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Guillain-Barré syndrome after heat stroke

Heat stroke is usually not listed among the events triggering Guillain-Barré syndrome. Two cases of a Guillain-Barré syndrome-like polyneuropathy after heat stroke are on record, although without reference to electroneurography.[1] We report on a patient, who developed Guillain-Barré syndrome 10 days after severe heat stroke. He had electrophysiological evidence of demyelination, increased CSF protein, and high anti-GM1 antibodies. Heat stroke activates the immune system by cytokine release,[2] opens the blood-nerve barrier, and exposes peripheral nerve antigens and thus may induce Guillain-Barré syndrome, as suggested by results from our patient.

A 28 year old drug addict was using anticholinergic drugs against sweating during levomethadone withdrawal. He was found unresponsive in a public garden on a hot summer day (ambient temperature 32°C) after ingesting cocaine. His core temperature was 42.5°C at admission. He was in deep coma with wide unreactive pupils and without corneal and pharyngeal reflexes. Tachypnoea had induced hypocapnia. Blood pressure was 85/30 mm Hg and heart rate was 165/min. He developed disseminated intravascular coagulation, thrombopenia below 10 000 MRD/ml, and metabolic acidosis. Creatine kinase rose from 128 to 751 U/l. Leucocytes and C reactive protein remained normal. After 4 days of coma he was transferred to a closed psychiatry ward because of agitation and frightening hallucinations. After 5 more days he complained of fatigue, myalgia, and arthralgia and, 3 days later, developed fever (38.9°C), tetraplegia, and dysphagia. He required intensive care within hours. Vital capacity was 1.5 l. Proximal arm muscles had MRC grades between 2 and 3. All other limb muscles had grades 1 or 2. Facial muscle weakness increased for 2 more days. There was no ophthalmoplegia, but areflexia and stocking glove hypaesthesia for vibratory and cold stimuli. He did not respond to early intravenous immunoglobulin treatment and underwent plasma exchange from day 14 to 18. Bulbar muscles improved on day 15. Head movements improved 1 week later. Minimal hand functions recurred after 4 more weeks. He still required help with dressing and was unable to stand 14 months after disease onset.

Protein comcentration in CSF was 480 mg/l 2 days after onset of tetraparesis, and 7300 mg/l 2 weeks later. Cell count was 6 cells/mm^3. He had high IgM (500 U/l; enzyme linked immunosorbent assay (ELISA); normal below 120 U/l) but only moderately increased IgA antibodies against GM1. 0nly one of 20 patients with Guillain-Barré syndrome examined in the same laboratory had higher anti-GM1 IgM antibodies. The anti-GM1 antibodies were normal 8 weeks after plasmapheresis.

Compound motor action potentials were<0.7 mV in all tested nerves from day 2 to day 95. Distal latencies were more than 150% above the upper limit of normal in the left peroneal nerve. Conduction velocities were below 70% of the lower limit of normal in the left peroneal and the left median nerve. F latency was above 150% of the upper limit of normal in the left ulnar nerve. F responses were missing in both median nerves and in the right peroneal nerve. A conduction block was present along the right ulnar nerve (wrist stimulation 0.69 mV; plexus stimulation 0.37 mV). Abnormal temporal dispersion and possible conduction block was present in the left ulnar nerve (wrist stimulation amplitude 0.36 mV; duration 8.6 ms; elbow stimulation amplitude 0.19 mV; duration 10.2 ms). Median sensory nerve conduction was normal and sural nerve conduction was moderately slowed (36 m/s) at day 2. Needle EMG disclosed abundant fibrillations and positive sharp waves in proximal and distal limb muscles at day 95.

Decreased sweating due to anticholinergic medication, cocaine induced increased heat production, and high ambient temperature precipitated heat stroke in our patient. Ten days afterwards he developed an acute neuropathy that met clinical and neurophysiological criteria for Guillain-Barré syndrome. Similar time delays have been seen in two other patients with Guillain-Barré syndrome-like neuropathies after heat stroke[1] and in the second of two patients reported as critical illness neuropathy after extreme hyperpyrexia.[3] This patient had increased CSF protein and fasciculations which are unusual in critical illness neuropathy. He may have had Guillain-Barré syndrome as well. Weakness evolved with delay in these four patients with Guillain-Barré syndrome-like neuropathies, whereas it was present immediately after hyperpyrexia in five more patients, who probably did not have Guillain-Barré syndrome. One patient with heat stroke was tetraparetic when he regained consciousness.[4] He had pyramidal and cerebellar signs and persistent atrophic weakness due to axonal or motor neuron loss and no neurophysiological evidence for demyelination. Four of 14 patients with cancer exposed to whole body hyperthermia and chemotherapy complained of weakness immediately after hyperthermia.[5] Their nerve conduction abnormalities are reported as "compatible with scattered demyelination".

Our patient had chronic HCV infection which may be associated with vasculitic neuropathy and cryoglobulinaemia, both absent in our patient. A connection between Guillain-Barré syndrome and non-A non-B hepatitis has been suggested,[6] but the close temporal relation makes heat stroke a more probable cause of the disease in our patient. His high anti-GM1 antibodies suggested immune mediation. Anti-GM1 IgA is increased after *Campylobacter jejuni* infection, whereas IgM dominated in our patient who had no evidence of *Campylobacter jejuni* infection. Heat stroke disrupts the gastrointestinal mucosal wall. Endotoxins enter circulation and stimulate macrophages, which release TNF-α, Il-1, Il-6, and IFN-γ. All these cytokines are raised after heat stroke[2] and open the blood-nerve barrier. This may have exposed the GM1 epitope in our patient. IFN-γ induces Schwann cells to express MHC class II gene product, inviting T cell attack. TNF-α is proinflammatory, myelinotoxic, and increased in Guillain-Barré syndrome.

Guillain-Barré syndrome-like neuropathies have been reported from Saudia Arabia,[1] where heat stroke is common, but they were not noted in connection with epidemic heat stroke in North America.[7] Our patient had all features associated with fatal heat stroke: long lasting coma, shock requiring intravenous catecholamines, metabolic acidosis, and disseminated intravascular coagulation.[7] Guillain-Barré syndrome may occur more often after heat stroke, if more patients survive extreme hyperthermia thanks to intensive care.

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Hydrodynamic performance of a new siphon preventing device: the SiphonGuard

Around 10% to 30% of shunt revisions may be attributed to posture related overdrainage. Of the various siphon preventing devices available at present, two construction types are the most prominent: those using a gravitational mechanism and those using a subcutaneous membrane. Gravitational devices such as Elekta-Cordis Horizontal-Vertical Valve, Chhabra Valve, Fuji Valve, or Miethke Dual-Switch Valve are widely used.[1] Their main drawback is susceptibility to malfunction when the shunt becomes displaced from its vertical axis after implantation and unpredictable operation during persistent bodily movements. The membrane devices: the Anti-Siphon Device (ASD, Heyer Schulte) or Siphon Control Device (SCD, Medtronik PS Medical) have generally proved clinically effective,[2 3] although in some cases these devices may obstruct the CSF drainage when the subcutaneous pressure increases or the scar tissue isolates the device from atmospheric pressure. The flow regulat-