PARACELSUS MEDIZINISCHE PRIVATUNIVERSITÄT

Karl Landsteiner Institut für Neurorehabilitation und Raumfahrt-Neurologie

The Extended Locked-In-Syndrom

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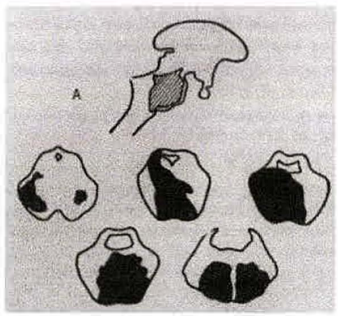
Locked-In Syndrome (Plum & Posner, 1966)

- No possibility to communicate with surrounding except with eye lids (blinking morse code) and vertical eye movements
- Consciousness and perception fully maintained
- Total paralysis of all extremities, trunk, neck and motor brain nerves, inclusive impairment of swallowing
- Spontaneous respiration possible
- Alpha-EEG

Etiology of Locked-In Syndrome

- Mosty caused by ischemic stroke or hemorrhage, affecting the corticospinal, corticopontine and corticobulbar tracts in the brainstem, e.g. infarction caused by basilar thrombosis (León-Carrión et al 2002)
- midbrain infarctions of the bilateral cerebral peduncles causing LIS have been reported (Karp and Hurtig 1974, Zakaria and Flaherty 2006)
- Traumatic lesion (León-Carrión et al 2002)
- Encephalitis, pontine abscess (Murphy et al, 1979)
- Brainstem tumor (Cherington et al, 1976)
- Central pontine myelinolysis, toxines and heroine abuse (Inci and Ozgen, 2003)

Pons and midbrain lesions causing LIS



Quelle: F. Plum, J.P. Posner 1972: Diagnosis of Stupor and Coma

Different Types of LIS (Bauer, Rumpl, Gerstenbrand, 1979)

- According to neurological symptoms
  - Classical Locked-In syndrome: total immobility except for vertical eye movements and blinking
  - Incomplete Locked-In syndrome: if any other movements are present
  - Total Locked-In syndrome: immobility, including all eye movements, combined with signs of undisturbed cortical function in the EEG.
- According to time course
  - Chronic Locked-in syndrome
  - Transient Locked-in syndrome

Additional clinical symptomatology in the Extended Locked-In-Syndrom

- Acinetic mutism (Cairns et al, 1941)
  - Lesion: region 3rd ventricle, periaqueductal
  - Clinic: Disturbance in the initiation of spontaneous and intentional movement, awareness undisturbed
- Stupor (Plum and Posner, 1972)
  - Lesion: intralaminar nucleus thalamus
  - Clinic: Deep sleep, unresponsiveness, temporarily arousable
- Hypersomnia (Jefferson, 1952)
  - Lesion: mesodiencephal
  - Clinic: Dormancy, continuously, not arousable
- Parasomnia (Facon et al, 1958)
  - Lesion: periaqueductal
  - Clinic: Permanent dormancy, awakes by himself after months

Patient S.M.: extended LiS

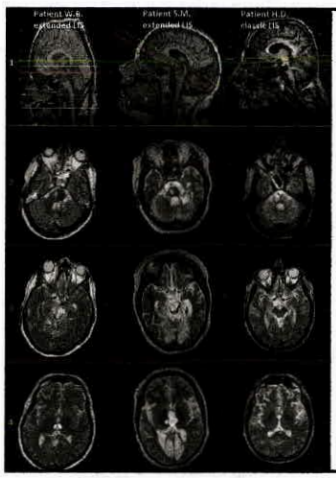
- Clinical characteristics: 21a, female, divergent bulbi, areagible pupils, anisocoria r > l, missing OCR, corneal reflex and gag reflex. Flaccid quadriplegia. Babinski's sign negative, distal flexor contractures, no reaction to noxious stimuli, distinct frontal release signs: orbicularis oris reflex, sucking reflex present. Additionally the patient shows a hypersomnia syndrome, an acinetic mutism and a bilateral thalamic hand: flexion in the MCP joint, extension in the distal joints.
- Imaging findings: Gliotic transformation on the right side of the pons, the left middle cerebellar peduncle, the bilateral dorsomedial thalamic nuclei, and bilateral occipital, mesiotemporal and cerebellar brain regions.
- EEG: severe abnormalities

Patient W.B.: extended LiS

- Clinical characteristics: 44a, male, divergent bulbi, arreagible pupils, anisocoriar > l, no visual pursuit, OCR and corneal reflex missing. Flaccid quadriparesis, distal flexor contractures, no reaction to noxious stimuli, Babinski's sign present, no communication with the patient possible – additionally the patient presented with a hypersomnia syndrome and an acinetic mutism.
- Imaging findings: Gliotic degeneration oft the upper 2/3 of the brainstem paramedian bilaterally, involvement of midbrain structures and dorsomedial thalamic nuclei bilaterally
- EEG: severe abnormalities

Patient H.D.: classic LiS

- Clinical characteristics: Round, isocor pupils, reactive to light, OCR and corneal reflex present, no paralysis of eye muscles, spontaneous movement of the right upper and lower extremity, retraction after noxious stimuli, plegia of the left upper and lower extremity, communication possible with the use of eye movements and movement of the right side extremities.
- Imaging findings: Isolated damage to the ventral pons, sparing of midbrain and thalamic structures, gliotic degeneration in the right cerebellar hemisphere and in the right upper and middle cerebellar peduncle.
- EEG: alpha-EEG



MRI Images

(Sagittal T1w, axial T2w):

In the vertical column, there are MRI Images of each patient at different levels:

1: Sagittal plane

2: Pons level

3: Midbrain level

4: Thalamus level

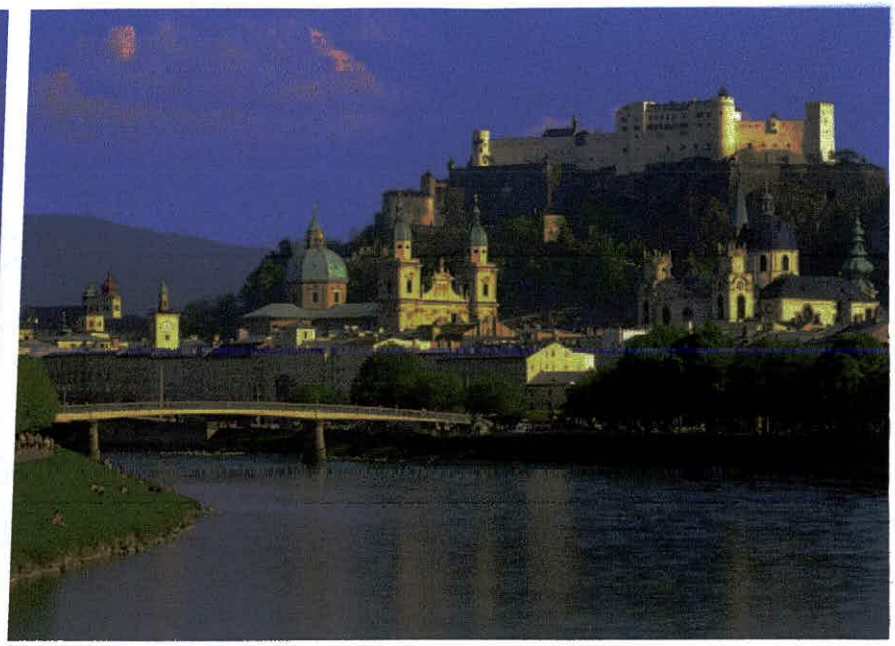
Discussion

- In order to meet the diagnostic criteria for classic LiS, brain damage has to be restricted to the ventral part of the pons, comprising the corticospinal and corticobulbar tracts, as well as the paramedian pontine reticular formation (PPRF), which is in particular responsible for horizontal eye movements and saccades.
- Vertical eye movements and blinking are controlled by the rostral interstitial nucleus of the medial longitudinal fasciculus (riMLF) , which is located cranial in the pontomesencephalic junction. Therefore involvement of paramedian mesencephalic structures can cause vertical gaze paralysis that lead to the clinical presentation of a total LiS.
- Sparing of PPRF and riMLF results in incomplete LiS presenting with horizonal and vertical eye movements.

Discussion

- Extended LiS: variety of additional extrapontine brain lesions with corresponding clinical symptoms:
  - Consciousness: hypersomnia, acinetic mutism, stupor, parasomnia
  - Frontal release signs
  - Thalamic posturing of hand and/or feet
  - Temporal and occipital signs may also be present in case of an involvement of occipital or temporal brain regions (e.g. basilar thrombosis with embolization in posterior ateries or into hippocampal/parahippo-campal regions)


Thank you for your attention!



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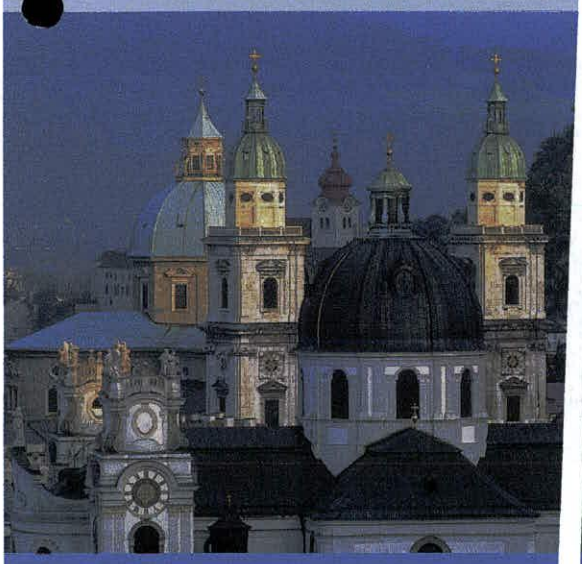

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