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GELASTIC SEIZURES HYPOTHALAMIC-HAMARTOMA SYNDROME: LATE ONSET SEIZURES

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Introduction:

Gelastic seizures hypothalamic-hamartoma (GS-HH) syndrome is one of the most intriguing condition in epileptology. Its clinical hallmark is represented by laughing attacks, presenting with an early onset and evolving towards a catastrophic generalized epilettic encephalopathy. The clinical presentation comprehends precocius puberty too. Sometimes gelastic seizures could present a late onset and be associated with other partial seizures, with frontal or temporal semeiology, usually pharmaco-resistant. Here we report the ictal semeiology and the clinical features of a small patient cohort with GS-HH and late onset epilepsy

Clinical cases:

M.C. – 67 years old

Onset at 20 years old, with gelastic seizures and face-flushing. Ten years later he presented seizures with loss of contact and oroalimentary automatism. MRI findings showed a sessile hypotalamic hamartoma attached to mammillary body, (diameter <5 mm), (Fig. 1a & b). The EEG recording of a pure gelastic seizure presented little or no epileptiform abnormalities, showing only a diffuse flattening of the cerebral activity (Fig. 1c). The patient has a normal cognitive assessment, a part of a mild deficit of the mnesic functions. The therapy with CBZ was able to control the complex partial seizures, resulting only in gelastic seizures persistence.

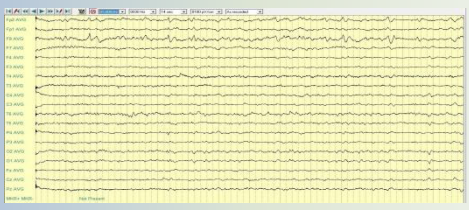

Fig. 2a - (M.V) –interictal EEG

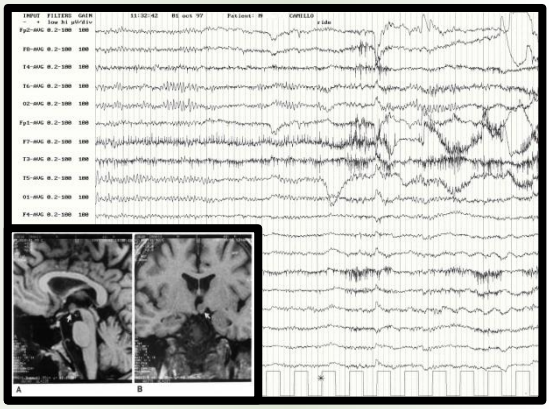

Fig. 1a e Fig. 1b MRI shows a little HH. Ictal EEG on backside

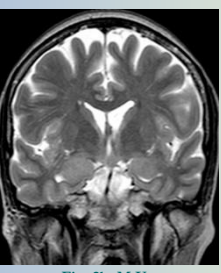

Fig. 2b M.V

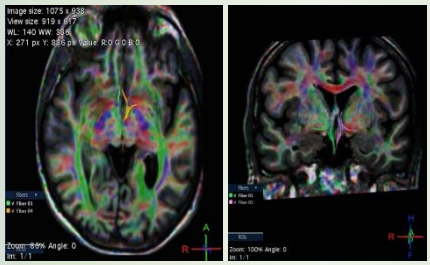

Fig. 2c e fig. 2d tractografy M.V.

M.V. - 66 years old (F)

Onset at 12-13 years old with sudden modification of facial expression, stretching of the mouth, miming a smirk of disgust, sadness and gooseflesh. This kind of seizures prosecuted unmodified in the years. The MRI showed a sessil hypothalamic hamartoma, oval shaped (diameter <1cm), attached to the left mammillar body *(Fig. 2b)*. Interictal EEG showed a right anterior temporal epileptic focus, while the ictal registration, just a a slowing of the background activity and muscolar artefacts.*(Fig. 2a)*. Cognitive assessment was normal. Seizures are not responsive to OXC.

B. G. - 59 years old (M)

Gelastic seizures since late childhood, with face flushing and sometimes lacrimation. Seizures went on unmodified in the years. The MRI showed sessil hypothalamic hamartoma (diameter < 1cm) attached to the right mammillary body*(Fig. 3a & 3b)*. The ictal EEG reported slowing of background activity and artefacts. Pet showed interictal hypometabolism of right ipothalamic temporal anterior zone *(Fig. 3c)*. The patient refused antiepileptic drugs

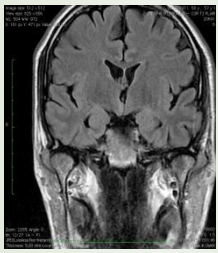

Fig. 3a

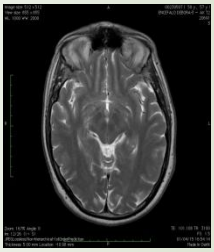

Fig. 3b

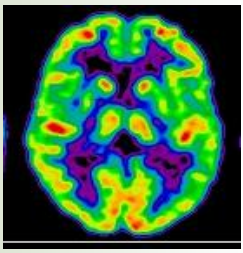

Fig. 3c

CONCLUSIONS:
Based on literature reports typical gelastic seizures usually become less clinically relevant or disappear in the adult age. In this small group of patients with late onset epilepsy, associated with Gs-HH syndrome, gelastic seizures persisted unmodified in the adult age, remarking the diagnostic importance of these seizures semeiology. Cognitive assessments are normal, or show only modest neuropsychological alterations. Moreover, the ictal EEG recorded during pure gelastic seizures could not show the evidence of paroxysmal activity

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