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Anatomical, radiological and clinical features of mesial temporal lobe epilepsy: two illustrated cases reports

Aspectos anatômicos, radiológicos e clínicos da epilepsia do lobo temporal mesial: relato ilustrado de dois casos

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ABSTRACT

Mesial temporal lobe epilepsy is the most commom form of focal epilepsy in adults. Its clinical features include focal seizure, dysmnestic symptoms – such as déjà vu or jamais vu – and autonomic or psychic aura. We reported two cases of mesial temporal lobe epilepsy with similar clinical features, but with entirely different etiologies. Mesial temporal sclerosis contributes up to 70% of all mesial temporal lobe epilepsy cases and MRI usually shows reduced hippocampal volume and increased signal intensity on T2-weighted imaging. Incomplete hippocampal inversion has uncertain relation with epilepsy and is characterized by an atypical verticalized and medially positioned anatomical pattern of the hippocampus and also a deep collateral sulcus.

Keywords: Temporal lobe, epilepsy, hippocampus, seizures

RESUMO

A epilepsia do lobo temporal mesial é a forma mais comum de epilepsia focal em adultos. Suas características clínicas incluem crises focais, sintomas dismnésicos - como déjà vu ou jamais vu - e aura autonômica ou psíquica. Relatamos dois casos de pacientes com epilepsia do lobo temporal mesial com manifestações clínicas semelhantes, mas com etiologias completamente diferentes. A esclerose mesial temporal contribui com até 70% de todos os casos de epilepsia do lobo temporal mesial e, geralmente, na ressonância magnética, apresenta atrofia do hipocampo e hipersinal na imagem ponderada em T2. A rotação incompleta do hipocampo possui uma relação incerta com a epilepsia e é caracterizada por alteração da estrutura interna do hipocampo, com um sulco colateral verticalizado e profundo.

Palavras-chave: Lobo Temporal, Epilepsia, Hipocampo, Convulsões

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INTRODUCTION

Temporal lobe has functions related to memory, language, emotions, and hearing[1]. A dorsolateral view of the brain shows two main sulci: superior temporal sulcus and inferior temporal sulcus, which limits three gyri: superior temporal gyrus, middle temporal gyrus, and the inferior temporal gyrus[1,2] (FIGURE 1). In a didactic manner, temporal lobe is divided in two regions: lateral temporal lobe and mesial temporal lobe (mTL). The mTL is divided into the following: parahippocampal gyrus, uncus, hippocampus, fimbria, amigdala, and dentate gyrus[2] (FIGURE 1).

Mesial temporal lobe epilepsy (mTLE) represents up to 21% among focal epilepsies, and epileptic focus of mTLE is mostly located in the mTL[3,4](TABLE 1). Epileptic seizures in mTLE present variable clinical features. Auras in mTLE are sensory or psychic phenomena and can manifest itself in isolation, but it can also be followed by motor manifestations, for example[5,6]. Furthermore, auras occur in 20% to 90% of patients with mTLE and can presents with abdominal discomfort, chest tightness, palpitation, fear, anxiety, déjà-vu or jamais vu, and hallucinations (usually auditory or visual). Gustatory and olfactory hallucinations may happen, but are less common[6,7] (TABLE1).

Some of the most common symptoms of focal epileptic due to mTLE are eye staring, unilateral dystonic posturing, impaired consciousness, rising epigastric sensation, oral automatisms (chewing and sucking) and unilateral limb automatisms. Autonomic signs such as pupillary dilatation, hyperventilation, piloerection and tachycardia can also occur. Subsequent amnesia, dysmnestic symptoms including déjà vu and jamais vu are other typical signs[5] (TABLE 1).

Treatment of mTLE includes antiepileptic drugs (AED), notwithstanding several cases can be considered as refractory epilepsy. Patients present refractory epilepsy when seizures do not respond to pharmacological treatment, even with appropriate trials of two AED, either alone or in combination. In these cases, a surgical approach is required, which presents high rates of success[8].

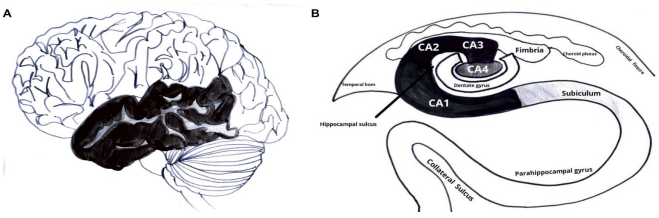

Figure 1: (A) Representation of the lateral view of telencephalon, showing the temporal lobe and its anatomical limits. Highlighted main temporal gyri: superior temporal gyrus, middle temporal gyrus, and the inferior temporal gyrus. (B) Graphical representation of hippocampal anatomy.

CASE REPORT

Find below two case reports describing patients with similar clinical features due to completely different etiologies.

CASE 1: A 49-year-old woman with an 8-year history of epileptic seizures characterized by oroalimentary and left upper limb automatisms, which last approximately 2 minutes and were followed by postictal amnesia. Seizures were often preceded by emotional aura (ictal fear) accompanied by palpitations. Patient presented a history of epilepsy in her childhood and simple febrile seizure. General and neurological exam were normal. Interictal electroencephalogram (EEG) revealed bilateral frontotemporal spikes. In addition, brain Magnetic Resonance Imaging (MRI) showed increased T2 and FLAIR signal in mesial temporal lobe, loss of internal architecture of hippocampus and reduced hippocampal volume, which are typical of mesial temporal sclerosis (FIGURE 2). Treatment with carbamazepine (600mg per day) was chosen, with a good outcome in the initial 2 years. After that, there was a gradual increase in seizure frequency. Then, the pharmacological treatment was changed for oxcarbazepine 1800mg per day associated with clobazam 20mg per day, achieving a satisfactory therapeutic response.

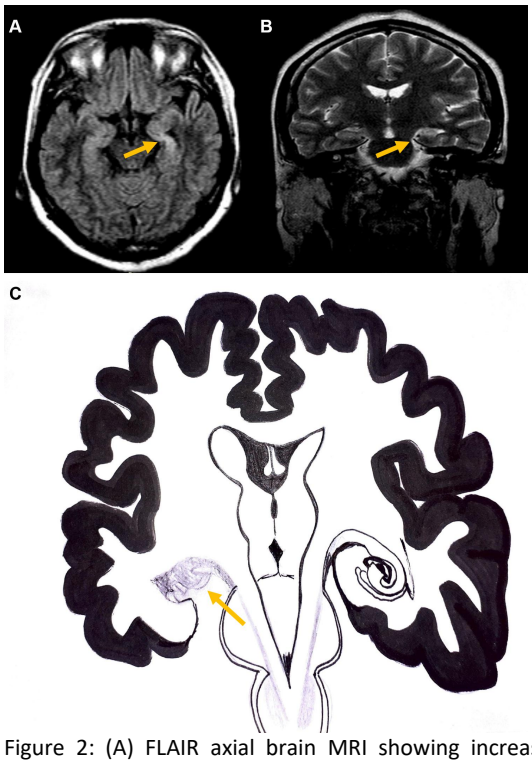

Figure 2: (A) FLAIR axial brain MRI showing increased signal in left mesial temporal lobe (arrow). (B) Coronal T2-weighted brain MRI showing loss of internal architecture of hippocampus (interdigitations), reduced hippocampal volume, and thin layer of white matter separating dentate nucleus and Ammon horn (arrow) which are typical of mesial temporal sclerosis. (C) Graphical representation in coronal section, showing altered signal intensity and loss of internal architecture of right hippocampus (interdigitations), typical findings of mesial temporal sclerosis (arrow).

CASE 2: A 49-year-old man with a 10-year history of epileptic seizures characterized by experiential aura (déjà-vu), followed by oroalimentary and left upper limb automatisms lasting less than 60 seconds, accompanied by postictal amnesia. He had an average of 1 seizure per 2 months, and he had sometimes experienced aura without a subsequent motor seizure. General and neurological exam were normal. EEG was normal, but brain MRI showed atypical anatomical pattern of the hippocampus with an abnormal medial location along the choroid fissure and a deep and verticalized collateral sulcus, supporting diagnosis of incomplete hippocampal inversion. (FIGURE 3). Patient was treated with carbamazepine 600mg/day with complete remission of seizures.

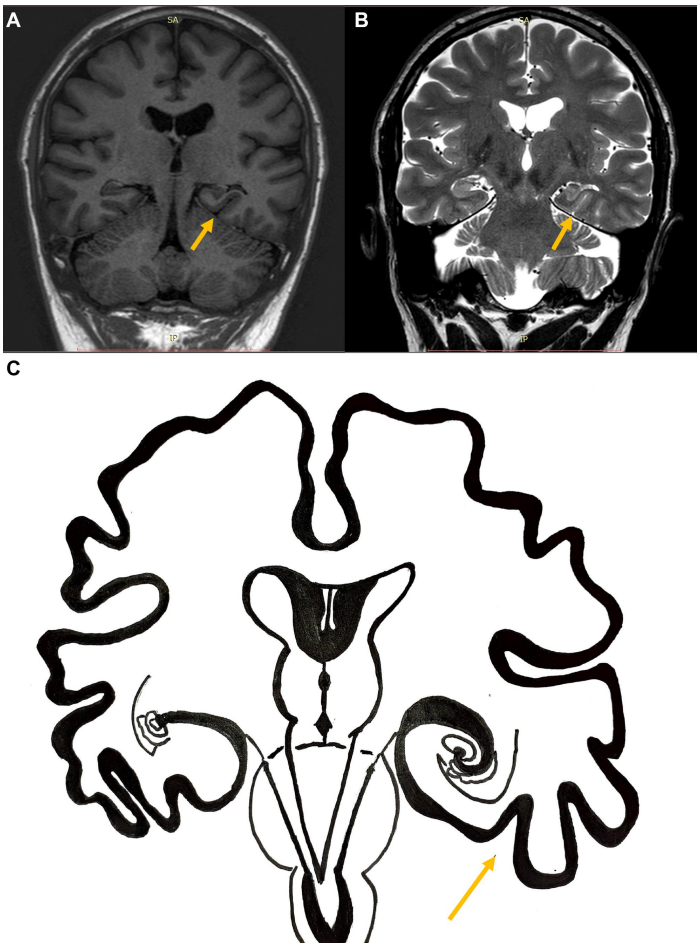

Figure 3: Coronal brain MRI showing atypical anatomical pattern of the hippocampus with an abnormal medial location along the choroid fissure and a deep and verticalized collateral sulcus (arrows), supporting diagnosis of incomplete hippocampal inversion (A) T1-weighted, (B) T2-weighted. (C) Graphical representation in coronal section, showing atypical anatomical pattern of the hippocampus with an abnormal medial location along the choroid fissure and a deep and verticalized collateral sulcus (arrow)

DISCUSSION

Mesial temporal sclerosis (MTS) is a neuroradiologic syndrome characterized by hippocampal sclerosis (HS), seizures and typical radiological findings; psychiatric symptoms may also occur[6]. MTS is the most common etiology of mTLE and it is present in 60-70% of patients with mTLE who have undergone surgery for treatment of drugs refractory seizures[9]. Histopathological features of MTS include neuronal loss, gliosis and granule cell dispersion. MRI typical findings includes: reduced hippocampal volume, increased signal intensity on T2-weighted imaging and loss of internal architecture of hippocampus[8].

Hippocampal inversion ends by the 21st week of embryological development. Incomplete hippocampal inversion (IHI) is also known as hippocampal malrotation, abnormal hippocampal formation and developmental changes of the hippocampus formation. IHI is most often unilateral and left-sided, characterized by an atypical round, verticalized and medially positioned anatomical pattern of the hippocampus. It is often accompanied by a deep and verticalized collateral sulcus[11-13].

Unlike MTS, the relation between IHI and epilepsy remains unclear. Some authors consider IHI a common finding in healthy subjects especially when it is unilateral[12,13]. Similarly, Bajic et.al. described IHI as common anatomical pattern, which does not represent a risk for development leading to epilepsy[14]. Also, authors suggested associations between IHI, occurrence of febrile status epilepticus and temporal lobe epilepsy[15]. In addition, other study demonstrated that IHI is associated with morphological changes outside the medial temporal lobe which can lead to epilepsy[11].

CONCLUSION

We report two cases of mTLE showing similar clinical manifestations, but entirely different etiologies. Therefore, we highlight that mTLE has a wide range of possible seizures, diverse etiologies and variable prognoses. We also point out the importance of recognizing the neuroimaging patterns of MTE and IHI.

The main cause of mTLE is mesial temporal sclerosis. However, the incomplete hippocampal inversion still has an uncertain relationship with epilepsy. Therefore, more studies are needed to validate/refute the hypothesis of the relation between epilepsy and IHI.

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Table 1: Summary table of mesial temporal sclerosis and incomplete hippocampal inversion.

Etiology	Mesial temporal sclerosis: A childhood history of febrile seizures, status epilepticus, encephalitis or trauma, generate spontaneous motor seizures leading to hippocampal sclerosis. Incomplete hippocampal inversion: It's a condition regarding to brain congenital malformations during embryological development.		
Incidence	Mesial temporal sclerosis: Mesial sclerosis is found in 50-70% of patients with mTLE. Incomplete hippocampal inversion: IHI is found in 30-50% of patients with mTLE and 9,37% among patients with intractable seizures.		
Clinical Features	Symptom/Signal		Clinical features
	Aura		
	Autonomic	Pupillary dilatation,	dilation, hyperventilation hyperventilation, piloerection and tachycardia
	Psychic experiences	Rising epigastric, panic	chest tightness, anxiety, fear,
	Ictal manifestations		
	Focal seizure	Eye staring, oral/upper limb automatisms,unilateral dystonic posturing, auditory and visuals hallucinations.	
	Dysmnestic symptoms	Amnesia, *Déjà vu* and *jamais vu*.	
Gender ratio	It does not seem to be a specific sexual predominance to Mesial temporal lobe epilepsy or incomplete hippocampal inversion.		
Age predilection	It does not seem to be a specific age predominance to Mesial temporal lobe epilepsy or incomplete hippocampal inversion.		
Risk factors	Mesial temporal sclerosis: A childhood history of febrile seizures. Other less important risk factors include head trauma, birth trauma, childhood central nervous system infection, and posterior cerebral artery territory infarcts. Incomplete hippocampal inversion: Genetic, environmental factors and presence of other malformations.		
Treatment	Mesial temporal sclerosis: Antiepileptic drugs such as carbamazepine, oxcarbazepine, levetiracetam, lamotrigine, and topiramate. In refractory epilepsy, surgical treatment may be indicated. Incomplete hippocampal inversion: Antiepileptic drugs such as carbamazepine, oxcarbazepine, levetiracetam, lamotrigine, and topiramate. In asymptomatic cases, drug treatment is not necessary.		
Prognosis	Mesial temporal sclerosis: Has a poor prognosis, considering its tendency of worsening epileptic seizures and a risk of becoming refractory epilepsy. Incomplete hippocampal inversion: Presents itself as an asymptomatic condition with a benign evolution. When epilepsy is present, it is usually easily controlled.		
Imaging findings	Mesial temporal sclerosis: Brain MRI shows increased T2 and FLAIR signal in mesial temporal lobe associated with loss of internal architecture and reduced hippocampal volume. Incomplete hippocampal inversion: The brain MRI shows abnormal medial location along the choroid fissure and a deep and verticalized collateral sulcus.		