

cortisol rise from 338 to 449 nmol/l over 1 hour). His plasma VLCFA profile was abnormal consistent with XL-ALD. Humphrey visual field testing demonstrated a left homonymous field defect. Brain MRI was abnormal (figure A and B). He was placed on a very low fat diet with supplements of glycerol trioleate oil.

By October 1996 his headaches had settled but his eyesight, memory, coordination, and walking were worse. Visual acuity was below 6/60 in both eyes. Brain MRI was repeated (figure C-E). By August 1997 there had been no new clinical developments (MRI figure F-G). In May 1998 he complained of navigational difficulties in familiar surroundings, further memory loss, and cognitive decline (MRI figure H-I).

In May 1996 (figure A) T2 weighted axial imaging showed high signal intensity areas in the region of the right lateral geniculate nucleus and left optic tract. The occipital white matter was normal. T1 weighted images with gadolinium contrast enhancement (figure B) showed bilateral enhancement of the intracerebral optic tracts.

By October 1996 (figure C) T2 weighted axial imaging showed spread of the areas of high signal intensity continuously from the lateral geniculate nuclei posteriorly along the optic radiations into the white matter of both occipital lobes, more prominent on the right. T1 weighted images showed contrast enhancement in the optic chiasm and optic tracts (figure D), lateral geniculate nuclei, origins of the optic radiations, and right occipital white matter (figure E).

By August 1997 (figure F) there had been further progression in the white matter changes in both occipital lobes, with spread to the splenium of the corpus callosum. Contrast enhancement (figure G) was seen in the optic radiations and right occipital white matter. The cerebellar white matter was of low signal intensity, with a small area of contrast enhancement above the fourth ventricle to the right of the midline.

In May 1998 (figure H and I) the changes in white matter were yet more extensive with the appearance of ring enhancement.

Our case illustrates MRI appearances typical of adrenoleukodystrophy and demonstrates in particular the evolution of these changes with time. The tendency of this condition to affect the visual pathways selectively is well illustrated as is the spread of disease along the fibre tracts of that system. This allowed visualisation of parts of the visual system, the anatomy of which is usually hidden—for example, the intracerebral portion of the optic tracts. The characteristic MRI appearances are thought to result from an advancing front of active demyelination, followed by an area of inflammatory cellular response demonstrating contrast enhancement, surrounded by areas of established damage, gliosis, and neuronal loss.⁷

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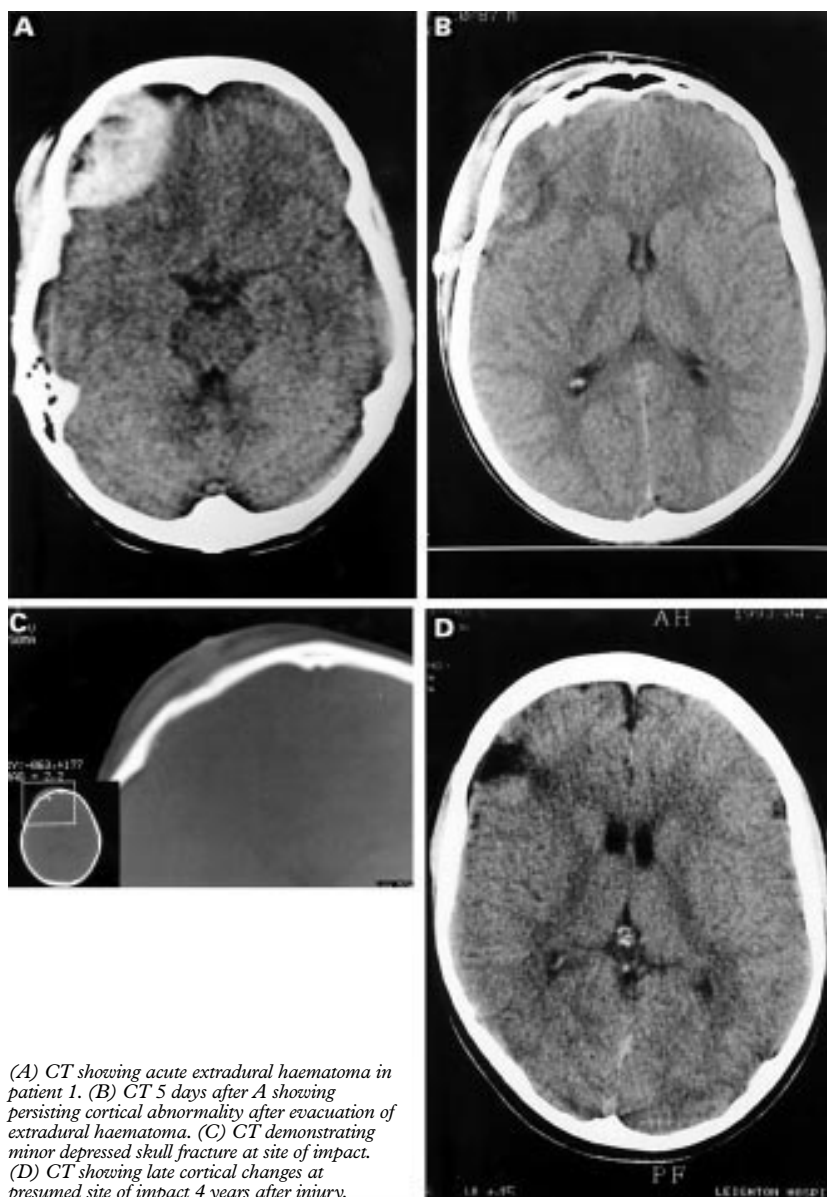
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Golf ball epilepsy

Blunt head injuries may cause epilepsy. We present the cases of four young people whose heads were all subject to contact with golf balls travelling at speed. Each had post-traumatic seizures, three early and one late, despite the apparent absence of post-traumatic amnesia. Although many patients who develop epilepsy recall some type of head injury preceding their first seizure, post-traumatic epilepsy probably accounts for less than 5% of all the epilepsies.¹ There is good

evidence that the risk of post-traumatic epilepsy increases with the severity of the injury. Thus, Jennett² identified the presence of intracranial haemorrhage, dural laceration, and early post-traumatic seizures as the chief risk factors for late post-traumatic epilepsy. Annegers et al also emphasised that in the absence of a post-traumatic amnesia of 30 minutes or greater, there was no significant increased risk for the development of post-traumatic epilepsy.³

From a practical point of view and for medicolegal purposes, it is necessary to decide if a seizure is post-traumatic. As a general rule it may be stated that if the person concerned does not give a history of a post-traumatic amnesia lasting for a significant period of time (an hour or more), and there is no history of a compound or depressed fracture with dural tear, it is reasonable to exclude the possibility that the epilepsy is post-traumatic. However, it is worth noting that this view is based on Jennett's work and precedes CT. There is no good evidence from a large series to indicate whether findings on acute imaging add anything to the prediction of post-traumatic epilepsy.



(A) CT showing acute extradural haematoma in patient 1. (B) CT 5 days after A showing persisting cortical abnormality after evacuation of extradural haematoma. (C) CT demonstrating minor depressed skull fracture at site of impact. (D) CT showing late cortical changes at presumed site of impact 4 years after injury.

Four examples of acute symptomatic seizures and epilepsy developing after head injuries with golf balls are described, which seem to be an exception to these clinical rules.

An 11 year old boy was struck on the right temple by a golf ball resulting in right frontal scalp contusion. His consciousness was not impaired until about 3 hours later when he became drowsy and had two focal motor seizures affecting the left arm. He was intubated and ventilated. A head CT showed a right frontal extradural haematoma with no skull fracture (figure A). The haematoma was evacuated (figure B). He was woken and extubated the next day and was discharged without neurological impairment two days later on phenytoin. No further follow up is yet available.

A 16 year old boy, who was a keen golfer with a single figure handicap, was struck on the head by a golf ball which rebounded several yards after striking him on the forehead. He experienced local pain, bruising, and swelling. Although he was never unconscious, some 4-5 hours later he developed repetitive jerking of the right face and arm. He was taken to his local casualty department where a diagnosis of serial focal motor seizures was made. His consciousness was then somewhat obtunded. A brain CT was performed which showed a small, discrete, spherical intracerebral haematoma in the left frontal lobe immediately beneath the skull at the point where he had been struck (similar in shape, but more hyperintense than the appearance in the figure B). The haematoma gave the distinct impression of a golf ball embedded in the surface of the cerebral hemisphere!

He was treated with parenteral anti-epileptic drugs and subsequently with thiopentone requiring ventilation for 48 hours while he was loaded with phenytoin.

He was maintained on phenytoin for 12 months but subsequently this was withdrawn and he has remained seizure free.

A 5 year old girl was struck on the forehead above the right eye by a golf ball struck 10 metres away. On arrival in the accident and emergency department she was fully alert, orientated, and neurologically intact. A laceration was present but there had been no apparent impairment of consciousness or vomiting. However, 90 minutes after the injury she had a generalised tonic clonic seizure lasting 25 minutes. She was intubated and ventilated. A CT scan showed a very small depressed fracture with minimal haemorrhagic contusion in the cortex of the right frontal lobe (figure C). She was woken and extubated later that day. She has had no further seizures.

A 12 year old boy was practising golf with a friend. He was struck on the front of the head by a golf ball which rebounded a considerable distance after striking him. He did not lose consciousness and had no more than localised pain, tenderness, and bruising at the site of impact. He did not seek any medical advice about the injury. Over the next 4 years he had three well documented tonic-clonic seizures that started during sleep.

A CT scan 3 years after the original injury showed a small, wedge shaped area of low density affecting the cortex close to the point at which he recalls being struck (figure D).

The heads of these four young people were all subject to contact with golf balls which at club level travel at speeds of up to 130 miles/hour. Each had post-traumatic seizures, three early and one late, despite the apparent absence of post-traumatic amnesia.

Patients 1 and 2 would indicate that this kind of injury is capable of transferring energy across the skull, independent of a skull fracture, to cause an acute extradural or cortical haematoma.⁴ In patient 4 the lesion identified at a later date by CT is consistent with the late consequences of a localised intracranial haematoma. It therefore seems reasonable to assume that the late epilepsy in patient 4 was also related to the initial golf ball injury.

In the third patient, a minor depressed fracture and contusion were again associated with an early seizure without evidence of intervening impairment of consciousness.

It does therefore seem that golf ball injuries are capable of giving rise to both acute symptomatic seizures and late epilepsy without causing post-traumatic amnesia, skull fracture or dural tear. CT evidence, however, would predict the possibility of seizures in these examples in whom the development of post-traumatic epilepsy probably results from the physical properties of golf balls and their ability to transmit considerable mechanical energy at a small site of impact. The problem is one of which spectators on golf courses (and their doctors) should be aware.

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Sensory predominant neuropathy with GM₁ antibodies, conduction blocks, and orbital pseudotumour

Two male patients developed multifocal sensory neuropathy with high titre IgM anti-GM₁ antibodies (up to 1: 64 000) and infiltrative orbitopathy. Nerve conduction studies showed multiple motor conduction blocks and evidence of a similar phenomenon in sensory nerves. Both patients deteriorated after corticosteroid administration but benefited substantially from intravenous immunoglobulin therapy. Our findings suggest the existence of a predominantly sensory subtype of multifocal motor neuropathy (MMN) and challenge the postulated motor specificity of anti-GM₁ antibodies.

Anti-GM₁ antibodies have been implicated in the aetiology of multifocal motor neuropathy (MMN) and are assumed to be specific for this disease when occurring at high titres.^{1,2} We report on two patients with high titre IgM anti-GM₁ antibodies and electrophysiological features typical of MMN presenting with severe sensory neuropathy.

Patient 1 was a man who developed asymmetric numbness of limbs and difficulty in performing fine motor movements around the age of 55. Sensory deficits showed a multifocal pattern (multiple mononeuropathy) and involved proximal limb regions, trunk, and

face. The course of illness was steadily deteriorating with some episodes of prominent disease progression usually preceded by minor infections. After 10 years he was unable to write, needed assistance for dressing and walking, and complained of diplopia. Neurological examination showed profound loss of all sensory modalities in the arms and legs and pseudoathetosis of the fingers and wrist. Deep tendon reflexes were preserved and muscle strength was normal. The patient showed marked protrusion and downwards and outwards deviation of the left eye with a complex impairment of all eye movements.

Patient 2, a 68 year old man, reported an insidious onset and gradual worsening of asymmetric sensory loss and painful patchy dysaesthesia. At a neurological examination 15 years after disease onset, pinprick sensation, vibration, and proprioception were prominently affected in his arms and legs. There was severe ataxia of gait. Postural maintenance and voluntary movements strongly depended on visual guidance. Muscle strength was normal except for slight bilateral paresis of the tibialis anterior and intrinsic hand muscles (4+/5 MRC scale). There was no evidence of autonomic nerve or pyramidal tract dysfunction. Tendon reflexes were absent. The patient had an incomplete left side third nerve paresis and ipsilateral visual loss.

General physical examination was normal in both patients, as were repeated screenings for neoplastic and connective tissue diseases. Enzyme linked immunosorbent assays (ELISAs)³ showed IgM antibody activity against GM₁, asialo-GM₁, and GD_{1b}, presumably recognising the Gal(β1-3)GalNAc group (table). Clonality of ganglioside antibodies was not investigated. Serum immunoelectrophoresis did not show monoclonal gammopathy. All the following laboratory indices were normal or negative: creatine phosphokinase, erythrocyte sedimentation rate, renal and liver function, antinuclear antibodies, thyroxine, vitamins B₁₂, B₆, B₁₂, folic acid, urine porphobilinogen, and serum cryoglobulins. Cerebrospinal fluid was acellular with 52 mg/dl and 90 mg/dl protein, respectively (normal <50 mg/dl).

Nerve conduction studies showed multifocal slowing of motor nerve conduction velocities and F wave latencies (table). Despite near normal muscle strength we found motor conduction blocks at sites not prone to compression. Sensory nerve potentials could not be elicited in the median, ulnar, radial, or sural nerves. Electromyography showed fibrillation activity, generalised fasciculations, and features of chronic neurogenic damage. Magnetic resonance imaging of the brain, spinal cord, and dorsal nerve roots did not show relevant abnormalities. Both patients had non-progressive orbital infiltration with slight gadolinium enhancement suggestive of ectopic lymphoproliferative tissue. Lesion extension was most pronounced at the apex orbitae and fissura orbitalis superior and caused compression of the left optic and oculomotor nerves in one patient and mechanical interference with eye movement in the other. Analogous orbital infiltration has been described in patients with paraproteinemic neuropathies and antibody mediated autoimmune diseases such as myasthenia.

Administration of methylprednisolone at dosages of 40 to 60 mg/day was followed by marked deterioration of sensory ataxia in both patients. By contrast, substantial and rapid benefit was achieved by means of intravenous immunoglobulin (IVIg) therapy (0.4