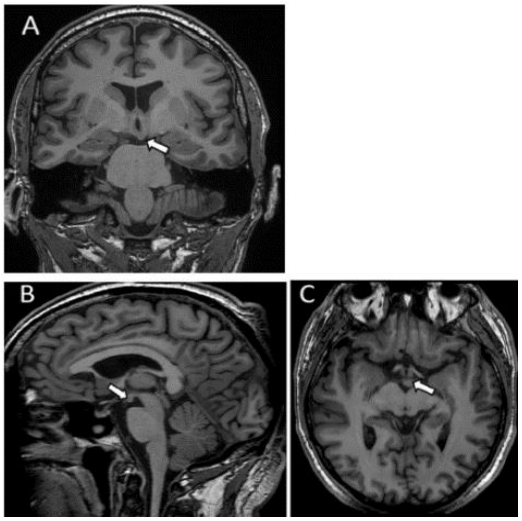

Figure 2: A) Coronal; **B)** Sagital; **C)** Axial three-Dimensional T1-weighted images fast spoiled gradient echo (FSPGR) (TR/TE= 9/3) shows a sub-centimetric nodular expansion of the left mamillary body, with isointense signal with respect to the contralateral, suggestive of hamartoma (arrows in A, B and C).

Discussion

Mammillary tract is an unusual localization of HH. Although there is evidence of the intrinsic epileptogenicity of the HH´s, we usually see typical clinical presentations with gelastic or dacrystic seizures, in the majority of these patients [2]. In a series of 5 patients, Kahane *et al.* studied with stereoEEG HH´s with refractory epilepsy and he proposed whether 4 of them had interictal and ictal discharges coming from HH´s, there was another type of seizures in which the HH wasn't involved. So, he proposed a theory of secondary epileptogenesis in which through the mammillary tract -Thalamus-cingulate gyrus, it can reach the cortex [5]. But these kinds of seizures were late in the evolution of epilepsy and they were happening at the same time as gelastic seizures. So, in our case we present a patient who only have clinically pseudo temporal lobe seizures and in EEG we can see interictal discharges in left temporal lobe and an ictal EEG which does not show a clear localization but we can see some late slow waves in the left hemisphere. As Kahane *et al.* described, the secondary epileptogenesis, if present, tends to be on the same side of the HH. So, we think this is an atypical presentation of an HH with secondary epileptogenesis.

Conflicts of Interest

None.

REFERENCES

1. Alomari SO, El Houshiemy MN, Bsat S, Moussalem CK, Allouh M et al. (2020) Hypothalamic hamartomas: A comprehensive review of the literature - Part 1: Neurobiological features, clinical presentations and advancements in diagnostic tools. *Clin Neurol Neurosurg* 197: 106076. [Crossref]
2. Munari C, Kahane P, Francione S, Hoffmann D, Tassi L et al. (1995) Role of the hypothalamic hamartoma in the genesis of gelastic fits (a video-stereo-EEG study). *Electroencephalogr Clin Neurophysiol* 95: 154-160. [Crossref]
3. Régis J, Scavarda D, Tamura M, Nagayi M, Villeneuve N et al. (2006) Epilepsy related to hypothalamic hamartomas: surgical management with special reference to gamma knife surgery. *Child's Nerv Syst* 22: 881-895. [Crossref]
4. Mullatti N (2003) Hypothalamic hamartoma in adults. *Epileptic Disord* 5: 201-204. [Crossref]
5. Kahane P, Ryvlin P, Hoffmann D, Minotti L, Benabid AL (2003) From hypothalamic hamartoma to cortex: what can be learnt from depth recordings and stimulation? *Epileptic Disord* 5: 205-217. [Crossref]