Neurologist-in-training

The aim of this section is to prepare the neurologist-in-training for the FMH examination, to confront her or him with specific problems of everyday neurological practice and to provide him or her with updates on recent controversies in clinical neurology.

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The missed "top of the basilar" syndrome

A 78-year-old woman was found by her husband to be unresponsive, without additional signs, following a nap. She was admitted to the emergency ward of the hospital with a GCS of 4, RR 174/75 mm Hg, miosis, O_2 saturation of 88%, no vomiting or further pathological signs.

CT-scan showed an old infarction of the temporo-parietal right cortex dating from 2008. No further lesions were reported. The basal arteries were assumed to be normal but artefacts prevented clear-cut judgement.

The patient was on opioid medication for long-standing back pain.

An overdose of opioids was assumed to be the possible cause of the unresponsiveness, and repeated doses of naloxone were therefore administered. At times the patient apparently became responsive for short intervals, but was agitated when awake.

She was transferred to the intensive care unit for further diagnostic workup and monitoring. The spinal fluid was unremarkable. Doppler sonography revealed no clear-cut pathology of the vertebro-basilar system, although the signal quality was restricted.

The patient stayed on the emergency ward for three days with alternating irregular periods of hypersomnia and unresponsiveness on the one hand, and agitation coupled with delirium on the other. Status epilepticus was ruled out by EEG.

The aetiology of the obvious disturbance of consciousness remained undetermined.

The patient was transferred to the neurological ward on day 4 with the following medication:

- aspirin 100 mg 1/d
- levetiracetam 500 mg 2/d
- lisinopril 10 mg 1/d

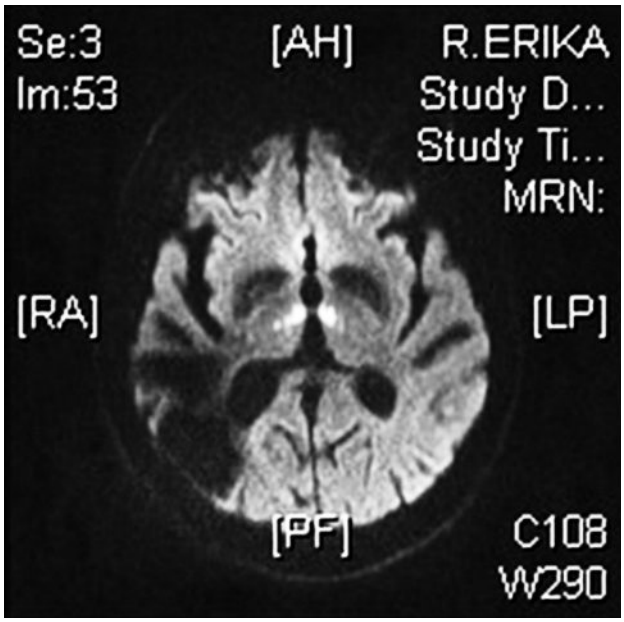
MRI (fig. 1) revealed symmetrical bithalamic lesions in the diffusion-weighted sequences six days after admission to hospital. MR angiography showed arteriosclerosis of the distal basilar artery. Cardiac workup was unremarkable.

Clinically the episodes of hypersomnia ceased and sleep-waking rhythm normalised. The neurological status was unremarkable, except for an incomplete vertical gaze paresis for upward movement and serious neuropsychological deficits persisting until transfer to the neurorehabilitation clinic after two weeks.

Neuropsychological tests one week after admission showed a distinct functional disturbance with hypersomnolence, significant memory deficits, disorientation and confabulation resembling Korsakoff's syndrome.

All symptoms showed a steady tendency towards recovery until discharge.

Figure 1
Axial diffusion-weighted images of MRI showing the acute bithalamic lesions of medial dorsal thalamus and the old ischaemic parieto-temporal lesion from 2008.



A) What has been missed?

It can be extremely difficult and challenging to judge an unresponsive patient in the emergency situation. Two precipitating factors might have been decisive i.e., first the patient's history of opioid intake of unknown amount and secondly the restricted quality of the initial CT-scan, which failed to reveal the vertebro-basilar pathology.

In the absence of a clear visualisation of the pathology of the distal basilar artery by CT-scan, the patient's clinical picture was misinterpreted as the consequence of opioid withdrawal.

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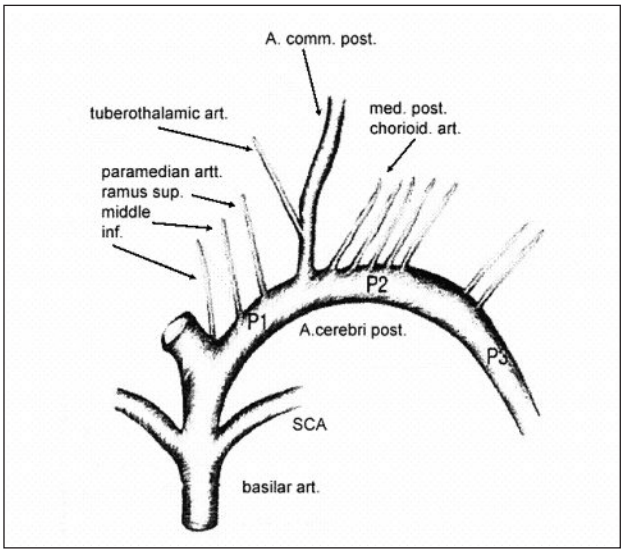
B) What is important to be kept in mind?

Every patient with a stroke-like episode and unresponsiveness, hypersomnolence, disorientation and gaze disorders should arouse suspicion of **"top of the basilar" syndrome** [1, 2, 3]. The syndrome may comprise a great variety of symptoms depending on the individual patient's vasculature at the distal basilar and posterior arteries and the degree of embolism. The posterior arteries and the thalamic perforating arteries (fig. 2) may be involved to a varying degree, thus causing syndromes which may additionally be characterised by visual disturbances and even more profound thalamic and frontal executive symptoms [1, 4, 5].

The pattern of bithalamic (dorsomedial thalamus) lesions of our patient point to a vascular embolic disease of the distal basilar artery with occlusion of the superior ramus of the paramedian artery (generally originating from the P1 segment of the posterior artery and corresponding to the posterior thalamo-subthalamic paramedian artery of Percheron). The paramedian arteries arise either as a pair from each P1 or originate from a common trunk of one P1, thus supplying the thalamus bilaterally [1, 6, 7]. Figure 2 demonstrates schematically the anatomy of the distal basilar artery and its branches.

Figure 2
Distal basilar artery (modified from Schmahmann et al. [2], reprint permission from Wolters Kluwer Health).



Abstract

The missed "top of the basilar" syndrome may be the most catastrophic event in a patient's and doctor's career. It must be assumed to be a common event anywhere leading to significant disability and death. This is due to a great variability of symptom presentation e.g., coma, somnolence, waking–sleep disturbances, visual disturbances, hallucinations, agitation, delirium and memory dysfunction, etc. The reason is a chiefly embolic obstruction of the distal basilar artery, the P1 arteries or their branches e.g., the different thalamic branches. MRI provides the best early diagnosis. Therapy must be introduced as soon as possible and patients should be transferred to a stroke centre providing adequate treatment.

MCQ

1. *Which symptom does not point to a "top of the basilar"?*
A Unresponsiveness
B Hemiparesis
C Severe memory loss
D Vertical gaze paresis
E Hypersomnolence

2. *What is the basic pathophysiology of bithalamic infarcts?*
A Heart embolism
B Thrombosis of the basilar artery
C A thalamic artery arising directly from the basilar artery
D Extraordinary vulnerability of the thalamus
E A paramedian thalamic artery arising as common trunk from P1

3. *Considering the variability of embolism of the distal basilar artery, which of the following symptoms is the most helpful for early diagnosis?*
A Seizure
B Hemianopia
C Unresponsiveness
D Tetraplegia
E Skew deviation

4. *What is the correct early management of suspected "top of the basilar"?*
A Transfer from the emergency room to an intensive care unit
B Careful study of the individual risk factors for embolism
C Intravenous thrombolysis and transfer to ICU
D MRI imaging
E Effective anticonvulsant medication

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Answers to MCQ

1. B
2. E
3. C
4. D