SURGICAL TREATMENT OF CERVICAL SPONDYLOTIC MYELOPATHY COMPLICATING ATHETOID CEREBRAL PALSY

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Operative treatment was performed in nine patients with cervical spondylotic myelopathy complicating athetoid cerebral palsy. The first two patients were treated by laminectomy, and the other seven by anterior interbody fusion. The symptoms in both the laminectomy patients improved after operation, but became worse again when cervical instability developed; they then had to have an anterior fusion in addition. In six of the seven patients who had primary anterior fusion a halo-cast (or a halo-vest) was used to keep the cervical spine immobile, and good bony fusion was obtained with satisfactory results. However, in one patient no halo apparatus was used, bony union did not occur and the radiculopathy reappeared.

In cervical myelopathy complicating athetoid cerebral palsy laminectomy is contra-indicated; anterior fusion combined with a halo apparatus is, however, satisfactory.

The degenerative changes of cervical spondylotic myelopathy are commonly observed in old people. But with repeated neck movements from childhood, as may occur with athetoid cerebral palsy, spondylotic changes are often noted even in the young. If the spondylotic is complicated by cervical myelopathy, the athetoid movements may aggravate the myelopathy; conservative treatment is then often ineffective, and operation is sometimes necessary. Such treatment has been reported by several authors, but each has described only one or two cases, with follow-up periods too short for definite conclusions to be drawn.

The purpose of this paper is to report the results of our surgical treatment, and to emphasise the importance of using appropriate techniques and of immobilising the neck after operation.

PATIENTS AND METHODS

Nine patients with athetoid cerebral palsy complicated by cervical spondylotic myelopathy were treated surgically from 1967 to 1982 at Okayama University Hospital, Okayama Rosai Hospital and Ryuso Orthopaedic Hospital. There were six men and three women; their ages ranged from 19 to 52 years (average 34.8 years). The follow-up period was 1 to 12 years (average 5.4 years).

Two patients in the early period were treated by cervical laminectomy, but both developed cervical instability after the operation, and anterior interbody fusion was subsequently performed. The remaining seven patients were treated by anterior interbody fusion as the primary procedure; in six of them a halo-cast (or a halo-vest) was applied about one week before the operation, and retained for three months afterwards, in order to immobilise the cervical spine. Robinson's technique (Smith and Robinson 1958) was used for all the anterior interbody fusions.

The symptoms of cervical spondylotic myelopathy complicating athetosis are not very different from those without athetosis, namely pain in the neck and upper limbs, restriction of cervical movement, sensory disturbance of all four limbs, muscle weakness, increased tendon reflexes, disturbance of delicate movements of the fingers, gait disturbance and dysuria; these are all in addition to the pre-existing athetosis. Applying compression to the neck is a useful diagnostic test.

Each patient had, in addition to plain radiographs, pre-operative myelography with iophendylate or metrizamide. The site of operation was determined by the neurological and radiological findings. The clinical data are shown in Table I.

Radiographic findings. In seven of the nine patients radiographs of the cervical vertebral bodies showed obvious deformation. The most deformed bodies were those of C3, C4 and C5, which characteristically showed flattening of the anterosuperior margin of the bodies and bead-like projections of the antero-inferior margins (see Figs 1 and 2). Intervertebral disc degeneration, as shown
by narrowing of the disc space, hypermobility between adjacent vertebrae and spur formation, were seen most often at C3-4 and C4-5; and the level showing an obvious block on myelography was also at C3-4 and C4-5 in most cases (Table II). Accordingly these were the levels at which operation was most often performed.

Table II. The number of instances at each level where degenerate discs and myelographic blocks were seen

<table>
<thead>
<tr>
<th>C2-3</th>
<th>C3-4</th>
<th>C4-5</th>
<th>C5-6</th>
<th>C6-7</th>
</tr>
</thead>
<tbody>
<tr>
<td>Number of degenerate discs</td>
<td>2</td>
<td>6</td>
<td>7</td>
<td>5</td>
</tr>
<tr>
<td>Number of blocks on myelography</td>
<td>1</td>
<td>7</td>
<td>7</td>
<td>2</td>
</tr>
</tbody>
</table>

RESULTS

Two patients (Cases 7 and 9) were unable to walk from birth, but could use their upper limbs well. Anterior interbody fusion was performed in one (Case 7) because motor disturbance of the upper limbs and intolerable brachialgia developed; and in the other (Case 9), because of motor disturbance of the upper limbs and dysuria. After operation Case 7 lost his brachialgia and noticed slight improvement in the motor function of the fingers in both hands; while in Case 9 motor function of the upper limbs and urination both improved.

Five patients could walk independently in spite of athetosis, until the onset of myelopathy. Then one (Case 1) could walk only with support, while the other four (Cases 2, 3, 4 and 8) became unable to walk. Cases 1 and 2 each had a laminectomy, after which both improved and were able once more to walk independently. However, the symptoms became worse again, and walking became impossible after six years in Case 1 and after one year in Case 2. Radiological examination showed instability in both these laminectomy patients, and anterior interbody fusion was added. After this second operation, Case 1 could walk, but in Case 2 improvement was only temporary. The other three patients had primary anterior interbody fusions; two (Cases 3 and 4) became able to walk independently, and one (Case 8) needed only one stick.

In the two remaining patients (Cases 5 and 6) the main symptoms of myelopathy were muscle wasting and weakness of one upper limb, while the lower limbs were only slightly affected. They had primary anterior interbody fusions and after operation had some recovery of power.

Of the seven patients who had primary anterior interbody fusions, in six a halo apparatus was used; in them the grafted bone crushed, but good bony fusion was obtained. The one patient in whom no halo apparatus
was used (Case 3) failed to unite because the graft absorbed; in him the radiculopathy recurred, though there was some improvement in the disturbance of gait.

Complications of the halo apparatus included breakage of a traction rod in one patient (Case 5) and, in two (Cases 4 and 5) hypermobility in the disc space immediately below the fusion; this developed several years after the operation, and was accompanied by pain which, however, improved after cervical traction.

ILLUSTRATIVE CASE REPORTS

Case 1. This 22-year-old woman, despite her congenital athetosis, could walk independently and do housework. One year before admission she developed pain in the neck, numbness of all four limbs and the trunk, and an abnormal gait. When admitted in 1971, she could hardly walk and had severe disturbance of fine movements of the fingers of both hands. Neurological examination revealed, in addition to athetoid movements, increased tendon reflexes and decreased sensibility of all four limbs as well as slight dysuria. Radiographs of the cervical spine showed marked degenerative changes and myelographic blocks at C3-4, C4-5 and C5-6. A laminectomy was performed from C3 to C5, and after operation she was able to walk and had improved sensation and urination. Numbness and muscular weakness appeared in the left upper limb four years after operation. After childbirth in 1977, she became unable to walk and was readmitted. Radiography then revealed marked instability at the site of her laminectomy, and myelography showed blocks at C5-6 and C6-7. Anterior interbody fusion was performed at these two levels; after this operation her symptoms improved and she was again able to walk. X-ray films taken in January 1983 show marked deformation of the cervical vertebrae; spontaneous union and growth of new bone are seen and the spine seems reasonably stable (Figs 1 to 5).

Case 3. This 29-year-old man had been athetoid since birth. He could walk and look after himself, but not work. Left brachialgia and numbness of all four limbs developed seven months before his admission in 1967, and he had become unable to walk. Sensation of the limbs was disturbed, and the tendon reflexes increased. Radiologically the cervical
vertebrae showed hardly any degenerative changes (Fig. 6), but the myelogram (Fig. 7) showed a marked block at C4-5. An anterior interbody fusion was performed at C4-5, but no halo apparatus was used. After the operation he became able to walk, and the sensory disturbance and brachialgia decreased, but the bone graft crushed and absorbed with failure of union, and the brachialgia reappeared (Figs 8 and 9).

Case 5. This 36-year-old woman had been athetoid since birth, but looked after her house and two children. Pain in the neck developed four years before her admission, and weakness of the right arm two years later. Her symptoms gradually progressed and she could not lift her right arm by the time she was admitted in October 1978. Neurological examination revealed, in addition to athetosis, sensory diminution of the right C5 dermatome, marked weakness and atrophy of the right deltoid muscle, slight weakness of the right biceps and increased tendon reflexes in all four limbs. The X-ray films (Fig. 10) showed irregularity of several vertebral bodies and degeneration of intervertebral discs, and the myelogram (Fig. 11) showed blocks at C3-4 and C4-5. An anterior interbody fusion was performed at these two levels; a halo-cast had been applied nine days before operation and was retained for three months afterwards. The traction rod broke two months after the operation, but the immobilisation was not interrupted. After the operation the graft at C4-5 crushed (Fig. 12), but good fusion was obtained; muscle power improved but the deltoid remained atrophied. Pain in the neck reappeared four years later, but improved after cervical traction. Radiographs taken in April 1983 show instability at C5-6 (Fig. 13).

Case 7. This 49-year-old man had been athetoid since birth. He could not walk, but could use his hands well, and was independent in the activities of daily living. Disturbance of fine movement and numbness appeared in both hands eight months before admission, by which time he could hardly use his hands, and complained of lightning pains from the chest to the abdomen as well as intolerable brachialgia on both sides. Neurological examination revealed severe athetoid movements, diminished sensation below the C5 dermatome, and marked weakness of all four limbs. The radial and triceps reflexes in both upper limbs and the tendon reflexes in both lower limbs were increased. Plain films of the cervical vertebrae showed degeneration at C5-6 and the myelogram showed a partial block at C3-4. For three weeks after admission cervical traction was given, but this was ineffective, and anterior interbody fusions were therefore performed at C3-4 and C4-5. A halo-cast had been applied nine days before the operation, and was retained for three months afterwards. After the operation the graft at C4-5 crushed, but union was good, the brachialgia disappeared, and he again became able to feed himself.

DISCUSSION

In 1956 Freiman and Luwisch described two patients with dystonia complicated by muscular atrophy, one of whom showed marked degenerative changes in the cervical spine. Since then several reports on cervical spondylosis complicating dyskinesia have appeared. In 1962 Anderson et al. reported two patients with cervical spondylotic myelopathy complicating atetosis. In 1970 Levine et al. performed a laminectomy in one patient with myelopathy complicating athetoid cerebral palsy, but no recovery occurred. In 1975 Nakahara et al. studied 66 adults with cerebral palsy, including four who had been operated upon; these four were similar to our Cases 1 to 4. They found, first, that those with spondylotic changes in the cervical vertebrae were mostly athetoid; secondly, that degeneration of the intervertebral disc was most common at C4-5, then at C3-4 and C5-6 in that order; and thirdly that spondylotic changes, though more frequent in old people, were by no means rare in the young. In addition, several authors in Japan reported operations on patients with myelopathy complicating athetoid cerebral palsy (Tezuka et al. 1978; Ooba et al. 1980; Ota et al. 1981; Miyamoto et al. 1982; Yoshikado 1982). However, each of these reports included only one or two cases, the follow-up periods were less than three years and definite conclusions could not be drawn as to the best operative technique or the importance of postoperative fixation. We have operated on nine patients including five with a follow-up of more than five years.

Cervical spondylotic myelopathy is most common in people in their fifties and the usual site is C5-6 (Clarke and Robinson 1956; Sakate 1973). However, in our series the average age was less (34.8 years), and four patients were under 30. The most frequently affected sites (C3-4 and C4-5) were also slightly higher than usual. These results are in accord with those of Nakahara et al. (1975).

It is suggested that athetoid movements accelerate
cervical degeneration. The difference in affected levels between athetoid and other cases seems attributable to the different patterns of movement of the cervical vertebrae. According to White and Panjabi (1978), if the atlanto-axial joint is excluded, flexion and extension of the cervical vertebrae is greatest at C5–6, while lateral bending and axial rotation are greater at C3–4 and C4–5 than at C5–6. In athetosis characterised by writhing movements, the ratio of lateral bending and axial rotation to flexion and extension is assumed to be greater than in normal adults, and this may be why C3–4 and C4–5 are more often spondylotic.

In patients with athetosis, laminectomy of the cervical vertebrae is contra-indicated, as it causes late instability. Anterior interbody fusion, however, resulted in partial recovery from the myelopathy. On the whole the operative results were good, probably because anterior interbody fusion not only removed the degenerated discs which were compressing the spinal cord, but also because fusion ensured stability of the cervical vertebrae.

Anterior cervical fusion in athetosis has two problems. One is the need for an apparatus to immobilise the neck after operation; particularly strong external fixation is needed because of the athetosis and a halo apparatus seems the best (Hartman, Palumbo and Hill 1975; Johnson et al. 1977). Moreover, the complications of the halo apparatus are relatively trivial, and it is well tolerated by athetoid patients.

The second problem is the recurrence of symptoms induced by increased degeneration of the intervertebral discs adjacent to the fused vertebrae. With athetosis, considerable stresses and strains are applied to these discs and therefore the likelihood of recurrence of symptoms is greater than with non-athetoid patients. In two of our cases hypermobility appeared in the intervertebral disc space immediately below the fused vertebrae, causing pain in the neck. Cervical traction has been successful so far, but the problem remains.

In conclusion, for patients with cervical spondylotic myelopathy complicating athetoid cerebral palsy, anterior interbody fusion together with halo immobilisation, while not necessarily the best treatment, is nevertheless very effective, and worth trying if conservative treatment is unsuccessful.

REFERENCES