Surgical relief of progressive upper limb paralysis in Arnold-Chiari malformation

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SUMMARY Two cases of delayed progressive paralysis of the upper limbs in an adult and a teenage patient, without neurological deficits in other regions of the body, are presented. In both cases, the pathology appeared to be a traction lesion of the middle cervical and lower cervical nerve roots, due to abnormal angulation of the nerve roots, which first ran up and then downward in the neural foramina and canal. Re-routing of the nerve roots by removing part of the floor of the neural canal, or by a facetectomy, appeared to offer extensive improvement or full recovery.

The many pathological manifestations of the Arnold-Chiari malformation, or cerebello-medullary ectopia, and associated syringomyelia or hydromyelia have been extensively described. Particularly notable are the reviews of List (1941), Logue (1971), and Barnett *et al.* (1973). Submerged in the large clinical complex is a distinct entity which is quite often mentioned but usually rather lightly passed over—the gradual onset of paralysis in the upper limbs accompanied by sensory deficits. This particular abnormality deserves special mention because it is very distinct, and apparently curable, although up to the present, no useful treatment has been described.

The condition has a delayed onset, occurring either in late childhood or at any time in adult life. It is manifested by increasing paralysis in the upper limbs accompanied by sensory deficits. It occurs in people who, although they have the Arnold-Chiari malformation, are otherwise neurologically intact, and have no decompensated hydrocephalus or increased intracranial pressure. The myelogram shows clear anatomical evidence of cerebello-medullary ectopia, namely downward displacement of the cervical cord, hydromyelia, and an upward running course of the cervical nerves as depicted in an illustration from Chiari's paper quoted by Carmel and Markesbery (1972).

The attempted treatment of this type of progressive paresis in the upper limbs was described by Hall *et al.* (1975). In case 5 of their series, they

describe a 12 year old boy who noted progressive weakness and wasting of his right hand in the C8 to T1 distribution. A myelogram revealed marked enlargement of the cervical cord from C3 to C7 segments. Over a period of months, the symptoms became more severe on the right, and he also developed clumsiness in his left arm. A suboccipital craniectomy and laminectomy was performed from C1 to T2 vertebrae, and an extensive communicating hydromyelia was found. Clear fluid was aspirated from the cord. The dura mater was left open. Postoperatively, the patient remained stable without improvement. Lassman *et al.* (1968) in their case 6, performed a similar procedure without any significant improvement. Banerji and Millar (1974) note that there have been few reports concerning the complications of Arnold-Chiari malformations in adults. They contribute to the literature 20 adult cases of whom four had upper limb symptoms because of what they called syringomyelic syndrome. In these four cases, they describe muscle wasting of the shoulder, the upper limb, and hand, with depressed tendon reflexes and variable sensory involvement. No details of treatment were given. Barnett *et al.* (1973) mention 20 patients in their series who had significant proprioceptive and other sensory deficits in the upper limbs with sparing of the lower limbs. Spillane *et al.* (1957) describe six cases with severe proprioceptive loss in the upper limbs associated with a variable loss of other sensory modalities, and good preservation of most functions in the lower limbs. Appleby *et al.* (1968) describe a patient (their case 2) with weakness,

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anaesthesia, and loss of reflexes in the left upper limb as the main manifestation of an Arnold-Chiari malformation in adult life.

We believe that there is a specific local pathology, of a remediable nature, in this upper limb syndrome. Two cases will be described. In both, the unnatural arched course of the nerve roots in the root canals appeared to cause tension and stretching of the roots. When this was relieved by partially removing the floor of the root canal or a facetectomy, a marked clinical improvement followed.

Case reports

CASE 1

This female patient (GB) was seen at the age of 13 years in April 1971 with a history of headaches from the age of 12 years. Investigation showed that she had an Arnold-Chiari malformation and an aqueduct stenosis. A Torkildsen's shunt and a decompression at the foramen magnum were performed. Though the shunt worked, it was found that a partially noncommunicating hydrocephalus was simply converted into a communicating hydrocephalus, and the symptoms of increased pressure persisted. Because of this, a lumboperitoneal shunt was performed later, after which all her increased intracranial pressure symptoms diminished satisfactorily. She developed normally although she had to have several revisions of the peritoneal end of the shunt.

The patient did well until the early part of 1975 when she developed, over a period of several months, a progressive weakness of her arms, particularly the left arm. On examination, the patient was a young female, well-nourished, approximately 145 cm in height. Her cranial nerves were unremarkable except for a partial bilateral deafness which, when investigated previously, was found to be due to an unexplained, probably congenital, lesion of the acoustic nerves.

Muscle strength testing revealed some degree of weakness in all muscles of both upper extremities, and of particular note was the marked weakness of the left biceps, the patient being virtually unable to bring her hand to her mouth while standing, though she could do this when reclining. The left deltoid was also noted to be moderately weak and eversion of the left forearm quite weak. The grip on the left was slightly weaker than on the right. Flexion and extension of the wrist and movement of the fingers all appeared to be mildly weak, but approximately equal on both sides. Coordination appeared normal on finger-to-nose testing, but there was slight atrophy of the intrinsic

muscles of both hands, more so on the left than on the right. Surprisingly, a biceps reflex could be obtained on both sides, but triceps reflexes were not obtained on either side. The knee jerks and ankle jerks were intact. Plantar responses were flexor and Hoffman's reflexes were negative. There was no evidence of hypertonus. Sensory examination showed no definite abnormalities of all modalities of sensation in the upper limbs.

The myelogram on this patient revealed the usual appearance of an Arnold-Chiari malformation with a distended hydromyelic cord with upward running nerve roots in the cervical spine (Fig. 1).

Fig. 1 Myelogram—case 1.

On 20 May 1975 in St Luke's Episcopal Hospital, Houston, an extensive cervical laminectomy was carried out from C4 to C7 vertebrae, inclusive, on the left side. The dura mater and the nerve roots were inspected. The dura mater was opened, and the spinal cord was found to be rather large, but soft, pulsating, and easily indented. It was quite obvious that the hydromyelic cavity communicated with the ventricles, and the pressure inside the spinal cord was very low in the

upright sitting position, with ample room in the spinal canal. Since there was no distention of the cord, there was obviously no reason to incise it or to open the hydromyelic cavity. The nerve roots were then inspected, and it was apparent that the roots were not only running upwards and laterally in the lower segments of the cervical canal, but also appeared to be unusually tense. The dura mater was then closed, and the dural sleeves of the roots were inspected. Again, it was noted that the nerve root sleeves appeared to be unusually tight as they ran up over the lower margin of the neural canal. Using small curettes, the lower lip of the neural canals was curetted away until it appeared that the nerve roots had been satisfactorily decompressed by taking away approximately 3 mm of the floor of the root canals, throughout their length, and it was seen that the nerve roots were much looser. The procedure was performed at the C5, C6, and C7 root levels on the left side. The wound was then closed in multiple layers. The patient was then observed for eight days, and it became quite obvious that, even at this early stage, the strength in the left arm was improving so that she could now lift her food to her mouth; but paradoxically, the right arm, which had started to deteriorate rather rapidly just before her admission to the hospital, now weakened even further until it became about as weak as the left arm had been before the operation. The only complication which was observed was that the grip of the left arm was weaker postoperatively than before the operation. This was thought to be due probably to a traction injury of the lowermost part of the cervical cord which resulted in a partial C8 root lesion.

Nevertheless, because of the marked improvement in the strength of the left biceps, the right side was explored on 29 May 1975. The nerve roots appeared identical to those of the left side, and an extensive C6, C7, and C8 foraminotomy was performed on the right side. The left C8 foramen was enlarged at the same time. The C8 foraminotomies were probably unnecessary because the nerve roots were running fairly horizontally at this level. At the end of the procedure, the C6 and C7 nerve roots on the right side appeared to be satisfactorily loose. The patient recovered from these procedures and was discharged home within a few days of the second procedure.

On 7 August 1975, she was doing remarkably well. The strength was returning to both hands and the biceps muscles had recovered to almost normal power. The deltoids also appeared to be of normal strength. By 11 November 1977 the patient had recovered completely normal power

in both upper limbs; even the grips had returned to normal strength. The reflexes are still very sluggish in the upper limbs but are symmetrical and obtainable at all levels. There is no sensory deficit, and for all practical purposes this patient now has normal upper limbs.

She still has a problem with her lumboperitoneal shunt which intermittently is blocking and also causes some discomfort in her low back. A lumbosacral brace helps her. Recently, a ventriculoperitoneal shunt has been added.

CASE 2

This patient (WK) was 46 years old when seen on 16 January 1975. He complained at that time of several months of increasing numbness in the upper limbs in the ulnar distribution, and parascapular pain. This improved after cervical traction. Examination showed some mild hypaesthesia in the C8-T1 root distribution, and rather sluggish reflexes in the upper limbs, but no definite weakness.

He was admitted to the Methodist Hospital in Houston where a myelogram was performed. This showed extensive dilatation of the cervical spinal cord, which was diagnosed as hydromyelia, with typical upward running cervical roots, as shown in Fig. 2. The EMG, at the same time, showed large polyphasic motor unit potentials in all the

Fig. 2 Myelogram—case 2.

muscles supplied by C5, C6, and C7 nerve roots, on both sides. At this stage, the question of operative treatment was discussed with the patient, but it was pointed out that no specific successful surgical treatment had been devised for this problem, though myelotomy and possibly drain insertion have been used in the past. The patient was then discharged from the hospital.

He was seen periodically until May 1976 when he returned having deteriorated considerably, and showing a very weak deltoid on the right side. The right biceps showed no more than one-third to a half of normal power, and the power of the right triceps was reduced to two-thirds of normal. In spite of this, the grip on both sides appeared to be satisfactory, and the left upper limb showed no motor weakness. The sensory examination showed some numbness in the C8-T1 root distribution, and there was marked loss of position sense in the whole of the right hand.

By this time, case 1 had been fully evaluated and, being satisfied with her progress, we discussed this new type of operative treatment with the patient. He consented to a surgical exploration of his cervical nerve roots.

Initial testing in August 1976 showed that the patient had only enough power in his right deltoid to lift a one pound weight (0.45 kg) and only enough power in his right biceps to lift a five pound weight in his hand. On the left side, he could lift approximately five pounds with his deltoid, and approximately nine pounds with his biceps.

On 20 August 1976 at the Methodist Hospital, Houston, an extensive cervical laminectomy was performed and also a facetectomy to uncover completely the C5, C6, C7, and C8 roots, on the right side. Again it was noted that the C6 and C7 roots, in particular, had a markedly arched course, running up and arching over the lip of the foramen and then running down and forward in the neural canal (Fig. 3). It was quite evident that the nerve roots were unusually tense. Since a complete facetectomy had been performed, the floor of the root canals could be removed easily at all four levels, and a multilayered closure was then performed.

Postoperatively, the patient's progress was entirely satisfactory, and showed improvement in the strength of the upper limb, though not as marked as in case 1. Two weeks after the right sided laminectomy, a left sided cervical exploration was performed. At this time, although the laminectomy was completed to the same extent as on the right side, the facets were deliberately left intact, and only the floor of the neural canals

Fig. 3 Arrows show upward and then downward course of the nerve root from its origin (lowest arrow).

was curetted out, as in case 1, at all four levels, again removing an estimated 3 mm of the floor of the canal.

After the operation the strength in both arms gradually increased. By 29 November 1976, less than three months after his last operation, the deltoid, on the right side, could lift two pounds of weight (0.9 kg) in the outstretched hand. The biceps, on the right side, could lift eight pounds, while on the left side strength had improved slightly but was close to normal. On 23 March 1977, the patient's left arm was judged to be completely normal in strength and sensory perception. The right deltoid still showed only one-third of normal power. The power of the right biceps and brachio-radialis were estimated at three-quarters of normal. The right triceps, wrist, and finger flexors appeared normal. Stereognosis in the right hand returned to normal. Whereas, originally, the patient had to look at his hand to see if he still held a small object or whether he had lost it, he was able to do his work as a draughtsman without hindrance when last seen.

Discussion

The entity of delayed gradual onset of paralysis or weakness of the upper limbs should be emphasised since this particular facet of the Arnold-

Chiari malformation syndrome appears to be amenable to surgical treatment.

The course of the cervical nerve roots is an abnormal upward arch, and in the two cases described here, this abnormal course resulted in a stretch lesion of the nerve roots in the middle and lower part of the cervical spine. If the angulation of the nerve can be altered by removing part of the floor of the root canal, the tension on the cervical roots can be relaxed and a considerable improvement in function can result. In case 1, the improvements amounted to complete recovery from a severe disability. In case 2, four months after operation, the improvement is marked, but not complete. However, it should be noted that in the first case it took over a year to complete the recovery and we hope that the second case will continue to improve.

Drainage of the hydromyelic cavity appears to be inappropriate in this condition since it has not proved helpful in the past, and has no rational basis. It is, of course, necessary that the hydromyelia is not part of the picture of increased intracranial tension. This has to be relieved first, if present. It also appears that the hydromyelia itself, in the absence of increased intracranial tension and of other neurological deficits, is not the main cause of upper limb paralysis.

The exact nature of the procedure which will help most is as yet difficult to determine, since only two cases have been treated surgically in this fashion. Whether a complete facetectomy is really necessary, or whether an extensive removal of the floor of the root canal is sufficient, remains to be determined. Fortunately, in the present two cases, an extensive removal of the floor of the foramen and canal appear to be just as effective as a facetectomy; therefore, it would appear to be preferable, since it would lead to less impairment of stability of the cervical spine. In case 2, the neurological deficit and the age of the patient appeared to be sufficient to warrant the most extensive decompression that could be devised, in order to preserve his earning capacity. For these reasons, a full facetectomy was performed, on the right side,

since he is right handed, and only infraradicular foraminotomies (to coin a term) were performed on the left side. Fortunately, both sides appear to be recovering well. Indeed, the left side is back to normal strength in all its muscle groups.

The question of aetiology has been left to the last, since there does not appear to be a very good explanation available. While it could be postulated that in case 1 gradual maturation resulted in increased angulation and traction on the nerve roots, it is obvious that in case 2 no such cause could be invoked. Thus, there is no clear explanation for the delay in the onset of the radicular phenomena in the upper limbs; but at least the distinct entity of a traction lesion of the cervical roots can be alleviated by altering the course of the cervical roots.

References

- Appleby, A., Foster, J. B., Hankinson, J., and Hudgson, P. (1968). The Chiari anomalies in adult life. *Brain*, **91**, 131-139.
- Banerji, N. K., and Millar, J. H. D. (1974). Chiari malformation presenting in adult life. *Brain*, **97**, 157-168.
- Barnett, H. J. M., Foster, J. B., and Hudgson, P. (1973). *Syringomyelia*. W. B. Saunders: London.
- Carmel, P. W., and Markesbery, W. R. (1972). Early descriptions of the Arnold-Chiari malformation. *Journal of Neurosurgery*, **37**, 543-547.
- Hall, P. V., Campbell, R. L., and Kalsbeck, J. E. (1975). Meningomyelocele and progressive hydromyelia. *Journal of Neurosurgery*, **43**, 457-463.
- Lassman, L. P., Michael-James, C. C., and Foster, J. B. (1968). Hydromyelia. *Journal of the Neurological Sciences*, **7**, 149-155.
- List, C. F. (1941). Neurological syndromes accompanying developmental anomalies of the occipital bone, atlas and axis. *Archives of Neurology and Psychiatry (Chicago)*, **45**, 577-616.
- Logue, V. (1971). Syringomyelia: a radiodiagnostic and radiotherapeutic saga. 14th Crookshank Lecture. *Clinical Radiology*, **22**, 2-16.
- Spillane, J. D., Pallis, C., and Jones, A. M. (1957). Developmental abnormalities in the region of the foramen magnum. *Brain*, **80**, 11-48.