Ounsted suggested that the gaze aversion of autistic children served to reduce arousal. Very little is known about control of eye movement in patients with head injuries. Our patient’s difficulty in disengaging from eye contact may be regarded as a form of the locking of fixation found in patients with Balint’s syndrome.1,2 He has suggested that the act of shifting visual attention requires three operations; disengagement of attention from its current focus, followed by movement of attention to the target, and finally engagement of attention on the target. The parietal lobe is specifically involved in disengagement of attention. Our patient seems to have lost control of this disengagement process.

Two processes seem to come together to result in our patient’s symptom. On the one hand brain damage resulted in disinhibition of relatively primitive neurobehavioural responses to gaze in which the gaze of another on oneself is regarded as a threatening stimulus. This will have been exacerbated by neural mechanisms which result in selective fixation on eyes; babies show preferential fixation on eyes by the second or third weeks of age.1 Thus the patient preferentially fixated on eyes, from which he had problems disengaging despite the profound arousal that he experienced on eye contact. On the other hand his premorbid socially avoidant personality traits and aversion to disability will have facilitated the distress he felt when he was self consciously aware that others were aware of his disability. The most potent cue for such self consciousness is eye contact. The development of increasing self awareness on recovery from his head injury was therefore necessary for the fully developed bilateral cortical syndrome and may partly explain why the symptom did not develop until three years after his head injury.

S FLEMING
Department of Psychological Medicine, St Bartholomew’s and the Royal London School of Medicine and Dentistry, London E1 2AD, UK
L MURPHY
Royal Hospital for Neuro-disability, Putney, West Hill, London SW15 3SP, UK
WA LISHMAN
Institute of Psychiatry, De Crespigny Park, London SE5, UK

Correspondence to: Dr Simon Fleming, Department of Psychological Medicine, St Bartholomew’s and the Royal London School of Medicine and Dentistry, Turner Street, London E1 2AD, UK

2 Perrett DI, Smith FAJ, Potter DD, et al. Visual cells in the posterior parietal cortex sensitive to face view and gaze direction. Proc R Soc Lond B Very Little is known about control of eye movement in patients with head injuries. Our patient’s difficulty in disengaging from eye contact may be regarded as a form of the locking of fixation found in patients with Balint’s syndrome.1,2 He has suggested that the act of shifting visual attention requires three operations; disengagement of attention from its current focus, followed by movement of attention to the target, and finally engagement of attention on the target. The parietal lobe is specifically involved in disengagement of attention. Our patient seems to have lost control of this disengagement process.

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S FLEMING
Department of Psychological Medicine, St Bartholomew’s and the Royal London School of Medicine and Dentistry, Turner Street, London E1 2AD, UK

Letter to the Editor

Arrested progression of the cauda equina syndrome of ankylosing spondylitis

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.1 The frequency of this complication is unknown, but it is probably higher than the pauciety of cases reported might suggest—for example, Thomas and colleagues found two cases among 45 patients with ankylosing spondylitis seen at this centre.1 The characteristic pathological findings include erosion of the vertebral pedicles, laminas, and parts of the sacrum (“ventral scalping”), 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.2 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), progression was slowly but relentlessly progressive without stabilisation or remission until complete sacral anaesthesia had developed, usually with double incontinence. The case 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 a conclusion in the remaining nine. Of these, 49, 47 eventually developed lumbar sacral sensory disturbance, 44 sphincter disturbance, 27 motor deficits, and 23 pain.2 Therapeutic interventions to try to arrest or reverse the lesion of the syndrome, 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 discomfort. Radiographs of his dorsolumbar spine showed fusion of sacroiliac joints, squaring of the vertebral bodies and syndesmophyte formation, and ossification of intervertebral 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 LS, 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 conducted via the related 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.1,3

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.1 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.
The pathogenesis of the cauda equina syndrome of ankylosing spondylitis is unknown. The long duration between the onset of ankylosing spondylitis and neurological symptoms (average 35 years in the Mayo Clinic series) argues against a shared inflammatory process as the cause of the cauda equina syndrome. Other conditions, such as trauma, infections, tumors, and previous surgery, may also be responsible. One long-term patient with a complication of ankylosing spondylitis has described painful, numbness, and paresthesia of the lower limbs, which became worse with standing or walking. This patient's symptoms were consistent with the diagnosis of cauda equina syndrome. However, the patient's symptoms improved with treatment, including physical therapy and medications, and did not recur after 1 year of follow-up.

Diffuse neurofibriobillary tangles with calcification in a non-demented woman

In 1994 we proposed the term “diffuse neurofibrillary tangles with calcification” (DNTC) for a new form of presenile dementia. This disease is clinically characterised by progressive cortical dementia. Neuropathological features consist of temporal atrophy and loss of myelinated axons, with rare neurofibrillary tangles, astrocytes, and calcifications throughout the brain. We have recently described a case of DNTC with calcification. The patient was a 78-year-old woman with a history of hypertension, diabetes mellitus, and hypercholesterolemia. She was admitted to the hospital with symptoms of memory loss, forgetfulness, and difficulty in performing daily tasks. MRI of the brain showed symmetrical atrophy of the temporal lobes, with a decrease in the T2 signal intensity in the bilateral entorhinal cortices. EEG showed no epileptic activity. The patient's clinical course was characterized by a gradual decline in cognitive function, with worsening of memory impairment and difficulty in performing daily tasks. She was diagnosed with DNTC with calcification, and no other cause of cognitive impairment was found.

Neurology of adult α-mannosidosis

Neurological findings in adult α-mannosidosis are rare. In a review of the literature, we found only a few case reports of patients with this condition. The most common neurological manifestations are motor and sensory impairment, such as muscle weakness, ataxia, and hyporeflexia. The disease is associated with a progressive decline in cognitive function, with symptoms of memory loss, forgetfulness, and difficulty in performing daily tasks. EEG showed no epileptic activity. The patient's clinical course was characterized by a gradual decline in cognitive function, with worsening of memory impairment and difficulty in performing daily tasks. She was diagnosed with DNTC with calcification, and no other cause of cognitive impairment was found.

Thanks are due to Dr Tony Finnegan, Barts Hospital, for his helpful comments and suggestions.

Correspondence to: Dr AJ Lamer, National Hospital for Neurology and Neurosurgery, Queen Square, London WC1 3BG, UK.