

TEMPORAL LOBE HYPOGENESIS ASSOCIATED WITH ARACHNOID CYST IN PATIENTS WITH EPILEPSY

Eliane Kobayashi, Leonardo Bonilha, Li M. Li, Fernando Cendes

ABSTRACT - Objective: To determine the frequency of temporal lobe hypogenesis (TLH) associated with arachnoid cysts (AC) in patients with epilepsy. **Method:** We retrospectively revised 655 consecutive MRI scans from patients followed in our epilepsy clinic. We identified patients with temporal AC and then performed careful visual analysis in a workstation. Patients with evident expansive or destructive lesions were excluded. **Results:** Only 4 (0.6%) patients had AC in the left temporal lobe, all associated with TLH. In addition, there were also ipsilateral dysgenetic characteristics in the ipsilateral hippocampus including abnormal shape and axis, and hyperintense T2 signal. In one patient this hippocampal abnormality was bilateral. **Conclusion:** AC with TLH was rarely found in our patients with epilepsy and it was always associated with hippocampal dysgenesis. Although volumetric reduction of the temporal lobe can be observed in patients with epilepsy and hippocampal abnormalities, the presence of adjacent AC points to a malformative etiology.

KEY WORDS: hippocampal dysgenesis, cortical development malformation, partial epilepsy, seizures, magnetic resonance imaging.

Hipogenesia de lobo temporal associada a cisto aracnóide em pacientes com epilepsia

RESUMO - Objetivo: Determinar a frequência de hipogenesia de lobo temporal (HLT) associada a cistos de aracnóide (CA) em pacientes com epilepsia. **Método:** Revisamos retrospectivamente 655 exames consecutivos de ressonância magnética de pacientes do ambulatório de epilepsia da UNICAMP. Identificamos os pacientes com CA na região temporal e então realizamos análise visual em uma estação de trabalho. Pacientes com lesões expansivas ou destrutivas evidentes foram excluídos. **Resultados:** Apenas 4 (0,6%) pacientes tinham CA na região temporal esquerda, todos associados a HLT. Além destas alterações, observamos características disgenéticas no hipocampo ipsilateral incluindo anormalidades no formato e eixo e hipersinal em T2. Em um paciente estas alterações hipocampais eram bilaterais. **Conclusão:** CAs associados a HLT foram raramente encontrados, e quando presentes, estavam sempre acompanhados de disgenesia hipocampal. Apesar da redução volumétrica do lobo temporal poder ser observada em pacientes com epilepsia e alterações hipocampais, a presença de um CA adjacente indica uma etiologia de malformação.

PALAVRAS-CHAVE: disgenesia hipocampal, epilepsia parcial, convulsões, cistos de aracnóide, ressonância magnética.

Arachnoid cysts are considered frequent and incidental findings in neuroimaging studies¹. The most frequent localization is in the middle fossa, corresponding to more than half the cases reported in the literature². The origin of middle fossa arachnoid cysts is related to two theories: primary malformation of arachnoid (Starkman et al.)³ or secondary development due to partial agenesis of the temporal lobe (Robinson et al.)⁴. Magnetic Resonance Imaging (MRI) demonstrated the presence of morphologic abnormalities in the temporal lobe and adjacent bone remodeling, which indicate a secondary origin of arachnoid cysts¹. Temporal lobe agenesis/hypogene-

sis is believed to be an important etiology of middle fossa arachnoid cysts^{4,5}.

The temporal region is quite important in epileptology, since temporal lobe epilepsy (TLE) is not only the most frequent form of epilepsy in adults, but also the refractory epilepsy with the best post-operative results⁴. In TLE the main feature is the hippocampal atrophy (HA), which can be associated with atrophy of the amygdala and the temporal lobe⁵. In HA we observe reduced hippocampal volume, with abnormal internal structure, which causes an abnormally flat shape and altered internal signal (hyperintense T2 and hypointense T1 signal). Studies on rela-

Department of Neurology, Faculdade de Ciências Médicas (FCM), Campinas State University (UNICAMP), Campinas SP, Brazil. This study was supported by FAPESP, São Paulo, Brazil.

Received 26 September 2002. Accepted 5 December 2002.

Dr. Fernando Cendes - Departamento de Neurologia, FCM - UNICAMP - Caixa Postal 6111 - 13083-970 Campinas SP - Brasil. FAX: 55 19 32891818. E-mail: fcendes@unicamp.br

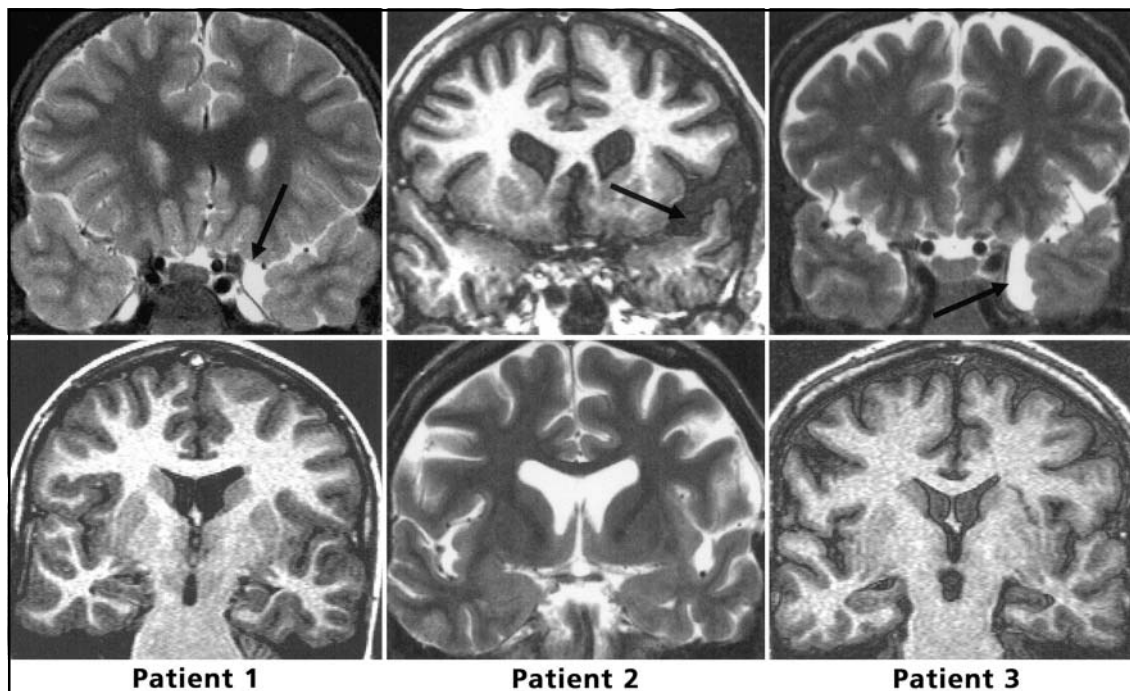


Fig 1. T1-IR and T2 weighted coronal images from patients 1, 2 and 3 with left temporal lobe hypogenesis associated with arachnoid cysts (arrows).

relationship between cysts and seizure foci, however, have proved negative^{6,7}.

The objective of this study is to determine the prevalence of temporal lobe hypogenesis associated with middle fossa arachnoid cysts in our patients with epilepsy.

METHOD

We retrospectively revised 655 consecutive MRIs from patients with epilepsy from UNICAMP Clinical Hospital. MRIs were acquired in a 2Tesla scanner (Elscent Prestige®), and kept in optical disk for revision in a workstation (OMNIPRO®). Exclusion criteria were evident destructive or expansive abnormalities identified in MRI.

Acquisition parameters were: (1) Sagittal T1 spin echo, 6mm thick, flip angle= 180°; repetition time (TR)=430, echo time (TE)=12, matrix 200X350, field of view (FOV)= 25X25cm; (2) Coronal images, perpendicular to long axis of hippocampus, defined on the sagittal images: (a) T2-

weighted and proton density "fast spin echo" (FSE), 3mm thick, flip angle= 160°; TR=4800, TE=108/18, matrix 256X256, FOV=22X22cm; (b) T1-weighted inversion recovery (IR), 3mm thick, flip angle=200°; TR=2800, TE=14, inversion time (TI)=840, matrix 130X256, FOV=16X18cm; (3) Axial images parallel to the long axis of the hippocampi: (a) T1-weighted gradient echo, 3mm thick, flip angle=70°, TR=200, TE=5, matrix 180X232, FOV=22X22 cm; (b) T2-weighted FSE, 4mm thick, flip angle=120°, TR=6800, TE=129, matrix 252X328, FOV=21X23cm; (4) T1-weighted 3D gradient echo with 1mm isotropic voxel, acquired in the sagittal plane for multiplanar reconstruction (1mm thick, flip angle=35°; TR=22, TE=9, matrix 256X220, FOV=23X25cm).

RESULTS

Only 4/655 (0.6%) from evaluated scans presented middle fossa arachnoid cysts, all in the left side and associated with signs of temporal lobe hypo-

Table 1. Data from the four patients with middle fossa arachnoid cysts.

Patient	Sex, Age (years)	Epilepsy syndrome	Outcome	EEG
1	M, 13	Simple febrile seizure	good	Left temporal slow waves
2	F, 38	Mesial TLE	poor	Bitemporal epileptiform discharges
3	M, 53	GTCs/sleep	good	Left temporal epileptiform discharges
4	F, 39	GTCs/sleep	good	Not available

TLE, temporal lobe epilepsy; GTCs/sleep, generalized tonic-clonic seizures during sleep.

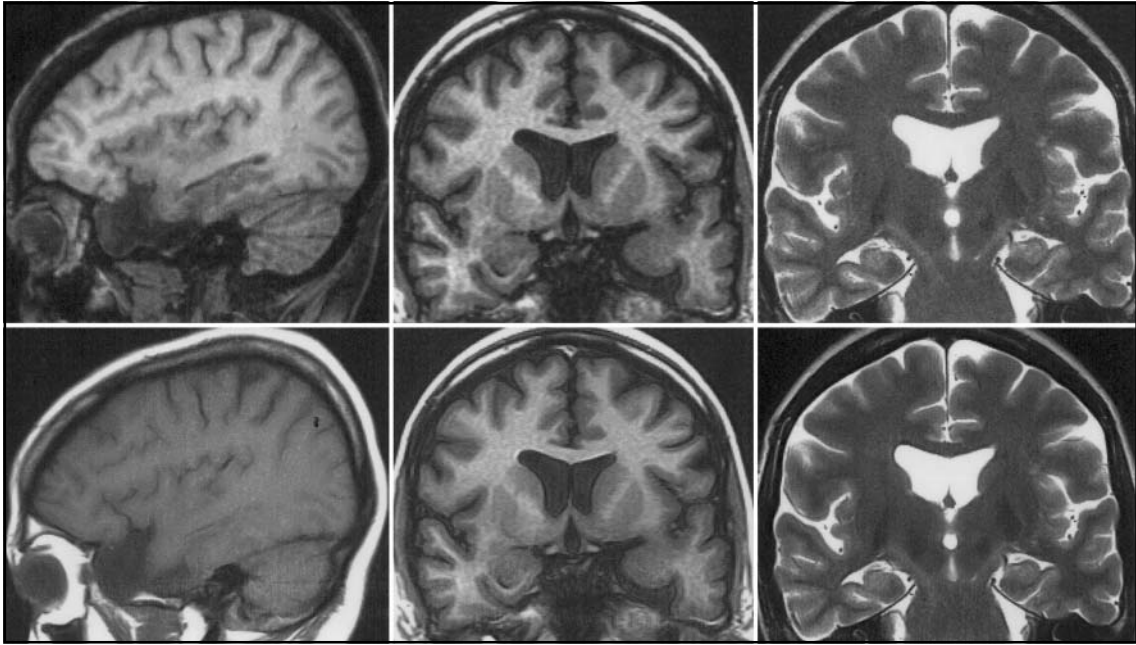


Fig 2. Images from patient 4 show left temporal lobe hypogenesis associated with arachnoid cyst, bilateral hippocampal abnormalities (rounded shape, vertical axis) and deep collateral sulcus.

genesis (Fig 1). A summary of patients characteristics is shown in Table 1.

Ipsilateral hippocampus had dysgenetic characteristics in all of them, with rounded shape, vertical axis and hyperintense T2 signal. In one patient this hippocampal abnormality was observed also in the contralateral hippocampus (Fig 2). The morphology of hippocampal abnormality found in these patients is quite different from that observed in hippocampal sclerosis.

DISCUSSION

Although temporal lobe abnormalities are frequently observed in patients with epilepsy, arachnoid cysts were rarely found among our patients. However, in the four patients with middle fossa arachnoid cysts, we found not only signs of temporal lobe hypogenesis, but also signs of hippocampal dysgenesis (bilateral in one of them).

Hippocampal dysgenesis is due to a malformation associated with abnormalities in the complex process of rotation and folding, which ends in abnormal shape, axis and internal structure^{8,9}. Such findings have been reported in asymptomatic individuals from families with either febrile convulsions or temporal lobe epilepsy^{10,11}. Recently, post-mortem pathological study of hippocampal malformation in an adult patient with temporal lobe epilepsy revealed abnormal position and complex convolutional malformations isolated to the hippocampal formation¹². All these evidences point to an inherited abnormality in the

mesial temporal region that is not necessarily associated with epilepsy.

Although temporal volume reduction can be found in patients with epilepsy and hippocampal abnormalities, the co-existence of arachnoid cysts indicates a malformative etiology. In the small number of patients with middle fossa arachnoid cysts identified in this study, the origin of the cyst can be attributed to temporal lobe hypogenesis.

REFERENCES

- Garcia Santos JM, Martinez-Lage J, Gilabert Ubeda A, Capel Aleman A, Climent Oltra V. Arachnoid cysts of the middle cranial fossa: a consideration of their origins based on imaging. *Neuroradiology* 1993;35:355-358.
- Wester K. Peculiarities of intracranial arachnoid cysts: location, sidedness, and sex distribution in 126 consecutive patients. *Neurosurgery* 1999;45:775-779.
- Starkman SP, Brown TC, Linell SA. Cerebral arachnoid cysts. *J Neuropathol Exp Neurol* 1958;17:484-500.
- Robinson RG. The temporal lobe agenesis syndrome. *Brain* 1964;88:87-106.
- Robertson SJ, Wolpert SM, Runge VM. MR imaging of middle cranial fossa arachnoid cysts: temporal lobe agenesis syndrome revisited. *Am J Neuroradiol* 1989;10:1007-1010.
- Cendes F, Andermann F, Gloor P, et al. MRI volumetric measurements of amygdala and hippocampus in temporal lobe epilepsy. *Neurology* 1993;43:719-725.
- Arroyo S, Santamaria J. What is the relationship between arachnoid cysts and seizure foci? *Epilepsia* 1997;38:1098-1102.
- Duvernoy HM. The human hippocampus: an atlas of applied anatomy. München: J. F. Bergmann Verlag, 1988.
- Baulac M, De Grissac N, Hasboun D et al. Hippocampal developmental changes in patients with partial epilepsy: magnetic resonance imaging and clinical aspects. *Ann Neurol* 1998;44:223-233.
- Fernandez G, Effenberger O, Vinz B et al. Hippocampal malformation as a cause of familial febrile convulsions and hippocampal sclerosis. *Neurology* 1998;50:909-916.
- Kobayashi E, Li M Li, Lopes-Cendes I, Cendes F. MRI evidence of hippocampal sclerosis in asymptomatic first degree relatives of patients with familial mesial temporal lobe epilepsy. *Arch Neurol* 2002 *in press*.
- Thom M, Sisodiya SM, Lin WR et al. Bilateral isolated hippocampal malformation in temporal lobe epilepsy. *Neurology* 2002;58:1683-1686.