

Short report

Periodic EEG discharges and status spongiosus of the cerebral cortex in anoxic encephalopathy: a necropsy case report

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SUMMARY A periodic EEG pattern very similar to the changes in Creutzfeldt-Jakob disease was seen in a case of anoxic encephalopathy. Necropsy revealed status spongiosus of the cerebral cortex. Generalised repetitive sharp transients in anoxic encephalopathy occur when the patient is comatose and carry a poor prognosis. They are not specific for Creutzfeldt-Jakob disease.

A periodic EEG pattern occurs in a heterogeneous group of diseases, such as subacute sclerosing panencephalitis, Creutzfeldt-Jakob disease, herpes simplex encephalitis, anoxic encephalopathy, metabolic encephalopathy, and drug intoxication. We now report a case in which an anoxic insult of the brain caused periodic EEG changes, strikingly similar to those seen in Creutzfeldt-Jakob disease. The EEG abnormality was correlated with neuropathological findings verified by necropsy.

Case report

A 52-year-old man with a long history of alcoholic abuse and malnutrition was admitted to the Saint Louis Veterans Administration Hospital with profound cyanosis. Since the age of 49 years he had experienced tremor of the hands and had been unable to walk straight. Two weeks prior to admission he became apathetic and stopped eating. He had a respiratory arrest in an ambulance on the way to hospital. On admission he was intubated and ventilation was maintained on a respirator. His blood pressure could not be obtained for 75 minutes despite the administration of dopamine. Initially he was stuporous and made occasional spontaneous movements of the extremities, but, with thiamine, he became gradually alert. On the 7th hospital day he was able to follow simple verbal commands. On

dolls' eye manoeuvre, or on request to move the eyes, only slight deviation from the primary position was noted. Caloric responses were absent. He moved all four extremities and had a symmetrical facial grimace. On the 12th hospital day he had a sudden cardiac arrest, but was resuscitated after three minutes of electrical silence in the ECG. Subsequently he remained comatose, responding only to painful stimuli with decerebrate posturing. Corneal and gag reflexes were preserved. Deep tendon reflexes were absent and plantar responses were extensor bilaterally. On the 13th hospital day the EEG showed generalised bilaterally synchronous spike-sharp wave complexes having a duration of 150-200 ms and an amplitude of 60-150 microvolts (fig 1). They occurred in a periodic fashion at the intervals varying between 1.1 and 1.7 second. The background activity between the periodic transients was markedly attenuated. The periodic discharges were not synchronous with ECG recording and were not modified by external stimuli. The EEG repeated three days later did not show such periodic discharges, but showed low voltage alpha-like rhythm of 7-9 Hz and 10-20 microvolts. This alpha-like rhythm was distributed diffusely and was not modified by painful stimuli. He never regained consciousness and died on the 40th hospital day.

Serial coronal sections of the brain revealed multifocal brownish cortical atrophy with softening throughout the cerebral cortex and cortical laminar necrosis (fig 2a, b). There was severe atrophy of the mamillary bodies and brownish softening of the periaqueductal grey matter and the subependymal grey matter around a third ventricle (fig 2c, d). Microscopic examination of the brain revealed neuronal loss, astrocytic gliosis, capillary proliferations and petechial haemorrhages in periaqueductal and periventricular grey matter. Loss of neurons and status spongiosus due to disintegration of the cortical layers were noted in every sample of the cerebral cortex (fig 2e, f). The

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Periodic EEG discharges and status spongiosus

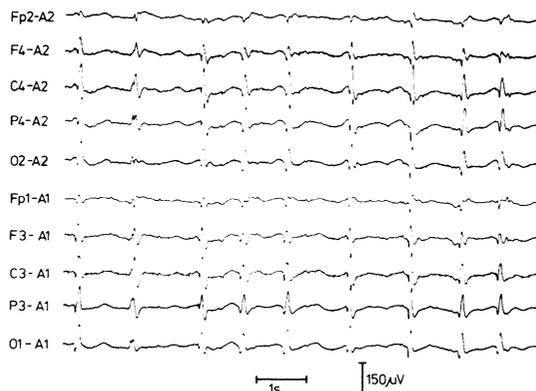


Fig 1 An EEG taken on the 13th hospital day. Note the periodic EEG discharges resembling closely to those seen in Creutzfeldt-Jakob disease.

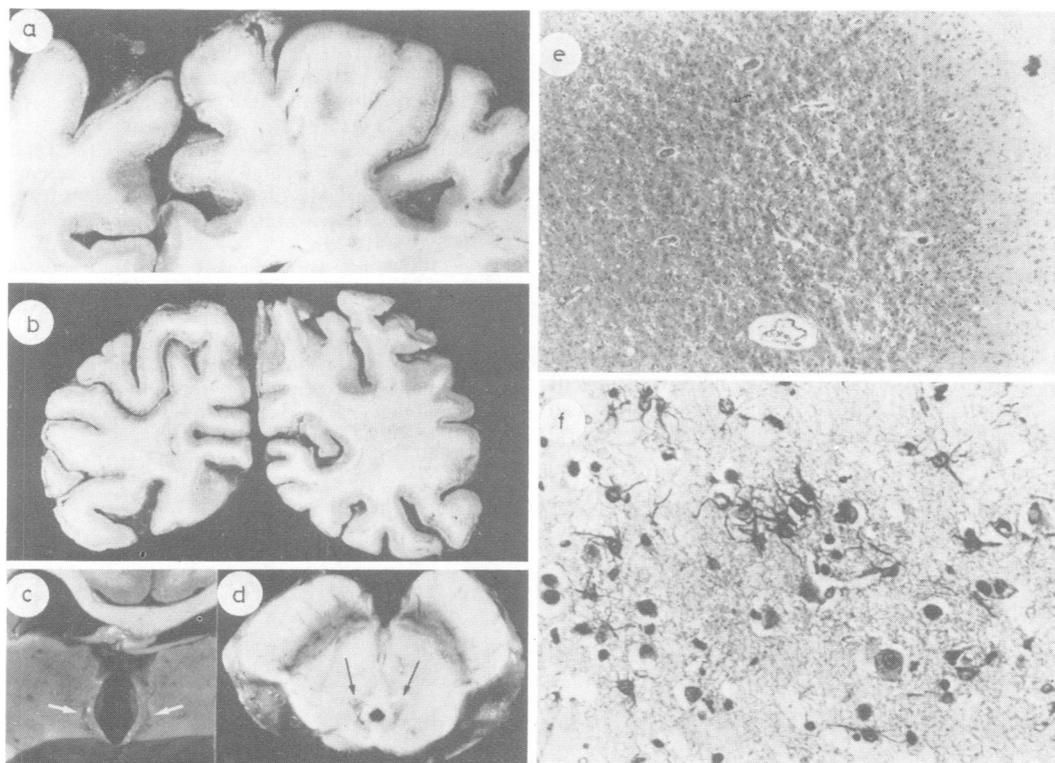


Fig 2 (a) Coronal slice of frontal lobes showing multifocal brownish cortical softening. (b) Coronal slice of occipital lobes showing cortical laminar necrosis. (c) White arrows indicate subependymal grey matter softening around a third ventricle. (d) Black arrows indicate brownish softening of periaqueductal grey matter in the midbrain. (e) Microscopic appearance of frontal lobe cortex. (H & E, $\times 27$) Note status spongiosus predominantly seen in intermediate cortical layers. (f) Microscopic appearance of frontal lobe cortex. (Holzer stain, $\times 170$) Note marked astroglyosis and status spongiosus.

third cortical layer was most severely involved. In contrast to the cortical damage, the cerebral white matter was normal, except for a small white matter softening in the right temporal lobe. Thalamic nuclei, basal ganglia and mesencephalic reticular formation were normal. In the cerebellum there was an extensive loss of Purkinje cells.

Discussion

This patient suffered from cerebral anoxia due to cardiorespiratory arrests. The history was also characterised by chronic alcoholism for many years and necropsy confirmed the presence of Wernicke's encephalopathy. An EEG on the day after a cardiac arrest was strikingly similar to that seen in Creutzfeldt-Jakob disease. In our earlier report,¹ we defined "Generalised Repetitive Sharp Transients" as sharp waves or spikes occurring synchronously over both hemispheres at the intervals varying between 0.5 and 2.0 seconds with attenuated background activity between transients. They are a useful diagnostic clue in Creutzfeldt-Jakob disease.²⁻⁴ However comatose patients suffering from severe anoxic encephalopathy can also present almost identical EEG changes, as seen in our patient. The similarity between the periodic discharges in our case and characteristic generalised repetitive sharp transients in Creutzfeldt-Jakob disease is so striking that there may be common mechanism for the generation of the transients in each. A few reports⁵⁻⁷ described their occurrence in anoxic coma. Generalised repetitive sharp transients in anoxic encephalopathy occur when the patient is comatose and carry a poor prognosis; every patient died. Two cases of Nilsson *et al*⁶ as well as our case had status spongiosus and loss of neurons throughout the cerebral cortex. Furthermore the cortical predilection of cerebral lesions in our case has been known to occur in Creutzfeldt-Jakob disease. Diffuse cortical involvement due to status spongiosus of the cerebral

cortex may be the common neuropathological factor for the generation of those transients. Our hypothesis agrees well with experimental work reported by Bignami and Palladini,⁸ who produced status spongiosus of the cerebral cortex in rats injected intracranially with ouabain. They showed that the EEG in the postictal stage of convulsion in such rats was similar to those seen in human Creutzfeldt-Jakob disease.

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