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CERVICAL SPONDYLOSIS WITH MYELOPATHY AS A COMPLICATION OF CEREBRAL PALSY[1]

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Abstract. Four cases of cerebral palsy with cervical spondylosis and myelopathy are described. Three patients had surgical decompression with poor results. The problems of diagnosis and management of such patients are discussed.

Key words: Cerebral palsy; Myelopathy; Cervical spondylosis.

Introduction

CERVICAL spondylosis is a relatively common cause of myelopathy in older individuals, especially those who have used their neck muscles for leverage or for heavy work for many years. The diagnosis is frequently delayed because the early symptoms are vague and progression may be slow and intermittent. There is no consistent neurological pattern and the findings may suggest a variety of other neurological diseases (Crandall and Batzdorf, 1966; Lees and Turner, 1963; Nurick, 1972; Peterson and Dayes, 1977; Valergakis, 1976; Wilkinson, 1976).

This paper calls attention to a group of individuals who are especially susceptible to this phenomenon because of frequent and severe stress on the neck, but who have not yet been identified as such in the literature, *i.e.*, the adult with cerebral palsy.

Case Reports

Case 1. B.B. was a 28-year-old female with athetoid cerebral palsy, more severe in the head, neck, and upper extremities, who was completely independent in self-care including ambulation without aid. She was a university doctoral degree candidate. In November 1977, she sustained a 'whiplash' injury in an automobile accident followed by pain in her neck and right arm. After about six weeks the pain subsided spontaneously with only occasional discomfort when under stress. In June 1978 (six months later), she awoke one morning with severe pain in her neck and inability to lift her right arm over her head. She was treated conservatively with little or no response for about two months, then she was hospitalised for evaluation. Radiographs of the cervical spine showed mild degenerative arthritis at C5–6. A myelogram showed a narrowed subarachnoid space with defects noted at C4–5 and C5–6 on the right and an anterior indentation at C4–5. The neurologist strongly recommended surgical decompression, but the neurosurgeon felt that her symptoms were not severe enough to justify the risk that she might lose function. She was treated with Haldol[1] and Sinemet[2] to

[1] Read at the Scientific Meeting of the International Medical Society of Paraplegia, Stoke Mandeville, July 1981.
[1] HALDOL (Haloperidol) McNeil Pharmaceutical, Spring House, P.A. 19477.
[2] SINEMET Merck, Sharpe and Dolune, West Point, P.A. 19486.

reduce the neck hypertonicity but the results were equivocal and she did not like the side effects, therefore these drugs were discontinued.

Both the weakness and the pain subsided gradually over the next few weeks and at follow-up a year later she was asymptomatic.

Case 2. M.M. was a 60-year-old female with athetoid cerebral palsy and an I.Q. of 34. She had been in numerous institutions since the age of six years, and it was impossible to obtain an accurate history, but it was documented in 1967 that she was able to walk, was independent in self-care, and 'did beautiful embroidery work.' Her speech was dysarthric and accompanied by much facial grimacing but was understandable. It is unclear whether the onset of deterioration in function was sudden or gradual, but by December 1976, she was totally dependent in self-care and an electric wheelchair was ordered. On 9 November 1977, she was admitted to hospital. A myelogram showed cord compression between C3 and 6 with almost complete block at C4–5 level. On 1 December 1977 she had a decompressive laminectomy from C2–7. The dura was not opened, but the cord was noted to bulge posteriorly. One week following surgery she was discharged back to the nursing home. It was reported that there was some sensory return post-operatively, but it lasted for only a few days, and was lost suddenly on 22 December and she was re-hospitalised for further evaluation. A repeat myelogram showed incomplete filling at the mid-cervical area, but no complete block. No further treatment was recommended. Since then there have been no major neurological changes. She remains essentially a C5 tetraplegic.

Case 3. J.H. was a 41-year-old male with athetoid cerebral palsy. His speech was dysarthric but easily understood and he appeared to be of average or better intelligence. He had always been dependent in self-care. The highest level of function he ever achieved was to be able to sit on the side of the bed unsupported and walk with moderate assistance from one person. As a teenager he could propel his wheelchair for short distances with his feet, but at no time could he feed himself or use his upper extremities purposefully. At the age of 18 years he acquired an electric wheelchair operated with tongue control, and he still uses it.

In 1960 (age 22) he had bilateral adductor tenotomies followed by a hip spica cast for 6–8 weeks. Whilst in the cast he spent most of his time prone with his neck hyperextended to enable him to see. During this time he first noticed numbness and increased spasticity in his legs. The neurological findings were: 'torticollis and lordosis of the neck, decreased sensation in the ulnar aspect of hands and forearms, no upper extremity reflexes, and 3 + lower extremity reflexes.' These symptoms improved after the hip spica cast was removed.

In 1968 (age 30) he noticed numbness and decreased athetoid movements in his arms, and there was urgency of micturition with occasional episodes of urinary retention. Over the next ten years he progressively lost motor and sensory function in all four extremities and became impotent. In April 1978 he was admitted to hospital. X-rays of the neck showed severe cervical spondylosis, and a myelogram showed a large extradural defect at C4–5 level, which was assumed to be due to a large anterior bony spur. He also had partial stenosis of the foramen magnum. On 31 October 1978 the C4–5 disc and anterior bony spur were surgically removed by an anterior approach. Post-operative X-rays six weeks later showed bony fusion at C4–5.

Although he reported some subjective improvement in sensation, there was no significant neurological change post-operatively, and he has remained stable since then with an incomplete C-5 tetraplegia.

Case 4. R.G. was a 46-year-old male with athetoid cerebral palsy, more severe in the head, neck, and upper extremities. His speech was almost unintelligible. Most attempts at functional activity such as eating, speaking, etc., resulted in severe

hyperextension of his neck. He appeared to be of average intelligence but had received no education because he was not accepted into a public school. He was completely independent in all self-care activities including ambulation without assistance. It was especially difficult to get a history from him because his 'yes' and 'no' responses often appeared the same.

About 1968 he first noticed neck pain, but he does not recall any other problems at that time. In 1972 he developed weakness of his right arm, and could no longer feed himself, and his balance was poor with difficulty in walking. Cervical spine X-rays showed spondylosis, but a myelogram was not done. A cervical collar was tried but could not prevent his hyperextension when he was active. In August 1972 he had a sudden exacerbation of symptoms with bowel and bladder incontinence, increased weakness and loss of sensation in all four extremities. This was diagnosed as 'hysterical weakness'. He remained incontinent for about two months, then gradually improved, but not to the previous level of independence.

For the next five years he remained fairly stable. He could no longer walk but could stand with minimal assistance to do a pivot transfer into bed and could propel his wheelchair with his feet. He could not feed or dress himself.

During 1978 he noted increasing weakness in his left leg, marked loss of sensation in both legs, and a degree of impotence. On 27 November 1978 a myelogram showed marked encroachment on interspaces from C3–7 and hypertrophy of the posterior ligaments. Surgery was recommended, primarily to prevent further neurological deterioration, and after much discussion he agreed.

On 31 January 1979 he had a bilateral decompressive laminectomy from C3–6. It was noted at surgery that his vertebrae were so sclerotic that it was difficult to find an instrument that could cut through the laminae. Post-operatively his neurological condition remained stable until after about a month when there was sudden improvement in the strength and sensation of his legs. He was again able to stand and to do a pivot transfer into bed. This lasted only a few days, when he began to have severe pain in his neck, especially when sitting, and over the next two months he deteriorated progressively until he became tetraplegic at C5 level. Attempts to immobilise his neck with bracing during this period were completely unsuccessful. On 11 May he was re-admitted to the hospital (three and one-half months post-operatively) because of respiratory difficulty. On the first day he had a cardiac arrest, but he was resuscitated and was placed on a respirator. He remained a C4 level tetraplegic until he died on 20 June 1979 (five months after the surgery).

Discussion

Patients 2 and 3 had an almost complete C5 tetraplegia at the time of surgery and no significant neurological benefit resulted from the operation except for (subjective) reports of temporary sensory improvement immediately after surgery which were not documented objectively in the medical records. It was felt that the surgery had been carried out too late because the diagnosis had not been made until the neurological damage had become irreversible.

When patient 4 was diagnosed, he was showing progressive deterioration, but he still had significant sensory and motor function in all four extremities. An extensive decompressive operation was done. This procedure has been shown to be effective without loss of spine stability or progression of neurological damage (Fager, 1976; Laterre and Stroobandt, 1976; Piepgras, 1976; and Stoops and King, 1962). But it is unlikely that the cases quoted by these authors were ever subjected to the extremes of

stress and range of motion, especially hyperextension, that occurred many times a day in this patient. The inability to immobilise his neck, even during the immediate post-operative period, was perhaps a factor in his rapid deterioration. In retrospect, the decision to operate was considered to be a mistake in judgment.

These four cases illustrate many of the problems in diagnosing neurological deterioration in someone with cerebral palsy, such as:

1. Poor verbal skills that make it difficult to obtain an accurate history.
2. New neurological symptoms tend to be ignored or overlooked.
3. Accurate neurological examinations on cerebral palsy patients are difficult to document so that minor changes may be missed.
4. Descriptions of previous levels of independence in self-care tend to be disbelieved unless they are confirmed by a reliable witness. However, such witnesses are difficult to find because these patients tend to move from one custodial institution to another and the medical records of these facilities are often poor.
5. Good X-rays are hard to obtain without general anaesthesia because of the involuntary movements occurring in these patients.

After making the diagnosis, the problem of management is even more difficult. It is not clear whether cervical spondylotic myelopathy is due to spinal cord compression or to ischaemia, or a combination of the two (Gooding, 1974; Gooding *et al.*, 1975, 1976; Hoff and Wilson, 1977; Nurick, 1972, 1976). With early diagnosis in an able-bodied person, the recommended treatment is to put the neck at rest with a collar, and to discontinue activities which cause particular stress on the neck, such as heavy lifting. In a patient with cerebral palsy this is impossible. Such necessary activities as eating and speaking may trigger violent muscle contractions in the neck often causing hyperextension which further narrows the cervical canal (Epstein *et al.*, 1970). Commercially available neck braces are not capable of immobilising the neck against the strong involuntary movements of these patients. Medications are usually minimally effective in reducing the force of the muscular contraction and certainly do not achieve a state of rest.

The results in the three patients who were treated surgically would make one hesitant to operate on an individual who still had partial neurological function, even with obvious progressive deterioration.

Summary

This paper presents a series of four patients with cerebral palsy, ranging in age from 28–60 years, who developed cervical spondylosis with myelopathy. One has responded well to conservative management (so far), but the three patients who were treated surgically all had poor results, with one disaster. These patients are being reported primarily to alert other physicians to the potentially poor results from surgical decompression in patients with uncontrollable spasticity of the neck.

Résumé

La papier presente une serie de quatre patients avec une paralysie cérébrale allant de

P'age de 28 à 60 ans qui a développé une spondylosis cervicale avec myelopathy. Un à bien répondu à un traitement consevati (traditionnel) (jusqu à maintenant) mais les trois patients qui ont subit une interrvention chirurguale ont eu de médiocre résultat dont un désastre. Les patients son surtout porteš à P'attention des autres docteurs pour prevenir des résultats médiocres possibles, dûs à la décompression chirurguale chez le patients atteints de spasme incontrolables du iou.

ZUSAMMENFASSUNG

Diese Arbeit stellt vier Fälle von Gehirnlähmung (cerebral palsy) dar. Bei den Patienten, die im Alter von 28 bis 60 Jahren stehen, ist Nackenspondylose mit Myelopathie (cervical spondylosis with myelopathy) aufgetreten. Von diesen ist einer mit bislang gutem Erfolg konservativ behandelt worden; bein den drei chirurgisch behandelten sind dagegen nur schlechte Ergebnisse, darunter eine Katastrophe, festzustellen. Dieser Bericht will andere Ärzte auf die Gefahren aufmerksam machen, die mit chirurgischer Dekompression bei Patienten mit unbeherrschter Nackenspastizität verbunden sind.

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