Malignant distress on eye contact after severe head injury

We report a patient with severe head injury who developed a very specific pattern of symptoms of distress on eye contact. It is likely that this neurobehavioural syndrome contributed to the decline of his mental and physical state which started three years after injury.

The patient was described as having been a quiet, private person who enjoyed solitary pursuits. He disliked being in the company of people who were disabled. At the age of 32 he had a severe closed head injury. Brain CT showed left temporal lobe contusions and an extradural haematoma at the base of skull, which was evacuated. He was in a coma for about four months. Two years after injury he was underweight and had severe contractures with no functionally useful movement.

At his best, about three years after injury, he was able to walk with the aid of a frame and assistance and was beginning to be able to feed himself. His most reliable cognitive response was following basic commands. At times he was able to indicate “yes” and “no,” and name objects and colours, but this was mostly unreliable. He had severe dysarthria.

His best verbal output consisted of singing a few words of a well known song. He had never, since the accident, shown any definite recognition of anybody, including his wife.

About two weeks after the injury, he began to show increasing signs of tension and irritability during therapy. It seemed as though he wanted to be left alone. It was noted that “eye contact was reported to provoke anger—noise”.

Since that time he has continued to show a behavioural syndrome consisting of symptoms of distress and agitation in relation to eye contact. Other social cues—for example, entering his room but avoiding eye contact—also cause distress but to a much lesser degree.

Symptoms of distress include wailing, grimming, episodic posturing, and repeated hitting of limbs, with facial engorgement and profuse sweating. These symptoms last for as long as the observer’s gaze is maintained. The symptoms subside within a few seconds of disengagement from eye contact. All three authors, as well as several nurses involved in his care, have independently noted the ability of eye contact to provoke distress. Nursing staff have found that averting their eyes tends to reduce distress. His eye movements lack normal fluency. He tends to fixate on objects, particularly faces and eyes, and has difficulty redirecting his gaze away. As a consequence eye contact tends to be maintained, with a fixed unblinking stare, until the observer looks away.

Specific behaviour therapy aimed at increasing exposure to eye contact was unsuccessful. The symptom was associated with a steady decline in his mental and physical state such that by 1994, seven years after injury, he had to be nursed continuously on his bed. His weight dropped 20 kg to 51 kg. Brain CT in January 1991 showed gross hydrocephalus with periventricular lucency but insertion of a ventriculoperitoneal CSF shunt at that time had no effect on his mental state. Over the past year combined treatment with trifluoperazine and moclobemide have been associated with a slight improvement in his mental and physical state.

To test the hypothesis that the distress was indeed related specifically to eye contact, rather than some other aspect of a person’s face or simply the presence of human contact, we carried out the following brief experiment. In a state of rest, lying awake in his recliner with his eyes open, the patient was approached by an unfamiliar member of staff who stood almost facing him about three feet away. In one condition the staff member looked directly at the patient, and thereby invariably made eye contact; in the other condition the staff member faced the patient at the same angle, but with eyes averted by about 30° from the patient. An observer, hidden from view, noted the time taken to become agitated (defined as banging his arm on the chair). The rate of eye blinking during the two conditions was also noted. The testing condition was terminated by the person walking away as soon as the patient became agitated, or after 10 minutes if there was no sign of agitation by this time. A total of 12 sessions were studied, six of each condition, carried out on three separate days. The order of testing was counter balanced.

The mean time to agitation was very significantly shorter in the eye contact condition than in the eyes averted condition (mean 93 (SD 114) s vs 510 (SD 221) s; P = 0.006; figure). The rate of eye blink was significantly slower in the eye contact condition (0.67 (SD 0.78) vs 1.87 (SD 0.42) blinks/min; P = 0.034).

Our patient showed two problems. One was distress on eye contact. The other was an apparent inability to disengage his eyes from eye contact.

It is known that eye contact may be associated with arousal and this link seems to develop early in life. In animals directed gaze at another may be used as a form of threat (Rudyard Kipling’s Mowgli “discovered that if he stared hard at any wolf, the wolf would look away”). The tendency to gaze aversion in monkeys develops during the first week of life. Cells in the superior temporal sulcus of the monkey are specifically tuned to detect eye contact or averted gaze. In humans various studies have shown that eye contact increases arousal. Hutt and
Arrested progression of the cauda equina syndrome of ankylosing spondylitis after lumbarperitoneal shunting

An idiopathic cauda equina syndrome is a rare but well recognised complication of longstanding ankylosing spondylitis, usually developing many years after disease onset, and after cessation of active pathology. The frequency of this complication is unknown, but it is probably higher than the paucity of cases reported might suggest—for example, Thomas and colleagues found two cases among 45 patients with ankylosing spondylitis seen at this centre. The characteristic pathological findings include erosion of the vertebral pedicles, lamination, and the superior articular processes (“vertical scalloping”), widening of the thecal sac, and the presence of multiple dorsal arachnoid diverticula, but direct compression of the nerve roots is very uncommon. Affected nerve roots may show fibrosis and lost myelin. Clinically, the differential diagnosis includes compressive lesions of the cauda equina—for example, tumours (especially in patients with ankylosing spondylitis previously treated with spinal radiotherapy) but CT/MRI establishes the diagnosis.

Neither the natural history nor the pathogenesis of this condition are well defined. In the largest series reported to date (14 patients) recovery was slowly but relentlessly progressive without substantial remission or resolution. The series was reported by Balint’s in 1935. In a subsequent series of 49 patients, and the investigation of 49 cases reported in the medical literature before 1990 concluded that 31 patients showed a progressive course, nine followed a stable course, and follow up was insufficient to permit meaningful conclusions. Of these 49, 47 eventually developed lumbar sacral sensory disturbance, 44 sphincter disturbance, 27 motor deficits, and 23 pain. Therapeutic interventions to try to arrest or reverse the course of the syndrome, for example, including the use of steroids, non-steroidal anti-inflammatory drugs, and surgical intervention, have produced disappointing results.

A 56 year old man with a 32 year history of ankylosing spondylitis presented with sensory disturbance in the right leg. Aside from developing the typical bodily habitus of ankylosing spondylitis and having recurrent attacks of iritis in the left eye with subsequent cataract formation, his disease had caused him few problems. He took no regular medication and was able to walk a distance of several miles without difficulty. Radiographs of his dorsolumbar spine showed fusion of sacrococcygeal joints, squaring of the vertebræ and syndesmophyte formation, and ossification of interspinous ligaments and apophyseal joints, appearances typical of the "bamboo spine" of ankylosing spondylitis. Tissue typing was positive for HLA antigen B27.

Four years before presentation he noticed persistent numbness over the lateral border of the right foot, and seven weeks before presentation numbness over the right buttock. Clinically, all sensory modalities were impaired in the right L5, S1, S2, and S3 dermatomes, and the right ankle jerk was lost, but power was preserved. His left leg was normal. Electrophysiological studies failed to detect somatosensory responses from the right S1 dermatome and the right posterior tibial nerve. Some loss of motor fibres in the right lumbar sacral distribution was indicated by delayed and abnormal F waves present via the right posterior tibial nerve in the foot and low amplitude evoked muscle action potentials in the abductor hallucis on stimulation of the posterior tibial nerve. Neurogenic abnormalities were found in the EMG of the right biceps femoris. Lumbar spine MRI showed erosion of the posterior lumbar arches with a wide and capacious spinal canal, in which multiple dorsal arachnoid diverticula were seen (figure). These electrophysiological and MRI findings are characteristic of the cauda equina syndrome of ankylosing spondylitis.

Further clinical deterioration occurred during follow up, with extension of the area of sensory impairment in the right leg to involve S4, sensory symptoms affecting the left buttock, and loss of vibration sense to the left ankle; the left plantar became extensor. These symptoms and signs were not influenced by an epidural steroid injection. In view of the progressive nature of the patient's neurological deficit and the excessive spinal subarachnoid space, the relatively simple procedure of lumboperitoneal shunting was considered. This was carried out under general anaesthesia without complication. Cerebrospinal fluid taken at operation showed a normal cell count and protein concentration.

During 36 months of postoperative follow up, the patient's neurological symptoms and signs have remained unchanged. There has been no recovery of the function lost before operation, but no new neurological deficit has developed. Electrophysiological studies and lumbar spine MRI are also unchanged. Although only limited information is available concerning the natural history of the cauda equina syndrome of ankylosing spondylitis, it seems to follow a slow but relentless progression in most cases, until complete sacral anaesthesia with impaired sphincter function is reached. Intermittent and spontaneous periods of stabilisation do not seem to occur. Hence, a 36 month period of neurological stability, as seen in our patient, would seem exceptional, the more so in view of his continuing deterioration before operation. We therefore think that lumboperitoneal shunting has at worst stabilised his neurological deficit for a time and at best arrested its progression.