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ORIGINAL ARTICLE

Cervical myelopathy in athetoid and dystonic cerebral palsy: retrospective study and literature review

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Abstract The early onset of degenerative cervical lesions has been well described in patients suffering from athetoid or dystonic cerebral palsy. Myelopathy can occur and aggravate of their unstable neurological status. Diagnosis and treatment are delayed and disrupted by the abnormal movements. This retrospective study was implemented to evaluate the symptoms, the anatomical findings, and the surgical management of seven patients from 20 to 56 years old suffering from cervical myelopathy and athetoid or dystonic cerebral palsy. The mean delay in diagnosis was 15 months and the mean follow-up was 33 months. The initial symptoms were spasticity, limbs weakness, paresthesias and vesico-sphinteric dysfunction. In addition to abnormal movements, imaging demonstrated disc herniation, spinal stenosis and instability. All patients were managed surgically by performing simultaneous spinal cord decompression and fusion. Two patients benefited from preoperative botulinum toxin injections, which facilitated postoperative care and immobilization. Strict postoperative immobilization was achieved for 3 months by a Philadelphia collar or a cervico-thoracic orthosis. All patients improved functionally with a mean Japanese Orthopaedic Association score gain of 1.5 points, in spite of the permanent disabilities of the myelopathy. Complications occurred with wound infection, metal failure and relapse of cervical myelopathy at an adjacent level in one case each. All the previous authors advised against isolated laminectomy but no consensus emerged from the literature analysis. Spinal fusion is usually recommended but can be complicated by degenerative adjacent deterioration. Surgical management provides good outcomes but requires a long-term follow-up.

Keywords Athetoid cerebral palsy · Dystonia · Cervical myelopathy · Abnormal movements

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Introduction

Even though degenerative spondylotic cervical myelopathy (CM) is a well-known pathology, diagnosis and treatment are challenging for patients suffering from athetoid and dystonic cerebral palsy (ADCP).

A perinatal anoxia can lead to brain suffering and damage the basal nuclei resulting in movement disorders. Chorea and athetosis manifest by abnormal movements, brisk and explosive or slow and repetitive, respectively, affecting the control of voluntary movement.

The early onset of degenerative cervical lesions has been well described and can result in or decompensate CM, thus deteriorating their neurological status and decreasing their already reduced autonomy. The management of this condition is often delayed and labored from diagnosis to treatment. The abnormal and uncontrolled movements impair proper neurologic evaluation, imaging exploration and postoperative care. Surgery is considered as mandatory

but surgical modalities are not consensual. Postoperative immobilization necessitates rigid orthoses that are usually not tolerated.

The purpose of this study was to evaluate the surgical outcome of seven patients suffering from CM and ADCP, treated with spinal cord decompression and spinal fusion.

Method

A retrospective cohort study design was implemented to include seven patients suffering from ADCP operated on between 2001 and 2008. The mean age of patients at the time of surgery was 40 years and all patients were scored ASA 2 [6]. The mean duration between the onset and the diagnosis of CM was 15 months (1–36 months) and the mean follow-up time was 33 months (9–99 months). Before the onset of CM, one patient was ambulant, two could walk with technical aid and four were not able to walk at all.

The magnitude of dystonic neck movements was evaluated according to two items of the Burke–Fahn–Marsden scale where the severity and the triggering factors of cervical dystonia are scaled from 0 to 4 (a higher number corresponding to a more severe handicap) [4].

The Japanese Orthopaedic Association (JOA) score modified by Haro was used to evaluate pre- and postoperative functional impact of CM in ADCP [13].

The first evaluation was performed with standard and dynamic radiographs, computed tomography (CT) and magnetic resonance imaging (MRI). Dynamic radiographs were performed for every patient but were only interpretable for five patients (patients 1, 3, 5–7). All patients required an adapted sedation up to general anesthesia to carry out MRI. Standard and dynamic radiographs were done regularly throughout the follow-up period. Surgical modalities were chosen according to the extent of CM and to a possible associated vertebral instability. Every decompressed level was systematically fused and anterior fusion was performed either by an autologous bone graft or by a titanium mesh cage with plate fixation or according to Cloward's technique [5]. Posterior fixation was achieved using a segmental device with articular screws from C3 to C6 and pedicular screws at C2, C7 and T1 levels. Postoperative immobilization was achieved for 3 months either by a thermoformed cervico-thoracic orthosis or by a Philadelphia collar. A specialized rehabilitation department, within the same hospital, managed postoperative care.

Clinical cases (Table 1)

Patient 1

A 39-year-old woman suffering from ADCP and right neonatal hemiplegia was able to use public transportation and to walk with crutches before she developed sphincteric dysfunction, weakness of her left hemibody and eventually lost her walking ability. Her cervical spine was painless and diagnosis was delayed for 3 years before she underwent an MRI, showing a C5C6 herniated disc and an associated myelopathy. An anterior cervical discectomy and fusion with a locking plate was performed followed by a 3-month postoperative immobilization with a Philadelphia collar. No complication occurred but worsening of lower limbs spasticity necessitated botulinum toxin injections and bilateral Achille's tendon lengthening. At the last

Table 1 Patients: anatomical findings, evaluation and surgical management

Patient	Age (years)	Anatomical findings	Preoperative Burke score (from 0 to 4)		Japanese Orthopaedic Association		Surgical procedure
			Triggering factor	Severity factor	Preoperative	Follow-up	
1	42	C5C6 herniation	2	2	7.5	8.5	Discectomy C5C6, autologous graft with plate fixation
2	56	C5C6 instability	4	3	2	4.5	C2T1 posterior fusion
3	44	C5C7 spinal stenosis	1	1	7	8.5	Corporectomy C6, posterior C5T1 fusion
4	38	C3C4 spinal stenosis	2	3	6	8.5	Cloward's C3C4, posterior instrumented fusion C3C4
5	20	C2C5 spinal stenosis	2	2	2	2.5	C2C5 posterior laminectomy and fusion
6	35	C3C4 herniation	1	1	6	8	C3C4 discectomy and autologous graft with plate fixation
7	44	C3C7 spinal stenosis	2	3	4	4.5	C3C7 laminectomy, C2C7 posterior fusion

follow-up, she had recovered her walking ability with technical aid but had a residual neurogenic bladder.

Patient 2

A 56-year-old wheelchair bound man suffering from ADCP was referred to our institution after having an isolated cervical laminectomy performed 6 months earlier. After this first surgery, neurological symptoms improved but worsened again a few months later. Preoperative radiographs were not conclusive regarding spinal instability and a CT-scan with the neck in flexion and extension had to be performed under sedation in order to demonstrate postoperative spinal instability at the C5C6 level (Fig. 1). The patient refused surgery until his neurological condition worsened with sphincteric dysfunction and Lhermitte's sign. Preoperative botulinum toxin injections were performed (100 units Botox, Allergan®, in each splenius muscle). A posterior arthrodesis from C2 to T1 was then carried out and immobilization was accomplished by a cervico-thoracic orthosis. During the follow-up, a pullout of the left rod from the C2 pedicular screw occurred, with no modification of the symptoms or of the contra lateral construct. The spine was stable on dynamic radiographs and therefore the therapeutic management was not modified.

Neurogenic pain resolved leaving behind a residual weakness of the left arm and an unimproved sphincteric function.

Patient 3 (Fig. 2)

A 44-year-old computer scientist suffering from ADCP secondary to neonatal anoxia was able to crawl and to drive his wheelchair with an occipital command. He presented with isolated paresthesia of the left arm. MRI demonstrated CM from C5 to C7 with no spinal instability on dynamic radiographs. A preoperative botulinum toxin injection was performed followed by a posterior decompression and fusion from C5 to T1 with a C6-corporectomy using a mesh cage and a locking plate. At the last follow-up, he had fully recovered.

Patient 4

A 33-year-old male who had suffered from neonatal hypoxia with resulting ADCP was a permanent wheelchair user and was working as an electrical engineer. He

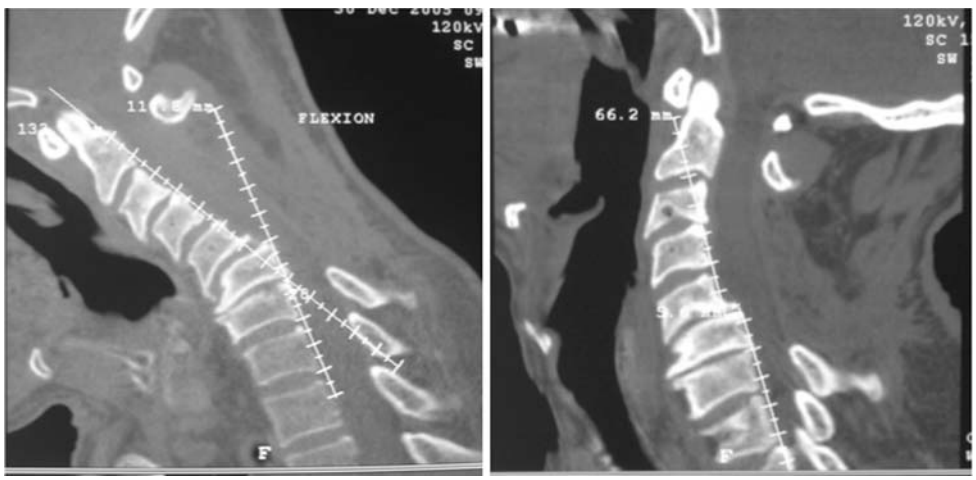

Fig. 1 Patient #2. Dynamic cervical CT-scan: C5C6 malalignment and instability after an isolated laminectomy

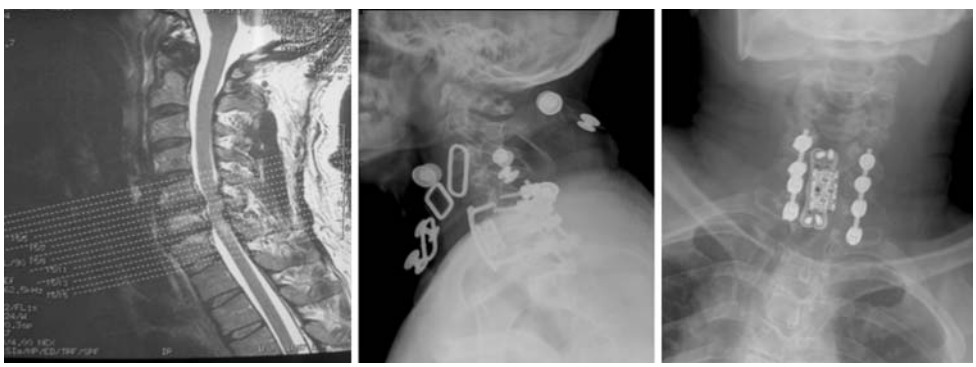

Fig. 2 Patient #3. Pre-operative sagittal T2-weighted MRI and postoperative radiographs: circumferential decompression and fusion for cervical stenosis without instability

experienced within a few months a deterioration of his neurological state with quadriparesis and sphincteric dysfunction. The MRI revealed a C3C4 disc herniation without instability. We performed an anterior cervical discectomy and fusion according to the Cloward's technique with a posterior C3C4 arthrodesis without decompression.

At 15 months follow-up, he developed a recurrence of CM with a relapsing quadriparesis and a deterioration of his vesico-sphincteric function. MRI and radiographs showed cervical instability and stenosis at the C4C5 level. A circumferential fusion and decompression improved his neurological functions but left him with a residual spasticity and neuropathic pain. He underwent surgical drainage and antibiotic therapy for a posterior wound infection. Postoperative immobilization was achieved by a cervico-thoracic orthosis. At the last follow-up, the patient improved but had a disabling residual spasticity.

Patient 5 (Fig. 3)

A 20-year-old man with ADCP secondary to neonatal infection used to walk on all fours and to speak with sign language. Swallowing and sphincteric functions deteriorated and the abnormal movements of both arms decreased progressively leading to diplegia. An MRI demonstrating cervical spine stenosis from C2 to C5, with no vertebral instability on plain radiographs, made the diagnosis of CM possible 5 months after the onset of the first symptoms. Posterior C2C5 spinal decompression and fusion was performed. Neurogenic pain and motor function got better but deglutition did not improve, necessitating permanent gastrostomy feeding. Residual spasticity developed and patient was permanently restricted to wheelchair.

Patient 6

A 42-year-old woman who had suffered from neonatal meningitis and secondary ADCP used to speak in sign language, to drive an equipped car and graduated high school. Her condition worsened with numbness and clumsiness of her four limbs and sphincteric dysfunction. In 1 month, quadriparesis set in and she lost her walking ability. MRI showed a C3C4 cervical disc herniation with myelopathy necessitating an anterior fusion with autologous graft and plating. A Philadelphia collar was provided for postoperative immobilization. At 6 weeks, she sustained algoneurodystrophy of her left arm and her neurological status slowly improved. At the last follow-up, fusion was achieved and she was able to walk, to use sign language again with her right arm but had a disabling residual spasticity of her left hemibody, necessitating regular botulinum toxin injections.

Patient 7

A 44-year-old male suffering from ADCP complained of an acute loss of his walking ability. He used to walk with a walker and to communicate with signs. Within 4 months, he developed quadriparesis and lost completely his autonomy, necessitating permanent nursing. MRI showed degenerative spondylotic CM from C3 to C7, with no vertebral instability. Posterior decompression from C3 to C7 and fusion from C2 to C7 were performed and a cervico-thoracic orthosis achieved postoperative immobilization. The postoperative course was uneventful. At the last follow-up, he was able to make a few steps with a walker but did not recover his ability to feed himself or to use signs.

Discussion

The incidence of CM in patients suffering from ADCP remains unknown [7]. Diagnosis is challenging and often delayed [15]. Anderson et al. [1] first reported this pathology in 1962 but since, it was only briefly touched upon in the literature.

Abnormal and uncontrolled movements interfere with voluntary command [7]. Dysarthria is also often present so the help of the entourage is necessary to detect early neurological changes and to perform an accurate clinical evaluation [7, 14, 16, 18]. Decrease in abnormal movements

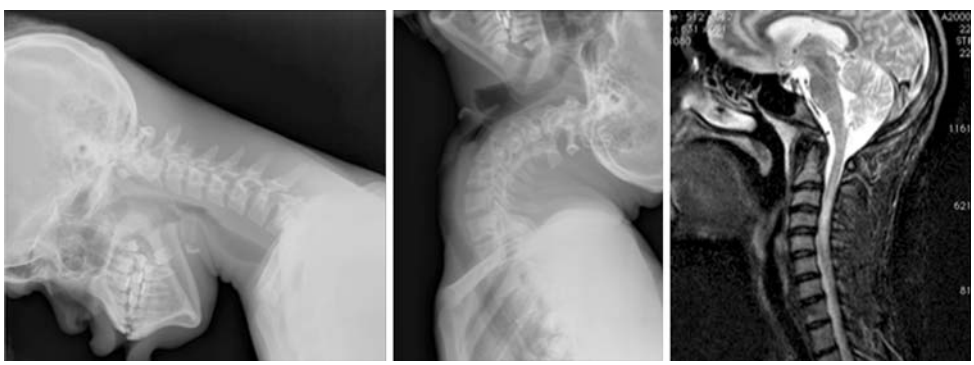

Fig. 3 Patient #5. Dynamic roentgenograms and sagittal T2-weighted MRI: congenital spinal stenosis without instability

must be considered as an alarming sign even though it could be interpreted as an improvement. An isolated radicular compression without myelopathy can sometimes occur [10].

In opposition to degenerative spondylotic CM of the elderly, these patients are younger and are generally in their fourth decade. Initial presentation is insidious and symptoms are lately detected leading to the delay of surgical management [16]. Better outcomes are reported if early surgical therapy is conducted. The only patient who did not suffer from neurological deterioration in our study was diagnosed early thanks to the vigilance of his entourage. For the others, residual symptoms persisted with major functional impact. In our experience, surgical management aims at first to prevent further neurological worsening and secondarily to give the patients the best ability to recover [8]. A complete recovery can only be expected with early diagnosis and treatment.

The majority of authors use the JOA score to evaluate the functional condition of their patients, especially the motor items [2, 23]. According to this score, all of our patients improved functionally with a mean gain of 1.5 points (0.5–2.5) but this score does not take into consideration the specificity of athetoid or dystonic movements that disrupt the voluntary command therefore rendering the results inaccurate. The frequency and the intensity of these parasitic movements may vary with stress and sensory stimuli, leading to fluctuation of motor function. Because of the initial neurologic condition, this score does not seem accurate enough. Our clinical feeling of major preoperative deterioration and of after-effects was not reflected by the improvement of the JOA score we had observed. Other authors who conducted similar studies evaluated surgical outcome by evaluating walking capacities or using a pain scale (Denis's scale) [19, 20]. Important residual neuropathic pain was observed for three patients and the ability to ambulate had worsened for six patients but even if surgery improved it for all of them, it was never the same as before the onset of CM.

The presence and the evolution of preoperative vesicospincteric problems were rarely reported which we think are of great importance in disability [2, 9, 16, 19, 22, 23]. Of the six patients who newly developed or had a worsening of sphincteric control, only three recovered their previous function.

The early onset of degenerative lesions in ADCP has been well described by Harada in a radiological study of over 180 patients, compared with controlled subjects [12]. Cervical spinal stenosis and instability were found to be more frequent in athetoid patients than in the controlled group [12, 13, 16, 18, 24].

The importance of spinal cord injury during these repetitive and uncontrolled movements should be underlined [14]. As an example, for patient #5, a permanent hyper extended posture and a constitutional spinal stenosis were the causative agents of his CM, with no significant degenerative lesion. In our study, only one patient presented with cervical instability precipitated by an isolated cervical laminectomy.

The inability to maintain a fixed posture challenges the imaging evaluation. Standard or dynamic radiographs are subjected to difficulty of interpretation, which might disrupt the detection of secondary destabilization. MRI is the only way to assess CM but is difficult to perform in these patients. This fact is rarely discussed in the literature and an adapted sedation or general anesthesia was always necessary in our experience, making MRI not suitable to screen CM for every athetoid or dystonic patient.

All authors advise against conservative therapy [7, 10, 19]. Surgical treatment always requires spinal cord decompression by anterior approach (corporectomy and discectomy) or by posterior approach (laminectomy and laminoplasty) [2, 7, 9, 13, 17, 19, 22, 23]. No isolated cervical laminectomy should ever be performed alone [2, 11, 14, 17, 18].

While some authors perform circumferential fusion [9, 16], others limit their fusion to either anterior or posterior elements [7, 10, 14, 17]. In our study, fusion was always limited to the liberated, malaligned or unstable spinal segments in order to preserve a residual mobility of the cervical spine which is required, for example, to swallow easily or to drive an electric wheelchair with a chin or occipital command. However, where long fusions might increase disability, short fusions may expose the cervical spine to further damages. A systematic procedure cannot be implemented for all patients and surgical therapy must be adapted for each of them [19].

Many authors reported a relapsing clinical and radiological deterioration with long-term follow-up. Spinal fusion increases strains on adjacent levels, predisposing to adjacent level instability or stenosis [2, 11, 13]. Azuma et al. [2] observed 80 and 60% of neurological and radiological deterioration, respectively, in a 15-year follow-up study with spinal fusion. Therefore, he recommended performing laminoplasty without spinal fusion but adjacent degenerative modifications developed as well. Ueda et al. [23] performed cervical laminoplasty with selective myotomies and obtained better clinical results than isolated laminoplasty. At 5-year follow-up, neither secondary deterioration nor spinal instability occurred. He supposed that cervical myotomies might decrease the intensity of abnormal movements and therefore protect the spinal cord. In the absence of preoperative instability, alternatives of spinal fusion might be discussed. However, the notion of high incidence of spinal destabilization secondary to isolated decompression may constrain the wide use of these techniques.

In athetoid or dystonic patients, strict postoperative immobilization is always necessary after spinal fusion [9, 16, 18, 23]. Some authors used a halo vest up to 6 months, but the rate of complications was as high as 40%, with pin-loosening or pin-site infection [11].

In our study, no complication of postoperative immobilization occurred. In one patient, unilateral implant failure proved the elevated constraints applied on the construct but this complication was also reported by authors using halo immobilization [9].

Botulinum toxin is widely used to decrease spasticity and dystonic movements. Many authors put the emphasis on the beneficial effects of its use during the perioperative period [3, 21]. Abnormal strains are reduced and the tolerance of the spinal orthosis improves. Because of recent changes in our management, only the last two patients benefited from preoperative botulinum toxin injection that will continue to be used in our practice.

Conclusion

CM in ACDP is a dreadful consequence of abnormal movements of the cervical spine. The loss of autonomy sets in as soon as the first symptoms occur.

Several anatomical patterns exist, from disc herniation to extensive spinal stenosis. Vertebral instability is not necessary for the development of CM in these subjects.

Even if the abnormal movements disrupt the radiological evaluation and the postoperative care, all patients should be managed surgically. Medullary decompression and spinal fusion is the recommended treatment and even though outcomes are satisfactory, it does not mean an absence of after-effects. The use of botulinum toxin increases the tolerance of postoperative immobilization. In the long term, clinical relapse can develop secondary to the degradation of adjacent levels. Therefore, follow-up must absolutely be prolonged, in order to detect possible recurrences. Screening of CM in ADCP could rely on oriented examination, on therapeutic education of the patients and on the distribution of information to the concerned associations.

Conflict of interest statement None.

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