UNILATERAL MESIAL TEMPORAL ATROPHY AFTER A SYSTEMIC INSULT AS A POSSIBLE ETIOLOGY OF REFRACTORY TEMPORAL LOBE EPILEPSY

CASE REPORT

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ABSTRACT - Mesial temporal sclerosis is the main pathological substrate present in refractory temporal lobe epilepsy and its presence is often related to the occurrence of febrile seizures in infancy. There is an on-going discussion on the nature of mesial temporal sclerosis as it related to epilepsy: cause or consequence. A previously normal child developed hyperosmolar coma after abdominal surgery at the age of 6. Three months afterwards he developed simple and complex partial seizures with an increasing frequency and refractory to multiple mono- and polytherapeutic drug regimens. He was evaluated for surgery at the age of 13. Ictal and interictal recordings showed left temporal lobe abnormalities. Early CT scanning suggested left temporal atrophy. MRI showed mesial temporal sclerosis. Neuropsychological testing showed verbal memory deficits and he passed a left carotid artery amytal injection. He was submitted to a cortico-amygdalo-hippocampectomy and has been seizure-free since then. The clinical data obtained from this patient suggest that at least in this case mesial temporal sclerosis would be related to the cause of epilepsy and not resultant from repeated seizure activity.

KEY WORDS: epilepsy, mesial temporal sclerosis, systemic insult, febrile convulsions.

Esclerose mesial temporal unilateral após insulto sistêmico como possível etiologia de epilepsia refratária do lobo temporal: relato de caso

RESUMO - A esclerose mesial temporal é o principal substrato anatomo-patológico envolvido na epilepsia refratária do lobo temporal e está frequentemente associada à ocorrência de convulsões febris na infância. Persiste até o momento intensa discussão se a esclerose mesial seria causa ou consequência da síndrome epileptica. Uma criança previamente normal, desenvolveu coma hipersonsmolar após intercorrência em cirurgia abdominal aos 6 anos de idade. Após 3 meses iniciaram-se crises parciais simples e complexas em frequência ascendente e refratárias a múltiplos esquemas terapêuticos em monoe- e politerapia. Ele realizou investigação pré-operatória para epilepsia aos 13 anos de idade. Registros eletrencefalográficos de superfície ictais e interictais mostraram anormalidades temporais esquerda. TC de crânio antiga, próxima ao insulto sistêmico, já sugeria atrofia do lobo temporal. RMN demonstrou esclerose mesial temporal. Testagem neuropsicológica mostrou déficit de memória verbal e este paciente apresentou o padrão passa-ipsi/falha contralateralmente no teste de amital sódico. Ele foi submetido a córtico-amigdalo-hippocampectomia e está sem crises desde então. Os achados anatomo-patológicos são de esclerose mesial. Os dados obtidos neste paciente sugere que, ao menos neste caso, a esclerose mesial temporal está possivelmente relacionada à causa da epilepsia e não à presença de crises recorrentes.

PALAVRAS-CHAVE: epilepsy, esclerose mesial temporal, insulto sistêmico, convulsão febril.

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Patients with refractory temporal lobe epilepsy represent the main set of patients currently being submitted to seizure surgery. These procedures generally include both neocortical and hippocampal resections and are generally performed with the aid of intraoperative electrocorticography.

Brain lesions observed previously only in pathological specimens, such as mesial temporal sclerosis, can now be detected “in vivo” after the availability of high resolution magnetic resonance imaging in clinical practice.

Mesial temporal sclerosis represents the pathological findings in up to 95% of the cases submitted to temporal lobe resections. Actually, its absence raises suspicion of non-temporal lobe seizures and epilepsy. On the other hand, there is still ongoing discussion on the issue of the nature of mesial temporal sclerosis as related to epilepsy. Data suggesting both that mesial temporal sclerosis is the cause of temporal lobe epilepsy or the consequence of repeated temporal lobe seizures are available in the literature.

This paper describes a previously non-epileptic patient in whom unilateral mesial temporal atrophy originated after a systemic insult, followed by the development of epilepsy, suggesting that at least in this case mesial temporal atrophy may be related to the cause of epilepsy.

CASE REPORT

CB, a 13-years-old boy was a normal child until the age of 6. By this time, he developed appendicitis and was submitted to surgery. In the immediate post-operative period he developed hyperosmolar coma after receiving several miscalculated bolii of intravenous glucose. The coma lasted for 3 days and was associated with no focal neurological deficits or seizures. He recovered uneventfully from a neurological point of view. Seizures started 3 months afterwards. They were uncinate and autonomic simple partial and complex partial seizures and had a post-ictal state characterized by confusion and disphasia. Initial seizure frequency was 1/week but within the following years it increased to up to 1/day with several clusters comprising 3-4 seizures/day, despite aggressive mono- and polytherapy drug treatment regimens. Memory complaints were initially recorded by the age of 10. By the time of pre-surgical evaluation (age of 13) he was experiencing 2-3 seizures/week. Neurological examination was normal. Neuropsychological testing showed verbal memory deficits. Dichotic listening testing showed left-hemisphere speech dominance. He passed a left carotid artery injection in the amygdaloid procedure and failed contralaterally from a memory point of view. Left hemisphere speech dominance was confirmed during the intracarotid amytal procedure. Interictal EEG abnormalities were restricted to the left temporal lobe with phase reversals over the left zygomatic electrode. Ictal recording showed consistent seizure onset over the left temporal lobe. A CT scan obtained 1 month after the onset of seizures suggested left temporal lobe atrophy. MRI obtained by the time of the pre-surgical evaluation showed mesial temporal sclerosis with further volumetric decrease of the parahippocampal and fusiform gyri and uncus. This patient has been submitted to a cortico-amygdalo-hippocampectomy under electrocorticographic guidance (Fig 1). Pathological examination disclosed hippocampal sclerosis. The patient has been seizure free since surgery (follow-up = 10 months).

DISCUSSION

Hyperosmolar coma is often related to focal neurological deficits and focal seizures without the evidence of definitive structural lesions. On the other hand, systemic stresses (primarily non-neurological) can give rise to focal brain lesions. Differential thresholds for lesion generation after systemic insults is well known and brain regions with terminal irritation, such as the temporo-parieto-occipital region and hippocampus seems to be very susceptible regions for ischemic vascular injury. The anatomical lesion present in our patient is very likely related to the systemic pathological episode that followed abdominal surgery.

The early appearance of seizures in this patient is compatible with a close relationship of the pathological substrate and the genesis of the epileptic syndrome. It is not clear, however, if the pathologic and hodologic abnormalities seen in mesial temporal sclerosis are responsible for seizure generation or are only an epiphenomenon of a previous structural lesion. Even if we consider that
Fig 1. A: CT scan 4 months after the hyperosmolar coma showing left temporal lobe atrophy. B: Pre-operative MRI showing left mesial temporal sclerosis. C: Post-operative MRI showing the extent of the cortico-amygdalo-hippocampectomy.
mesial temporal sclerosis might be a consequence of ongoing seizure activity, it should be noted that 3 months (the latent period in our patient) is a very short period of time for the progressive development of such a lesion and that a limited number of seizures had occurred. It is very likely that this lesion was generated by the time of the hyperosmolar coma.

This young boy disclosed an epileptic syndrome very similar to the adult temporal lobe epilepsy type. Fifty percent of the adult refractory temporal lobe epileptic patients have had febrile convulsions during infancy and this could represent the analogous systemic insult responsible for the pathological substrate generation in these patients. There appears to be a genetic basis for the occurrence of febrile convulsions but it is still unclear how a systemic injury as this could induce the appearance of focal unilateral brain lesions. On the other hand, these patients typically have a latent period of years for seizure development. Our patient had no febrile convulsions but had a clear systemic insult. The different nature of the insult might has been responsible for the early appearance of seizures (short latent period = 3 months).

The extremely low prevalence of MRI abnormalities such as mesial temporal sclerosis in the normal (non-epileptic) population makes it very unlikely, although possible, that the MR abnormalities seen in this patient were present before the hyperosmolar coma. This is in striking difference with lesions such as arachnoid cysts or cortical dysplasias which are mainly congenital.

In summary, the clinical findings in our patient strongly suggest that unilateral mesial temporal sclerosis would have a close relationship to the systemic causative event that led to seizure development in this case and not to the effect of on-going seizure activity.

REFERENCES