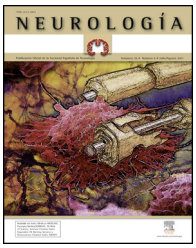
Neurología. 2015;30(2):90—96

NEUROLOGÍA

www.elsevier.es/neurologia



ORIGINAL ARTICLE

Corpora amylacea in the neocortex in patients with temporal lobe epilepsy and focal cortical dysplasia☆

B.O. Estupiñán-Díaz[a,*], L.M. Morales-Chacón[b], I. García-Maeso[c], L. Lorigados-Pedre[d], M. Báez-Martín[b], M.E. García-Navarro[e], O. Trápaga-Quincoses[b], N. Quintanal-Cordero[c], J. Prince-López[c], J.E. Bender-del Busto[f], Grupo Interdisciplinario de Cirugía de Epilepsia, Centro Internacional de Restauración Neurológica (CIREN)

[a] *Laboratorio de Anatomía Patológica, Centro Internacional de Restauración Neurológica (CIREN), La Habana, Cuba*
[b] *Servicio de Neurofisiología Clínica, Centro Internacional de Restauración Neurológica (CIREN), La Habana, Cuba*
[c] *Servicio de Neurocirugía, Centro Internacional de Restauración Neurológica (CIREN), La Habana, Cuba*
[d] *Laboratorio de Neuroinmunología, Centro Internacional de Restauración Neurológica (CIREN), La Habana, Cuba*
[e] *Departamento de Neuropsicología, Centro Internacional de Restauración Neurológica (CIREN), La Habana, Cuba*
[f] *Servicio de Neurología, Centro Internacional de Restauración Neurológica (CIREN), La Habana, Cuba*

Received 14 March 2013; accepted 1 June 2013
Available online 4 February 2015

KEYWORDS
Corpora amylacea;
Focal cortical dysplasia;
Temporal lobe epilepsy;
Clinical outcome;
Drug-resistant;
Histopathology

Abstract
Introduction: Corpora amylacea (CoA) are present in about 60% of atrophic hippocampi resected from patients with drug resistant temporal lobe epilepsy (DRTLE). They have also been described in the lateral temporal neocortex, although less frequently.
Objective: The objective is to measure the presence, distribution and density of CoA in the lateral temporal lobes of patients with DRTLE and focal cortical dysplasia (FCD), also examining how CoA density may be linked to demographic and clinical traits.
Methods: Resected tissue from 35 patients was analysed. CoA density was assessed with a semi-quantitative scale according to the criteria established by Cherian et al.
Results: Presence of CoA in the neocortex of nine patients was associated with hippocampal sclerosis (FCD type IIIa, seven cases), dysembryoplastic neuroepithelial tumour (FCD type IIIb, one case), and cavernous angioma (FCD type IIIC, 1 case). The meningeal surface (MS) was involved in all cases, and eight cases displayed CoA in the cerebral parenchyma (white matter) and around blood vessels. CoA density on the MS showed a negative correlation with age at seizure onset ($r = -0.828$, $P < .05$) and a positive correlation with disease duration ($r = 0.678$, $P < .05$) but not with postoperative clinical outcome.

☆ Please cite this article as: Estupiñán-Díaz BO, Morales-Chacón LM, García-Maeso I, Lorigados-Pedre L, Báez-Martín M, García-Navarro ME, et al. Cuerpos amiláceos en la neocorteza de pacientes con epilepsia del lóbulo temporal y displasia cortical focal. Neurología. 2015;30:90—6.

* Corresponding author.
E-mail address: baby@neuro.ciren.cu (B.O. Estupiñán-Díaz).

Conclusions: Patients with DRTLE and a primary lesion (hippocampal sclerosis, tumour, vascular malformation) associated with mild FCD were shown to have CoA deposits in the neocortex. No association was found between presence of CoA and clinical outcome 1 year after surgery.

PALABRAS CLAVE
Cuerpos amiláceos;
Displasia cortical focal;
Epilepsia del lóbulo temporal;
Evolución clínica;
Farmacorresistencia;
Histopatología

Cuerpos amiláceos en la neocorteza de pacientes con epilepsia del lóbulo temporal y displasia cortical focal

Resumen
Introducción: Los cuerpos amiláceos (CoA) se presentan en aproximadamente el 60% de los hipocampos atróficos resecados de pacientes con epilepsia del lóbulo temporal farmacorresistente (ELTFR). Su presencia en la neocorteza temporal lateral ha sido observada con menor frecuencia.
Objetivo: El objetivo es evaluar la presencia, la distribución y la densidad de CoA en el lóbulo temporal lateral de pacientes con ELTFR y displasia cortical focal (DCF) y la relación de su densidad con variables demográficas y clínicas.
Métodos: Analizamos histológicamente el tejido resecado de 35 pacientes con ELTFR. La densidad de los CoA fue evaluada con una escala semicuantitativa según los criterios de Cherian et al.
Resultados: La presencia de CoA en la neocorteza de 9 pacientes estuvo asociada a esclerosis hipocampal (DCF tipo IIIa, 7 casos), tumor neuroepitelial disembrioplásico (DCF tipo IIIb, un caso) y angioma cavernoso (DCF tipo IIIc, un caso). Todos los pacientes tuvieron afectación de la superficie meníngea (SM) y en 8 casos se localizaron en el parénquima cerebral (sustancia blanca) y alrededor de los vasos sanguíneos. La densidad de los CoA en SM tuvo una correlación negativa con la edad de inicio de las crisis ($r = -0{,}828$, $p < 0{,}05$) y positiva con la duración de la enfermedad ($r = 0{,}678$, $p < 0{,}05$) pero no con la evolución clínica postquirúrgica.
Conclusiones: En pacientes con ELTFR con lesión principal (EH, tumor, malformación vascular) asociada a DCF ligeras se constata la acumulación de CoA en la neocorteza. No se encontró una asociación entre la presencia de CoA y la evolución clínica al año de la cirugía.

Introduction

Surgery constitutes a valid treatment alternative in patients with drug-resistant temporal lobe epilepsy (DRTLE). Its most frequent neuropathological substrate is hippocampal sclerosis (HS), also known as mesial temporal sclerosis,[1] and it is detected by MRI in 87% to 89% of all patients.[2]

Robitaille et al. proposed the umbrella term 'polyglucosan bodies' (PB) to refer to Lafora bodies, Lafora-like bodies, Bielschowsky bodies, and corpora amylacea (CoA), since all of these structures show biochemical similarities with no histochemical or ultrastructural differences. PBs are rounded, amorphous, laminated, and basophilic. They range from 10 to 50 μm in diameter and they are composed of glucose polymers.[3]

PBs are considered non-specific structures.[3] When present in greater quantities, they are considered a pathognomonic symptom in several different diseases, and the specific term describing them will depend on the coexisting symptoms. For example, PBs are called Lafora bodies in Lafora disease,[4] Bielschowsky bodies in choreoathetosis and cerebral palsy,[5] and CoA in normal ageing and neurodegenerative diseases.[6]

Focal cortical dysplasia (FCD) is a specific subtype among the malformations of cortical development. It is highly epileptogenic and a frequent cause of drug-resistant epilepsy.[7]

CoA serve as a marker of HS when neuronal loss and gliosis are difficult to assess due to surgical resection of hippocampal tissue.[8]

CoA can be seen in the hippocampus and lateral neocortical tissue in DRTLE. However, there are few published studies describing them at either of these locations.[9–11]

In light of the above, our study aims to assess the presence, distribution, and density of CoA in the lateral temporal lobes of surgically treated patients with DRTLE and FCD, as well as to determine the connection between CoA density and demographic and clinical variables (age at seizure onset, epilepsy duration, and post-operative results).

Subjects and methods

We consulted the Centro Internacional de Restauración Neurológica (CIRN) database for patients with DRTLE who had undergone temporal lobectomy with electrocorticography and shown no response to any antiepileptic drugs (AED) for at least 2 years before the surgery. Of these surgically treated patients, we performed histological studies on those with FCD and CoA.