Cognitive neurology and neuropsychology

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BEHAVIORAL AND NEURO-STRUCTURAL MARKERS OF EARLY EMPATHIC IMPAIRMENT IN THE BEHAVIORAL VARIANT OF FRONTOTEMPORAL DEMENTIA

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Background: Changes in social behaviour, loss of empathy and impairment of social insight are early and consistent symptoms of the behavioural variant of frontotemporal dementia (bvFTD), thus constituting key-elements for differential diagnosis.

Materials and methods: We investigated alterations of empathy, both in its cognitive (Theory-of-Mind) and affective (emotional empathy) components, in 13 mild bvFTD patients (CDR≤1) and 39 age-, gender- and education-matched healthy controls. They performed a cartoon-task, requiring the identification of the correct ending (among three possible choices) of comic strip lacking verbal components and sub-divided in 3 conditions (Theory of mind-ToM; empathy-Emp, causal inferences-Caus), with either one or two interacting characters. Voxel-Based-Morphometry (VBM) was used to correlate performance with grey-matter (GM) density, both in whole-brain and in a-priori regions-of-interest.

Results: Behavioural results highlighted deficits in bvFTD, compared with controls, in all conditions and particularly in Emp with one character. VBM analyses showed significant correlations between both Emp and ToM performance and GM-density in the left temporo-parietal junction, right temporal pole and amygdala, structures previously associated with mentalizing. Anterior cingulate cortex and bilateral amygdala and insula (involved in emotional processing), were associated with performance in Emp conditions, while medial prefrontal regions were specifically associated with Emp involving two characters.

Discussion: We provide behavioural and neuro-structural evidence, supporting the early impairment of cognitive and emotional empathy in bvFTD, associated with overlapping but distinct brain-networks. The emotional component of empathy (mainly associated with prefrontal, temporal and limbic damage) is particularly disrupted in mild bvFTD, thus representing a potential marker for early diagnosis.

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THE "EXTENDED LOCKED-IN SYNDROME"

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The locked-in syndrome is one of the most devastating neurological conditions. However, despite thorough description of the condition and its clinical appearance, the classic locked-in syndrome, which is defined as quadriplegia, only vertical eye movement and blinking possible with preserved cognitive abilities, seems to be infrequently present. Since MRI verified isolated damage to the pons poses the finding in this certain case, the question arises, how the symptomatology increases, if additional lesions are found in cranial brain areas. Therefore, the terminus complete Locked-in syndrome was proposed in 1979, characterizing the total loss of voluntary muscle movement. However, as damage can spread to occur in main brainstem arousal centres and in more cranial brain areas, disablement of cognitive functions can be expected. Here, brain areas of special interest constitute the mesencephalic structures, especially the periaqueductal grey matter to the thalamic nuclei. In such remarkable cases, brain damage does not only affect the patients' motor abilities, but also their consciousness. We demonstrate five cases of locked-in syndrome, each with different patterns of structural lesion, as obtained by 3T MRI in great detail and discuss how clinical appearance and imaging results relate to each other. The question will be approached if it is useful to differentiate extended forms locked-in syndrome with differentiated severe chronic disorders of consciousness as for instance akinetic mutism and parasomnial syndromes. We analyzed the lesions from the brainstem and propose a hierarchical scheme of consciousness.

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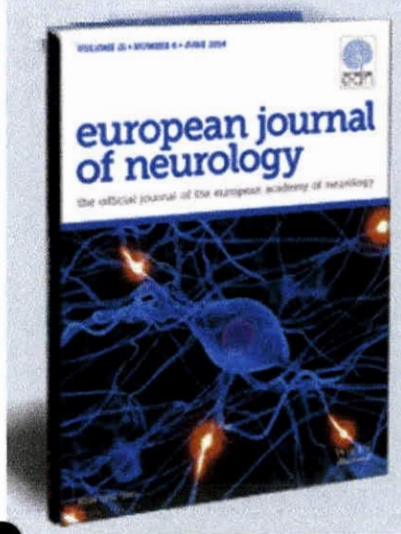

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POSTER SESSIONS

Poster Session 2, Monday 12 September

The Extended Locked-in Syndrome

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Introduction:

The locked-in syndrome is one of the most devastating neurological conditions. However, despite thorough description of the condition and its clinical appearance, the classic locked-in, which is defined as quadriplegia, only vertical eye movement and blinking possible with preserved cognitive abilities, seems to be infrequently present. This syndrome is also referred to as bilateral ventral pontine syndrome, which in respect neuroanatomically explains the symptomatology. Since isolated damage to the pons poses the finding in this certain case, the question arises, how the symptomatology increases, if additional lesions are found in cranial brain areas. Therefore, the terminus complete locked-in was proposed in 1979, characterizing the total loss of voluntary muscle movement [6]. However, as damage can spread to more cranial brain areas, disablement of cognitive functions can be expected. Here, brain areas of special interest constitute the mesencephalic structures, especially the reticular formation and thalamic nuclei. In such remarkable cases, brain damage does not only effect the patients' motor abilities, but also reveals symptoms of apallic syndrome. This combination of a Locked-in Syndrome and symptoms of apallic syndrome is what we propose as an "Extended Locked-in Syndrome". We present two cases.

Case one:

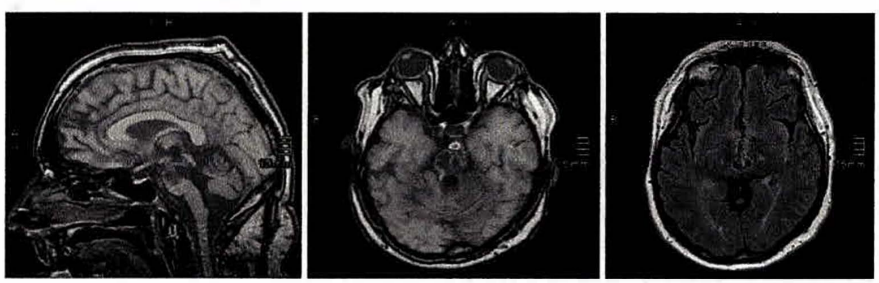
A forty-year-old male was admitted to the hospital with symptoms of vertigo, slurred speech, hemiplegia on the left side, with horizontal gaze palsy, somnolent but sociable. An admission CT scan with i.v. contrast showed occlusion of the distal part of the basilar artery. The posterior cerebral arteries were supplied by the carotid arteries, only the superior cerebellar arteries showed diminished flow. There was no sign of intracranial hemorrhage and an infarct zone had not been demarked. The patient was previously treated with antibiotics for a febrile infection and had a history of nicotine dependency. Premedication included NSAIDs, PPI and Tetrazepam for chronic back pain.

Case two:

A twenty-nine-year- old female was admitted to the hospital intubated and sedated, after she was found with a Glasgow Coma Scale value of 4. Both pupils were dilated with anisocoria (the right pupil larger) and showed no direct or indirect reaction to light. Oculocephal -, swallowing –and gag reflex were absent. Due to wide spread lesions in the initial CT scan and multiple vascular occlusions in DSA no intra-arterial therapy was performed. The patient suffered from obesity and mild nicotine dependency.

[1] "Top of the basilar" syndrome. Caplan LR. Neurology. 1980 Nov;30(11):72-9.
[2] Hypersomnia following paramedian thalamic stroke: a report of 12 patients. Bassetti C, Vella S, Donati F, Wielepp P, Weder B. Ann Neurol. 2000 Apr;47(4):433-41.
[3] Hypersomnia and thalamic and brain stem stroke: a study of seven patients] Silveira M, Espinola M, Arpa J, Serrano P, Rodríguez-Alfaro A. Neurologia. 1999 Jun-Jul;14(6):307-14. [Article in Spanish]
[4] Pseudohypersomnia and pre-sleep behaviour with bilateral paramedian thalamic lesions. Castellanos NP, Cano-Salva MA, Goldberg MP. Brain. 1993 Dec;116 (Pt 6):1549-63.
[5] Persistent akinetic mutism after bilateral paramedian thalamic infarction. Crespo AC, Bertges L, Casanova S, Servo S, Strigaro G, Monaco F. J Neuropsychiatry Clin Neurosci. 2008 Summer;20(3):351.
[6] Varieties of the locked-in syndrome. Bauer G, Gerstenbrand F, Rumpl E. J Neurol. 1979 Aug;221(2):77-91.

Magnetic resonance imaging case one:

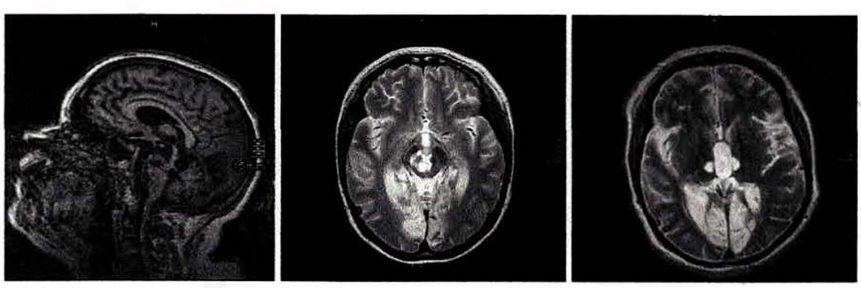


Findings:

Cystic-gliotic degeneration of the bilateral paramedian pons and mesencephalon. Bilateral paramedian thalamus affected as well (dorsomedial nuclei, see third picture). Dorsally, the degeneration of the brainstem reaches the fourth ventricle. Small islands of preserved tissue. High degree of atrophy of the cerebellar peduncles, the cerebellum and the medulla oblongata. Degeneration of bilateral temporomesial structures, especially the parahippocampal gyrus.

Negligible small chronic vascular lesions in supratentorial white matter. The corticospinal tract is affected at a mesencephalic and pontine level.

Magnetic resonance imaging case two:



Findings:

Cystic-gliotic degeneration of the bilateral paramedian pons and mesencephalon. The right side being more affected. Bilateral paramedian thalamus affected as well (dorsomedial nuclei, see third picture). Infarctions in the posterior cerebral artery territory bilaterally, as well as in the left middle cerebellar peduncle and the left cerebellum. Expansion of the third ventricle. The corticospinal tract seems to be completely cut at the level of the pontomesencephalic junction. Slight atrophy of the posterior part of the corpus callosum, the cerebellum and brainstem.

Clinical presentation and discussion:

What both patients have in common, are widespread brain lesions after basilar artery thrombosis, exceeding those described as „top of the basilar syndrome" [1]. Both patients clinically present unresponsive to auditory, visual and noxious stimuli. Blinking –and gag reflexes are present in both patients. Both patients also show increased signs of sleepiness, having the eyes closed most time of the day, unable to be aroused by any kind of stimuli ((Pseudo-) Hypersomnia, due to disruption of the mesencephalic reticular formation and/or paramedian thalamic damage [2,3,4]). Within this clinical presentation, the syndrome of an akinetic mutism, which has been described after bilateral paramedian thalamic infarction [5] may be hidden. It is important to mention that both patients do neither have spontaneous nor provoked limb movement. Due to radiologic evidence of a disruption of the corticospinal tract, the overall picture fulfills the condition of a Locked-in Syndrome. We propose the terminus „Extended Locked-in Syndrome" for patients with evidence of disconnection of the corticospinal tract and symptoms of apallic syndrome, which may be due to damage to the reticular formation or to bilateral paramedian thalamic nuclei.

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