Short report

Pain in motor neuron disease

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SUMMARY Twenty-seven of 42 patients with motor neuron disease had significant pain. The nature and duration of the pain are described along with an illustrative case-report. The aetiology and most effective treatment of this common complication of motor neuron disease remain unclear.

Pain is a serious complication of motor neuron disease. References to pain in the published literature are few though one account details the occasional occurrence of sensory hypersensitivity, neuralgia, joint pains, painful muscle cramps and frozen shoulder. A survey of hospice experience found 40% of patients to have had pain in the terminal phases. Our recent experience with motor neuron disease patients' suggested pain is far more frequent than usually recognised, with up to 64% having persistent symptoms. This report describes the frequency, nature and management of pain in motor neuron disease.

Patients and methods

Using data from our recent study of 42 motor neuron disease patients we reviewed our records of those with persistent pain of more than trivial severity. We sought details of the pain duration, timing in the course of the disease, pain quality, site and efficacy of treatment measures.

Results

Pain was a major symptom in 27 patients (64%), 10 of whom were completely independent for all self-care activities. Three patients presented with pain at the time of diagnosis. The table shows further details. Pain was felt principally in a single site by 17 patients (the legs in seven, miscellaneous sites in five, shoulders in three and the back in two). Ten patients experienced pain in more than one site (the neck, shoulders and back in eight, and the back and limbs in two). Pain was described as predominantly aching (14 cases), cramping (eight cases), or burning, shock-like or indescribable (five cases). Some patients described more than one type of pain. All patients had tried drug treatment. This was successful or partially helpful in 15; all these patients rated their pain as mild. Nine patients had moderate pain and three complained of pain severe enough to dominate their lives. Of these latter 12 patients, eight had tried many prescribed and proprietary analgesics as well as a variety of non-steroidal anti-inflammatory drugs, but had abandoned their regular use. This was done because of ineffectiveness (6), side-effects (1) or spontaneous improvement in severity (1). All of the eight found that symptoms were worse at night and one man used to cry at night with the pain in his hips.

The following case report is an example of the problems for patient and doctor that pain in motor neuron disease can bring.

Case Report

A white male aged 59 years presented with 18 months generalised weakness and cramp-like pain in the neck, lower back and legs. Examination revealed a spastic paraparesis but myelography did not show cord compression. When seen 3 months later he was having frequent extensor

Table 1 Patients with significant pain. N = 27

<table>
<thead>
<tr>
<th></th>
<th>Male (19)</th>
<th>Female (8)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Mean age-yr (ISE)</td>
<td>60 (3)</td>
<td>67 (2)</td>
</tr>
<tr>
<td>Mean duration of disease-mth (ISE)</td>
<td>43 (10)</td>
<td>39 (9)</td>
</tr>
<tr>
<td>Mean duration of pain-mth (ISE)</td>
<td>18 (6)</td>
<td>12 (5)</td>
</tr>
<tr>
<td>Mean interval between onset of motor neuron disease and start of pain-mth (ISE)</td>
<td>32 (9)</td>
<td>26 (5)</td>
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</tbody>
</table>
Pain in motor neuron disease

spasms and persistent pain in the back and all four limbs. Trials of lioresal, dantrolene and diazepam partially ameliorated the spasticity and spasms but had no effect on the pain which he variously described as tight and aching but also had an unpleasant burning quality which varied unpredictably. At this stage he had evidence of a spastic tetraparesis, dysarthria, wasting of intrinsic hand muscles and fasciculation but no sensory abnormalities. Electrophysiological studies demonstrated fibrillation, reduced numbers of motor units and some giant motor unit potentials in all four limbs. The diagnosis of motor neuron disease was confirmed by the subsequent course with the development of widespread muscle wasting and progressive bulbar palsy. Pain control was not achieved with the use of different anti-inflammatory or codeine compounds nor with anti-depressants or tranquillisers. He was now sleeping in his wheelchair because of night pain and there was considerable marital discord, exacerbated by his demanding behaviour. This appeared to stem from anxiety that his wife would be unable to cope with his increasing dependency. Further investigations showed normal routine tests including blood count and viscosity. The serum creatine kinase and calcium levels were normal as was an isotope bone scan. Plain radiographs of spine showed minor degenerative changes only and a further myelogram did not show root compression or arachnoiditis. An inpatient trial of prednisolone (30 mg daily for 2 weeks) had no effect and injection of local anaesthetic and steroids into tender trigger points in spastic extensor muscles in the back gave improvement for 12 hours only. Transcutaneous nerve stimulation did not help. Mechanical measures such as a change of cushion or wheelchair backrest, and regular change of position afforded slight relief but 5 years after symptoms began he remains in moderately severe chronic pain.

Discussion

Pain is a major symptomatic complication of motor neuron disease. This is not widely recognised and certainly no clear account of its nature and management is to be found in standard texts such as that by Walton.4

The aetiology of the pain is not definitely known. Pain may be part of the sensory phenomena noted by up to 11% of patients.5 Charcot6 stated that it was “not unusual for patients to complain of paraesthesias, sensations of pulling, burning, prickling and formication with vague painful discomfort either spontaneous or provoked”. Three of our patients had subjective sensory disturbance and pain long before serious disability occurred, and this pattern has been noted by other authors.7,8 Degenerative changes in sensory elements of spinal cord9 and peripheral nerve10 have been infrequently described in necropsies of motor neuron disease patients and may be relevant to symptoms. It is possible that this can occasionally give rise to a specifically “neurological” cause of pain associated with motor neuron disease and there is a case report11 of chronic pain in a female patient in which the author speculates that degenerative change in sensory pathways might cause modulation of normal pain transmission. Our case has some similar features although much of the cramping experienced was clearly related to gross spasticity. Unfortunately, control of this aspect did not give control of the pain.

Pain may also stem from abnormal stresses on the musculo-skeletal system imposed by weak musculature. It is particularly relevant in this respect that most of the patients developed pain in the later stages of the disease at the time when they were largely immobile and unable to change the mechanical stresses on their bodies by change of position. Saunders et al2 found 40% of patients in the late phases of the disease complained of pain and this appeared to arise from stiff joints, muscle cramp or skin pressure. Three of our patients had frozen shoulders and this is certainly a risk for anyone who needs lifting. The frequency of night pain exacerbation is probably related to lack of ability to change position, and an electrical patient-operated turning bed could be of help in this respect. There is likely also to be a psychological element to any such chronic pain, particularly if there is co-existing depression, anxiety or opportunity for manipulation or gain.

Unrelieved pain is a distressing and debilitating phenomenon. Management is not easy and may require the services of doctor, physiotherapist, pain clinic and psychiatrist. Intervention may take the form of reassurance, simple analgesics and non steroidal anti-inflammatory drugs, muscle relaxants, physiotherapy with mechanical measures such as passive movement and heat or specialised techniques of nerve blockade and stimulation. Our experience suggests that anything more than mild pain is resistant to complete control. Much attention should be given to prevention. Early education of the family about lifting methods, regular passive movements, and frequent changes of position should be given priority. These manoeuvres may avoid some of the worst symptoms and prevent the development of the chronic intractable pain state. If required, conventional analgesics, anti-inflammatory and anti-spasticity agents may be tried, in sequence or in combination, in the knowledge that a proportion of patients will benefit (55% in our series). For those who remain troubled by pain, there would seem a strong argument for giving opiates on a regular basis, as in malignant disease. The effectiveness of this approach, later in the course of the disease, has been described by Saunders.12 Particular care should be taken in cases with bulbar involvement; depression of respiration and coughing.
are of obvious clinical relevance. It becomes an ethical decision for the individual clinician as to whether or not opiates should be offered to a patient with bulbar problems whose life is significantly marred by pain. It seems likely that, after discussion, many of the most severely affected patients would accept the risk and be grateful for the relief of pain.

References


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_J Neurol Neurosurg Psychiatry_ 1985 48: 838-840
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