Long-segment Spontaneous Cervical Arthrodesis in Juvenile Rheumatoid Arthritis

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Objective: To describe the first case of long-segment spontaneous fusion of the cervical spine in juvenile rheumatoid arthritis and the major pitfalls of its treatment.

Materials and Methods: A 27-year-old female with juvenile rheumatoid arthritis presented with two-month duration of posterior neck pain and progressive weakness. Imaging studies showed spontaneous apophyseal joint fusion from occipital condyle to C6 and instability at C6/7 level. During operation, endotracheal intubation was not feasible due to stiff neck and the authors should select the way of tracheostomy. Moreover, severe osteoporosis restricted surgical option. At finally the patient was approached posteriorly with interspinous process wiring with titanium-braided cable and integrated bone graft was performed at C6/7 level.

Results: Fortunately, no postoperative complication were noted. While over 6 months of follow-up, pathologic reflexes disappeared and activities of daily living improved. Follow up computed tomography showed well-established bone fusion.

Conclusion: It is suggested that spontaneous long-segment cervical arthrodesis is related with severity and rapid progression of the disease, which seems to be a late manifestation of cervical involvement.

Key Words: Spontaneous arthrodesis - Fusion - Cervical spine - Juvenile rheumatoid arthritis

INTRODUCTION

Juvenile rheumatoid arthritis is a systemic progressive polyarticular inflammatory condition causing peripheral and axial joint destruction and has a relatively more favorable prognosis than adult type rheumatoid arthritis. Recent advances in medical treatment enable more favorable prognosis, but rapid-progressing cases may result in severe complications such as joint deformity, arthrodesis of joints, osteoporosis, and decreased bone mass. The common clinical manifestation of spinal involvement is instability of high cervical spine. The cervical apophyseal joint fusion is rare and only seen in juvenile type rheumatoid arthritis. One or two level fusion are observed in upper cervical spine but fusion over 3 level in cervical spine has not been reported yet. We report herein a case of spontaneous cervical arthrodesis from the occiput to C6 with subjacent instability at C6/C7 level in juvenile rheumatoid arthritis.

CASE REPORT

A 27-year-old female presented with two-month duration of posterior neck pain and progressive weakness. She had juvenile rheumatoid arthritis for 10 years and showed contractions in major joints including the hip, knee, elbow, ankle, and mandible. The patient had been treated with methotrexate, steroids, hydroxychloroquine, NSAIDs, and folic acid. Symptoms aggravated rapidly despite medications; the patient became wheelchair-bound for 3 years before admission. There was no family history of rheumatoid or other forms of arthritis.

On neurological examination, the patient showed reduced motor power, and the contracture of the joints restricted the range of motion. Motor power was checked within the possible range of motion. The examination revealed both paresis and dysesthesia below the C7 dermatome. Neck motion was severely restricted, and only simple 'yes' or 'no' motion was possible.
The patient showed loss of lordosis and sclerotic changes of the facet joints in cervical simple X-rays 5 years before admission (Fig. 1A). The atlanto-axial distance was within the normal range. Simple X-rays of the cervical spine on admission showed spontaneous apophyseal joint fusion from the occipital condyle to C6 (Fig. 1B). Flexion and extension X-rays showed wide apophyseal joint space between C6 and C7, suggesting instability and excessive mobility (Fig. 1, B and C). Thoracic and lumbar spinal X-rays showed no notable findings of apophyseal joint fusion.

Computed tomography of the cervical spine revealed the apophyseal fusion of the atlanto-occipital and atlanto-axial joints. The pannus between the atlas and the odontoid process was calcified and became a bony bridge. An atlanto-axial distance interval of >5 mm suggested previous atlanto-axial instability (Fig. 2A). The apophyseal and uncovertebral joints from the atlas to C6 were also fused (Fig. 2, B and C). Cervical magnetic resonance imaging showed disc degeneration, spondylotic change, hypertrophy of the ligamentum flavum, and spinal cord compression at C6/7 level (Fig. 2D).

The C6/7 apophyseal joint was the only mobile segment maintaining cervical spinal motion. Excessive motion and loading caused the degeneration of disc and facet, which resulted in instability. Anterior spinal interbody fusion with posterior spinous process wiring was planned to decompress the spinal cord and to stabilize the instability.

During general anesthesia, patient's neck was excessively stiff, and mouth was not opened because of the contracture of the temporo-mandibular joint. A tracheostomy was performed to maintain the airway. The trajectory for anterior interbody fusion was interfered with the tracheostomy wound. The anterior approach was abandoned to avoid the tracheostomy site-related complications.

For posterior approach, the pedicle or lateral mass screw was not indicated because of severe osteoporosis. After decompressive subtotal laminectomy and removal of hypertrophic ligamentum flavum, interspinous process wiring with titanium-
A braided cable and integrated bone graft was performed at C6/7 level.

During 6 months of follow-up, pathologic reflexes disappeared and activities of daily living improved. Computed tomography showed well-established bone graft (Fig. 3).

**DISCUSSION**

Juvenile rheumatoid arthritis is also known as juvenile idiopathic arthritis. The prevalence is about 1 per 1,000 children, and the annual incidence is about 1 per 10,000 children. Juvenile rheumatoid arthritis is defined as the presence of joint swelling or 2 of the following: joint tenderness, decreased range of motion, pain on range of motion, or joint warmth for at least 6 weeks without another cause in children younger than 16 years of age. The course, progression, and joint involvements are similar to adult rheumatoid arthritis.

Cervical stiffness is the most common finding, and its occurrence has been reported to be 46% to 60% in patients. Radiological changes are not seen as frequently as in adult rheumatoid arthritis. Subluxation or spontaneous posterior fusion is seen in the late stage of the disease. Neurological complications are also less common than adult type.

Rheumatoid arthritis of the spine was once believed to be a benign disease process. However, many studies have shown that the spectrum of joint erosion, deformity, ensuing neurological deficits, and underlying pathologic mechanisms warrant concern, especially in the cervical spine. The pannus is composed of proliferating fibroblasts and inflammatory cells, which appears as a granulation tissue during the inflammatory process. The pannus produces collagenases and other proteolytic enzymes that are capable of destroying cartilages, ligaments, tendons, and bones. This destructive synovitis leads to ligamentous laxity and bony erosion, which may lead to instability and subluxation in the cervical spine.

Cabot et al. reported the 6 major categories of cervical involvement in severe rheumatoid arthritis: a) atlanto-axial subluxation; b) subluxation; c) upward translocation of the odontoid (cranial settling); d) odontoid erosion; e) apophysial joint fusion; and f) miscellaneous findings of osteoporosis, endplate erosion, and disk space narrowing without osteophytosis.

Generally, neurological symptoms are caused by atlantoaxial subluxation, cranial settling, and subaxial subluxation. Apophysial joint fusion of the cervical spine is a late and uncommon complication of rheumatoid arthritis, which is rather common with ankylosing spondylitis. Classic patterns of ankylosing spondylitis are progressive ossification of spinal ligaments and intervertebral discs as well as ankylosis of facet joints from the sacro-iliac joint to cephalad. The cervical spine involvement is the last manifestation in ankylosing spondylitis, whereas rheumatoid arthritis commonly involves the cervical spine rather than other spine. The mechanism of apophysial joint fusion of the cervical spine in rheumatoid arthritis is still unclear, but findings are similar to those of ankylosing spondylitis.

Most previous studies have focused on cervical instability. However, Cabot et al. reported that 11% of patients with juvenile rheumatoid arthritis showed apophysial joint fusion involving the C2-3 or C1-3 facets and/or spinous processes. However, they described only short-segment cervical fusion, and long-segment cervical fusion was beyond their descriptions. This case is a first-reported case of long-segment cervical fusion, and it is suggested that long-segment cervical fusion may be related with rapid progression of disease. Although calcification of the pannus and the alar ligament indicated previous cervical instability, the patient had not shown any instability symptoms before. Progressing stiff neck and disability might have obscured instability symptoms. It is also suggested that long-segment cervical apophysial joint fusion is the last consequence of disease, following cervical instability, atlanto-axial subluxation, and atlanto-axial arthrodesis.

The 2 major difficulties of cervical operation are stiff neck and osteoporosis. Neck stiffness along with the loss of lordosis, micrognathia, and arthrodissis of the temporo-mandibular joint makes intubation difficult. Laryngeal airway mask, awake-fiberoptic intubation, light source-guided intubation, and many other techniques are tried to overcome the stiff
neck intubation problem. If every effort fails, tracheostomy is the only way to maintain the airway and achieve adequate oxygen saturation. Emergency kits for tracheostomy and crico-
stomy should be prepared. We underestimated this problem. Despite every effort to perform intubation using the afore-
mentioned techniques, adequate oxygen saturation was not maintained. An emergency tracheostomy was required to save the patient.

When the patient has a stiff neck and an arthrodesis of the temporo-mandibular joint, the entrance for anterior approach is narrow. If the tracheostomy is performed, anterior approach is hardly available and troubled with a high risk of wound infection. If such patients are treated with immunosuppres-
sive drugs such as methotrexate, the risk of wound infection is much higher.

Posterior approaches can be performed without an addi-
tional risk in tracheostomy patients and can usually provide adequate stability without anterior approach. However, patients with longstanding rheumatoid arthritis have osteoporosis resulting from chronic inflammation, inactivity atrophy, and steroid use. To prevent fusion failure, selection of appropriate instrumen-
tation is crucial. Pedicle or lateral mass screw is commonly used for a variety of indications to achieve segmental stabilization and fusion. Despite being the most rigid form of poste-
rior instrumentation, pedicle or lateral mass screw sometimes achieve poor initial fixation because of severely compromised screw-bone interface in patients with osteoporosis.

Although sublaminar or interspinous wires have the potential for increased neurological risk, these wires are ideal for use in patients with osteoporosis because fixation points are maxi-
mized and each wire can be sequentially and repeatedly tightened. Recent development of titanium-branched cable enables safer and more reliable stabilization.

CONCLUSION

This case suggests long-segment spontaneous cervical arthro-
desis may be the last manifestation of cervical involvement, which seems to be related with the severity and the rapid progression of juvenile rheumatoid arthritis. The instability of subjacent levels needs surgical stabilization with precautions against stiff neck and severe osteoporosis.

REFERENCES

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