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Review Article

Cervical Hyperextension and Scoliosis in Muscular Dystrophy: Case Report and Literature Review

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Abstract

Introduction: Cervical hyperextension is rarely seen, there has been a paucity of data on the association of this condition in the published literature. The current case is even rarer, because it presented not only a cervical hyperextension but also a scoliotic curve involving C1-T5.

Materials and methods: This article presents a case of cervical hyperextension and scoliosis in a 19-year-old patient with Emery-Dreyfus muscular dystrophy who underwent surgical correction using navigated pedicle screw placement. The Patient was followed for 2 years. A thorough review of the literature was conducted using the Medline database between 1988 and 2014.

Results: The time consumption for the surgery was 320 min. Blood loss was 600 ml, which was collected by cell saver. No surgical complications occurred. Both the cervical hyperextension and scoliosis were significantly corrected. The scoliotic curve was corrected from 35.0° to 6.2°, and the hyperextension was corrected from 56.6° to 24.5°. After surgery, the patient's trunk had been well balanced on both frontal and sagittal view. And he was able to walk in an upright position with looking straight ahead on his own accord.

Conclusions: Cervical deformity associated with muscular dystrophy is rare. The rigid and significantly increased cervical lordosis often forces the patients to bend their trunk forward and assume awkward compensating postures in order to look straight ahead, worsening significantly their quality of life. Posterior cervical muscle release and instrumented fusion are effective for correcting such deformities. The selection of candidates for surgery depends mainly on the type of myopathy and cardiorespiratory function. Preoperative CT scans of patients should be thoroughly analyzed and close attention paid to the pedicle diameter.

Keywords: *Cervical Spine; Cervical Scoliosis; Cervical Hyperextension; Sagittal Imbalance; Cervical Correction; Muscular Dystrophy*

Introduction

The management of muscular dystrophy and spinal deformity can be challenging for spine surgeons. Scoliosis in thoracic or lumbar spine is the most common spinal deformity associated with muscular dystrophy. However, either hyperextension or scoliosis in cervical spine is rare. The reported experience for treating cervical hyperextension is limited in the literature. Only a few case series and case reports have been published so far. Giannini., *et al.* [1] reported the first case of surgically treated cervical hyperextension in 1988. And in 2005 and 2006, they published 2 series of 7 patients [2,3]. All the patients were treated with a posterior cervical muscle release, non-instrumented fusion, and postoperative plaster jacket immobilization. In 2005, Arkader., *et al.* [4] reported a case treated with a posterior soft tissue release, osteotomies, and instrumented occiput-cervical fusion. In 2009, Poulter., *et al*. [5] reported 3 cases and described 4 major complications they have encountered.

Cervical hyperextension in muscular dystrophy is due to weakness of the neck flexors, and contracture of the neck extensors. The rigid and significantly increased cervical lordosis often forces the patients to bend their trunk forward and assume awkward compensating postures in order to look straight ahead, worsening significantly their quality of life [3].

To our knowledge, only 3 cases of cervical hyperextension treated by instrumented fusion have been described in the literature, The majority of the reported cases were treated by non-instrumented fusion, which could be due to those surgeries' being performed before 2002 when cervical instrumented fusion was not widespread yet. With the development of instrumentation and navigation, surgical treatments for such cases can be different. This article presents a case of cervical hyperextension and scoliosis in a 19-year-old patient with Emery-Dreyfus muscular dystrophy who underwent surgical correction using navigated pedicle screw placement.

Methods

The current case is a male with Emery-Dreyfus muscular dystrophy. He was 19 years old at the time of surgery. His major complaint was an increased difficulty of looking forward. In order to look straight ahead, he had to bend his trunk which was awkward and uncomfortable (Figure 1). He also suffered from poor head control in extension with frequent backward falling of the head.

Figure 1: *The patient had no problem with standing up straight, but when he was doing so, he could only look upward. To look straight ahead, he had to bend his trunk forward, which was awkward and uncomfortable.*

The lateral radiograph showed a hyperextension in the cervical spine, and the anterioposterior radiograph revealed a scoliosis involving C1 – T5 (Figure 2).

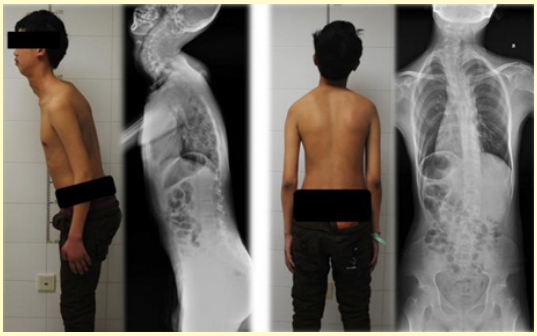

Figure 2: *The lateral radiograph showed a cervical hyperextension, and the C2-7 angle was 56.6°. The anterioposterior radiograph revealed a scoliotic curve involving C1 – T5, the Cobb angle was 35.0°.*

The patient was ambulant with both 4/5 upper and 4/5 lower extremity weakness. When checking his neck, we found that his neck lordosis was very rigid (Figure 3), the range of motion of the neck was limited. Both Hoffmann's sign and Babinski's sign were negative.

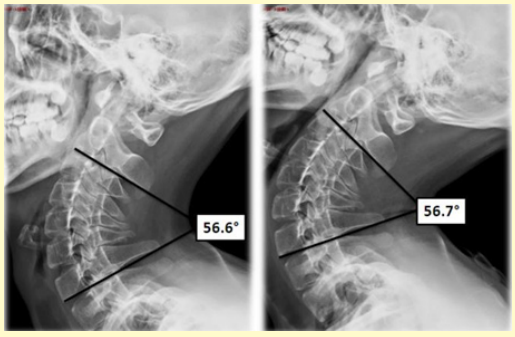

Figure 3: *Cervical forward and backward bending radiographs were taken, on which we found that there was very limited flexibility between C2 and C7. The difference in C2-7 angle was only 0.1°between the two radiographs. However, significant change in occiput-C2 angle could be seen when the neck moved from extension to flexion position*

The MRI of the spine showed no abnormalities of the spinal cord. The MRI of extremities revealed a decrease in the muscle mass, and fatty infiltration in the biceps femoris and semi membranous muscles bilaterally, consistent with the muscle dystrophy pattern. The cardiac function of the patient was evaluated by electrocardiography and echocardiography, and the results were normal. The respiratory function was evaluated by spirometry, which showed restrictive ventilatory defects with significantly decreased FVC (forced vital capacity). The FVC (1.92 L) was 57.5% less than expected for his age and gender (Figure 4). The measured SaO_2 (arterial O_2 saturation), however, was normal, which was 96% on room air.

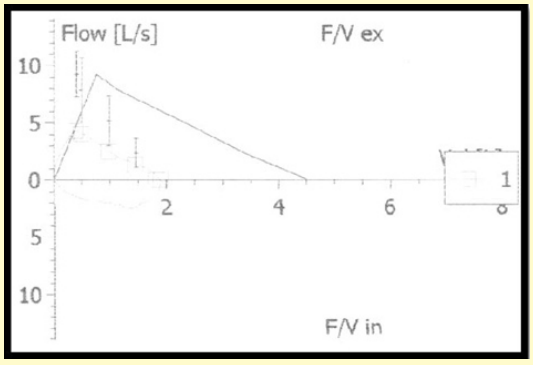

Figure 4: *The results of spirometry showed restrictive ventilatory defects. The FVC(1.92 L) was 57.5% less than expected for his age and gender.*

Surgical Techniques

Surgery was performed with the patient under general anesthesia and in prone position on a carbon-fiber operating table. A longitudinal median incision was made. The paravertebral muscles were detached from the spinous processes and then the laminas of vertebrae C2–T5 were bilaterally exposed. A small part of paravertebral muscles at C5 level was transversally sectioned. Unlike the previously reported procedures, we neither applied traction nor detached interspinous Ligament. The head of the patient was supported by a soft

holder during the surgery, so that the gravity could force the neck in anterior flexion. During the exposure, the neck hyperextension was gradually neutralized due to both the muscle release and gravity.

Pedicle screws were bilaterally inserted at C2, C5, and T1-T5 levels under navigation guidance (Stryker Navigation System II) (Figure 5). Lateral mass screws were bilaterally inserted at C3, C4, and C6. The screw diameters were 3.5 mm in the cervical spine and 5.5 mm in the thoracic spine, respectively.

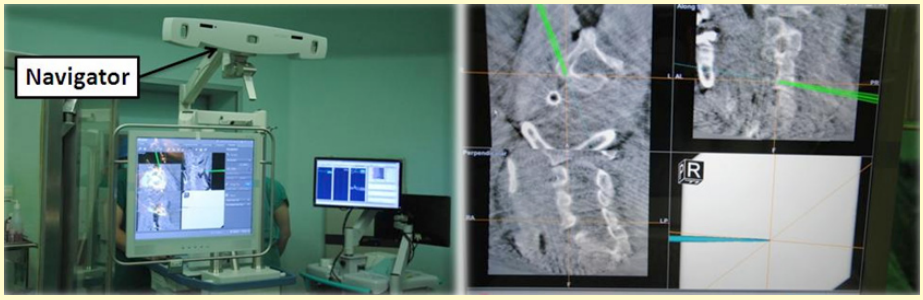

Figure 5: *Pedicle screws were inserted under navigation guidance.*

A special rod was used, half of which was 3.5-mm-diameter and the other half was 5.5-mm-diameter. After instrumentation of a rod on the convex side, the rod was derotated, and in the same time, an assistant applied some force on the head to bend the neck into a neutralized position. The screw nuts were then locked on the convex side. In order to achieve further correction, in-situ bending on the convex rod was performed. Meanwhile, the rod on the concave side was instrumented, and all the screw nuts were locked. Routine wound closure and postoperative radiographs were performed.

Somatosensory-evoked potential (SSEP) and Motor evoked potentials (MEP) monitoring were employed for the case and remained normal throughout the procedure.

Results

The time consumption for the surgery was 320 min. Blood loss was 600 ml, which was collected by cell saver. No surgical complications occurred. Both the cervical hyperextension and scoliosis were significantly corrected. After surgery, the patient's trunk had been well balanced on both frontal and sagittal view. And he was able to walk in an upright position with looking straight ahead on his own accord. (Figure 6).

The patient was followed for 2 years. The results of angle measurements are listed in Table 1.

	A-P Radiograph	Lateral radiograph		
	Cobb angle of Scoliotic curve	C2-7 angle in flexion position	C2-7 angle in neutral position	C2-7 angle in extension position
Preoperative	35.0°	46.8°	56.6°	56.7°
Postoperative	6.2°	N/A	24.5°	N/A
2 years	6.5°	N/A	25.5°	N/A
N/A -not available.				

Table 1: *The Angle Measurements in the anterioposterior and lateral radiographs.*

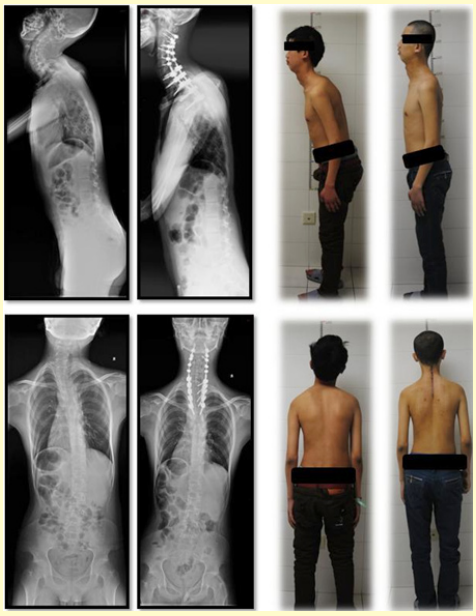

Figure 6: *Both the cervical hyperextension and scoliosis were significantly corrected.*
The scoliotic curve was corrected from 35.0°to 6.2°, and the hyperextension was corrected from 56.6° to 24.5°.

Discussion

Cervical hyperextension is rarely seen, there has been a paucity of data on the association of this condition in the published literature. The current case seems even rarer, because it presented not only a cervical hyperextension but also a scoliotic curve involving C1 – T5. To correct such a curve, we believe that cervical pedicle screws are necessary to provide enough corrective forces. Considering the risk of neural and vascular injuries associated with cervical pedicle screw placement, we used navigation during the procedure. Another key point for correcting such a curve is using the special rod, half of which was 3.5-mm-diameter and the other half was 5.5-mm-diameter. With the rod, we were able to perform derotation and in-situ bending, which were essential for achieving a satisfactory correction.

Which Patients Are Indicated for Surgical Correction?

The selection of candidates for surgery depends mainly on the type of myopathy and cardiorespiratory function, because the type of myopathy is associated with their life expectancy, and the cardiorespiratory function determines if the patients are able to tolerate the surgeries. As summarized by Giannini., *et al*. [2], Emery-Dreifuss muscular dystrophy, merosindeficient congenital muscular dystrophy, and meros in positive congenital muscular dystrophy were considered indications for surgery, because in all of these myopathies, with the usual course of the disease, the life expectancy of the patients would be acceptable.

In the current case, the respiratory function was evaluated by spirometry, which showed restrictive ventilatory defects. The FVC (1.92 L) was significantly decreased. However, the patient turned out to tolerate the surgery very well. After surgery he was extubated without difficulty, and no intensive care or mechanical ventilation was needed afterwards. In our institution, a measured vital capacity ≦ 35% of the estimated value is considered as one of the contraindications for general anesthesia and surgery. For a patient with VC ≦35%, full-time non-invasive ventilator support is usually applied for 1-2 months, followed by an evaluation of respiratory function. If the respiratory function has improved with VC ≧35% and SaO2 ≧ 95% on room air, surgical treatments can be performed.

Patients with Emery-Dreifuss muscular dystrophy usually have a cardiomyopathy with conduction defects. And sometimes a pacemaker is needed. The cardiac function of the current patient was evaluated by electrocardiography and echocardiography, and the results were normal. This could be due to the patient's still being young, the cardiac abnormalities have not been fully established by his second decade.

Surgical Techniques: Instrumentation or Non-instrumentation?

To our knowledge, only 3 cases of cervical hyperextension treated by instrumented fusion have been described in the literature, 1 of them was reported by Arkader., *et al*. in 2005 [4], and the remaining 2 were reported by Poulter., *et al*. [5] in 2009. All the cases reported by Giannini., *et al*. [1-3] were treated by non-instrumented fusion. The difference in the surgical methods could be due to Giannini's cases being treated before 2002 when cervical instrumented fusion was not widespread yet.

Generally speaking, instrumentation provides more corrective forces and more secure maintenance of correction than non-instrumentation does. Instrumentation also makes postoperative plaster jacket immobilization unnecessary. Besides, considering cervical instrumentation having been grasped by many spine surgeons nowadays and being a safe and quick procedure, we prefer instrumentation to non-instrumentation for cervical hyperextension cases. As for the current case, the scoliotic curve couldn't be corrected by non-instrumented procedure, because most of the correction was achieved by derotation and in-situ bending. When performing cervical fusion procedure, we need to consider proximal instrumented level. Both Giannini., *et al*. and Poulter., *et al*. chose C2 as proximal instrumented level, while Arkader., *et al*. chose occiput instead. As we know, the less levels fused, the more motion of the spine saved. Giannini *et al*. [1-3] have found that there is considerable motion remained between occiput and C2, which should be reserved if possible. Besides, most of the reported patents were well treated by C2-C7 fusion. As such, we don't think it is necessary to fuse up to occiput, C2 should be routinely chosen as proximal instrumented level.

Navigated Pedicle Screw Placement in Cervical Spine

Pedicle screw instrumentation is essential for correction and stabilization of spinal deformity. However, inserting pedicle screws in cervical spine is challenging due to the small osseous morphometrics and the close proximity of neurovascular elements. There are a variety of descriptions in the literature telling us how to find entry points and insert cervical pedicle screws. When intra-operative navigation is applied, the procedure becomes simpler, because the navigator can help us identify the best available bone stock for inserting pedicle screws in altered anatomy. Besides, navigation has been found to improve the accuracy of pedicle screw fixation with significantly reduced misplacement rates.

Preoperative CT scanning of cervical spine is mandatory for determining if the pedicles are large enough to accommodate the screws. In the current case, the width of the C2 pedicles is 6.7 mm, and that of C3-C7 increased caudally from 4.95 to 5.57 mm, which is consistent with the findings in the previous reports (Table 2).

Cervical Level	Current Case	Ebraheim., *et al.*[6]	Jones., *et al.*[7]	Panjabi., *et al.*[8]	Karaikovic., *et al.*[9]		Onibokun., *et al.*[10]	Su., *et al.*[11]
					Female	Male		
C3	4.95	4.7	4.8	4.3	5.3	4.5	4.7	4.9
C4	4.97	4.7	5.2	4.4	5.4	4.5	5.0	5.1
C5	5.03	4.9	6.1	4.9	5.7	5	5.5	5.8
C6	5.13	5.1	6.5	5.1	5.9	5	5.7	6.0
C7	5.57	N/A	6.9	5.6	6.7	5.9	6.5	6.5
N/A -not available.								

Table 2: *The pedicle width of C3-7 measured in the current case and literature.*

As shown in the literature, C2 usually has large enough pedicles and can be instrumented with pedicle screws in the majority of patients. Smith., *et al*. [12] found that the mean pedicle width of C2 was 5.8±1.2 mm, and in more than 75% of patients the width of C2 pedicles is ≧ 5 mm. Wu., *et al*. [13] found that the mean C2 pedicle width measured directly and by CT scan was 7.8 and 6.6 mm. Regarding the pedicle width of C3-C7, all the studies showed that there was a trend toward increasing size proceeding caudally. The C3 and C4 pedicles are narrow in many patients making screw insertion risky and unsafe. Inserting pedicle screws at C5-C7 levels is optional, but considering the wide variations in pedicle anatomy between individuals, preoperative CT scans of patients should be thoroughly analyzed and close attention paid to the pedicle diameter. We usually consider 5.0mm as the minimum pedicle width required for 3.5-mm screw insertion, leaving at minimum 0.5 to 0.75mm of medial and lateral cortical wall thickness.

Conclusion

Cervical deformity associated with muscular dystrophy is rare. The rigid and significantly increased cervical lordosis often forces the patients to bend their trunk forward and assume awkward compensating postures in order to look straight ahead, worsening significantly their quality of life. Posterior cervical muscle release and instrumented fusion are effective for correcting such deformities. The selection of candidates for surgery depends mainly on the type of myopathy and cardiorespiratory function. Preoperative CT scans of patients should be thoroughly analyzed and close attention paid to the pedicle diameter.

Declaration of interest

The authors report no conflicts of interest.

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