

Hippocampal sclerosis, ILAE Type 2 (CA1 - predominant neuronal loss)

Clinical history. A 56-year-old female patient with right-sided temporal lobe epilepsy since the age of eight. She suffered from many epigastric auras and automotor seizures, and also experienced a generalised tonic-clonic seizure. Repeated MRI at 3T revealed no hippocampal abnormality (T2-weighted turbo spin-echo MRI) (*figure 1A; white arrow*). Neuropsychological examination disclosed no cognitive dysfunction of the right hemisphere. At the time of surgery, she was treated with LEV.

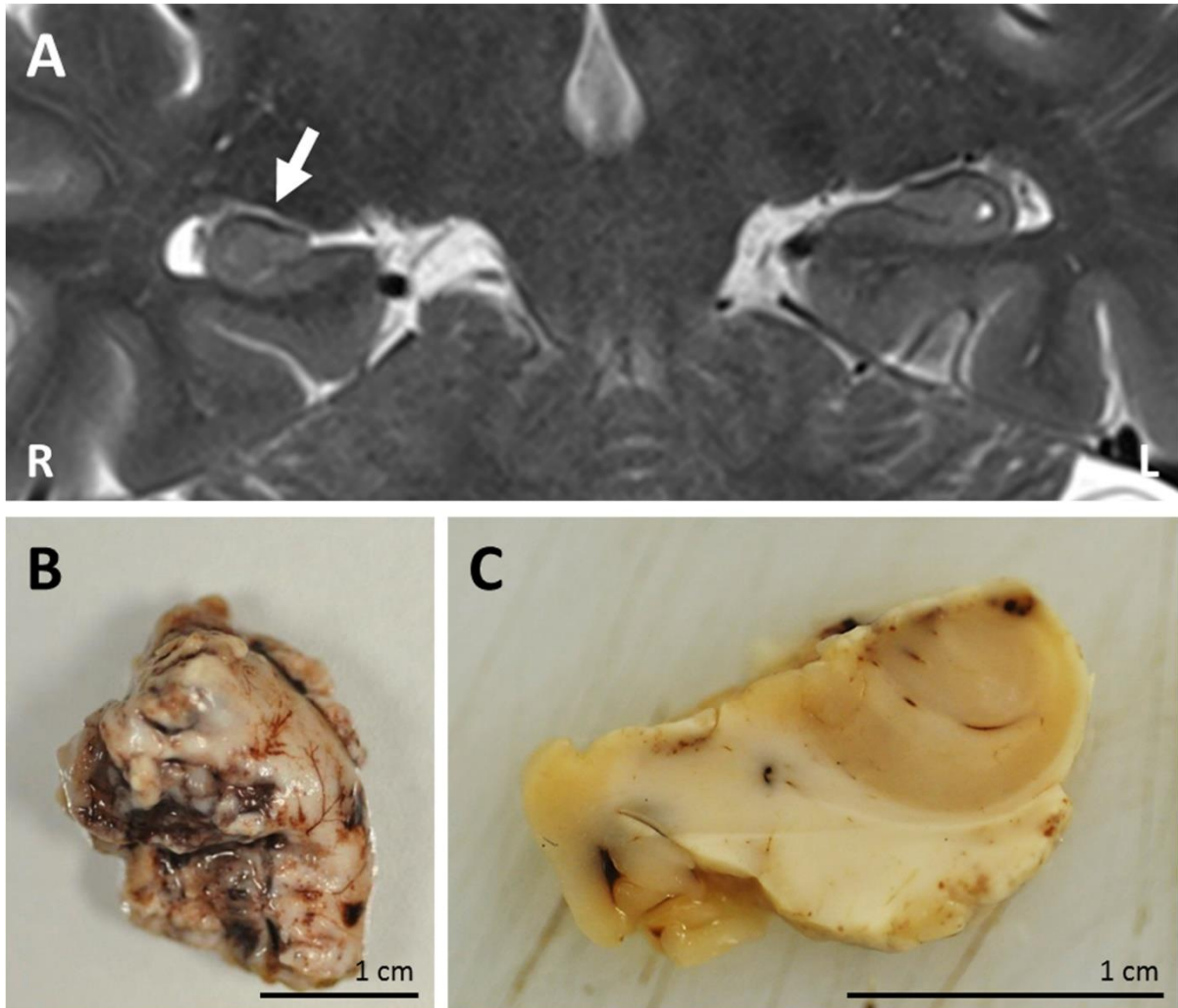


Figure 1.

Histopathology and immunohistochemistry. We received en bloc resected hippocampal tissue (3x2.2x1 cm) (*figure 1B*). The specimen was dissected perpendicular to the anterior-posterior axis into six 5-mm thin sections (*figure 1C*). All samples were fixed in formalin and embedded into paraffin. H&E staining revealed a well preserved hippocampal anatomy with severe neuronal loss predominately in sector CA1, whereas CA2, CA3 and CA4 remained

preserved. CA1 showed concomitant fibrillary gliosis (GFAP). The granule cell layer showed a few areas of granule cell dispersion.

Comments. Neuropathological examination revealed a predominant pattern of segmental pyramidal cell loss and fibrillary gliosis in the hippocampal CA1 area, consistent with the diagnosis of HS ILAE Type 2 (Blumcke *et al.*, 2013). The dentate gyrus showed no granule cell depletion but foci of granule cell dispersion. No evidence of tumour or acute inflammation was seen.

Microscopic findings in HS ILAE Type 2

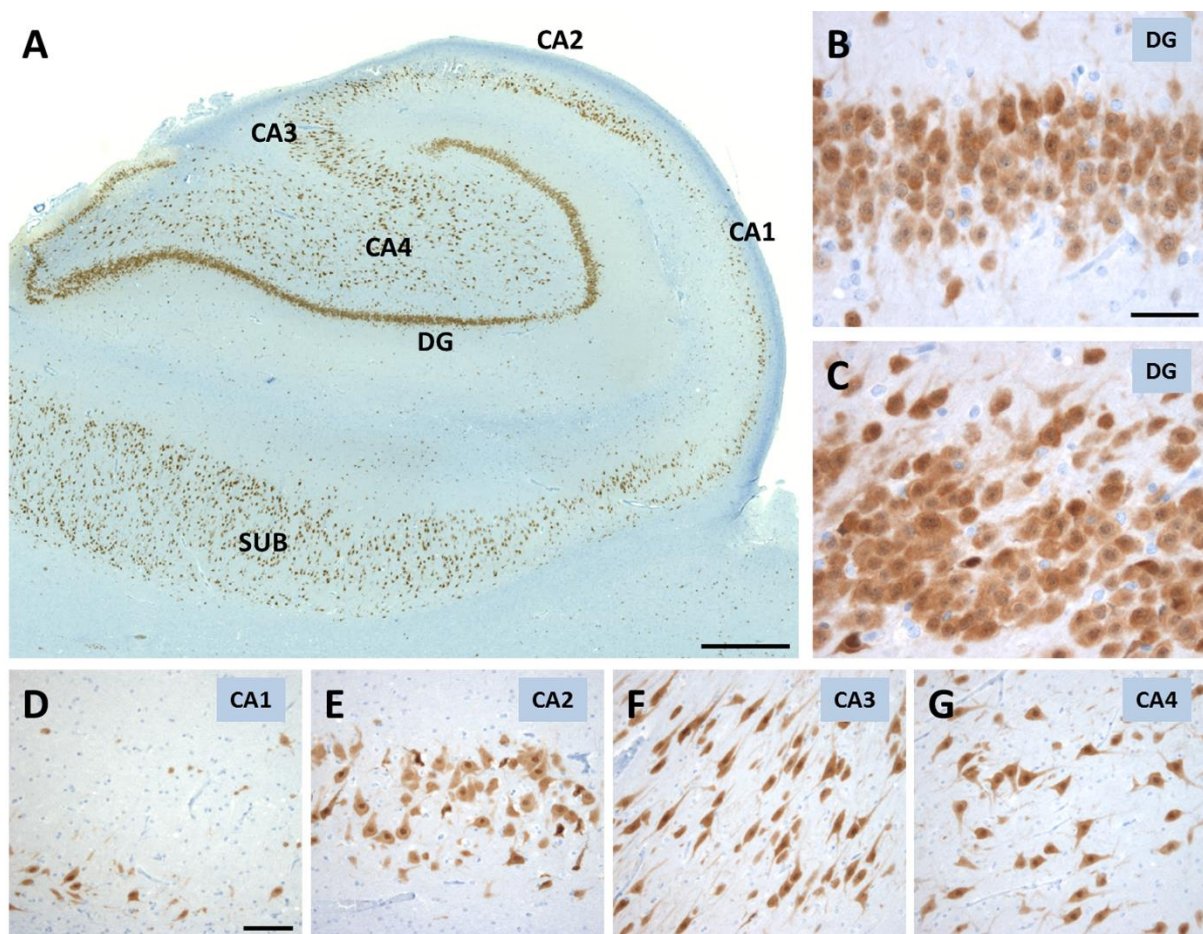


Figure 2. Microscopic findings; NeuN immunohistochemistry. (A) Segmental pyramidal cell loss predominantly affecting sector CA1. (B, C) The granule cell layer mostly has a normal granule cell density (B), with a few areas of granule cell dispersion (C). (D-G) Severe neuronal loss predominately in sector CA1, whereas CA2, CA3 and CA4 are preserved. Scale bar in (A): 1000 μ m. Scale bar in (B, C): 50 μ m. Scale bar in (D-G): 100 μ m.