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Management of cervical spinal lesions in rheumatoid arthritis

Abstract We review the management of cervical spinal lesions in rheumatoid arthritis. Surgical treatment for cervical lesions presents several potential problems, such as spinal cord injury during surgery, prolonged and painful postoperative immobilization, poor outcome incompatible with surgical invasiveness, and a high failure rate of arthrodesis. The introduction of spinal instrumentation techniques to surgery for cervical lesions has solved some of these problems. Rigid spinal fixation using screws, rods, and wires has made surgery more reliable, and has freed patients from painful postoperative immobilization methods such as the Halo vest. However, the effects of surgical treatment have not been clearly examined using methods of evidence-based medicine. There is a need for clinical studies of treatment for cervical lesions, in order to help establish better methods for the management of cervical spinal lesions.

Key words Atlantoaxial subluxation (AAS) · Natural history · Rheumatoid arthritis (RA) · Subaxial subluxation (SS) · Vertical subluxation (VS) of the axis

Introduction

The cervical spine is frequently involved in rheumatoid arthritis (RA). The reported incidence of cervical spine involvement in RA varies from 25% to 90%, depending on diagnostic criteria.^{1–5} Such cervical spinal lesions cause not only severe and sometimes intractable neck pain, but also quadri- or paraplegia that may result in severe disability or death. However, surgical treatment for these cervical le-

sions is sometimes decided against for two main reasons: (1) difficulty in differentiating neural symptoms from complex disability caused by multiple joint involvement; (2) the relatively high rate of pseudoarthrosis is incompatible with the great invasiveness and risks of surgery, which include intra- and postoperative death. However, owing to recent advances in medical and surgical management of RA, it is now possible to preserve a high level of function in the extremities in many rheumatoid patients. This has increased the relevance of cervical lesions, because neurological symptoms due to such lesions can compromise preserved or reconstructed joint functions that have been obtained with considerable effort. Several surgical techniques for treating cervical lesions have been developed as a result of advances in spinal instrumentation surgery. There is a need for clear guidelines for the treatment of cervical lesions in rheumatoid patients.

Cervical lesions and clinical presentation

Cervical involvement due to synovitis in the atlantoaxial (central and lateral) and atlantooccipital joints, and bursitis between the odontoid and the transverse ligament, leads to instability of these joints (atlantoaxial subluxation (AAS) and vertical subluxation (VS) of the axis). Subaxial subluxation (SS) results from destruction of the facets, interspinous ligaments, and intervertebral discs.^{6–8}

The clinical presentation of cervical lesions includes neck pain and neurological deficit. Neck pain is reportedly common (40%–88% of RA patients), whereas neurological deficit is rarely reported (7%–34%).³ In addition, studies have indicated that cervical lesions can be lethal.⁹ Mikulowski et al.¹⁰ reported that 10% of RA patients may die of unrecognized medullary brainstem compression. Marks and Sharp¹¹ reported that half of patients with symptoms of spinal cord compression died within 1 year of diagnosis. Redlund-Johnell and Pettersson¹² reported that medullary brainstem compression accounted for 50% to 100% of sudden deaths of RA patients. Thus, cervical

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lesions, especially upper cervical lesions, can be life-threatening for RA patients.

Natural history

Cervical lesions occur during the early stage of RA,¹³ and are reportedly progressive.¹⁴⁻¹⁶ Winfield et al.¹⁴ reported that in a 7.2-year follow-up study, 12% of RA patients developed AAS and 20% developed SS. Smith et al.¹⁶ reported that in a 7.8-year follow-up study, radiographical progression of cervical lesions was detected in 34% of patients. Fujiwara et al.¹⁷ examined patterns of progression of upper cervical lesions based on a transverse study, and reported that upper cervical lesions progress from reducible AAS to irreducible AAS, then to AAS associated with VS, and finally to VS alone. Oda et al.¹⁸ conducted a longitudinal study and reported results similar to those of Fujiwara, noting that a few cases showed rapid progression directly to VS. They also analyzed relationships between the disease subsets proposed by Ochi et al.¹⁹ and the mode of progression of upper cervical lesions, and found that patients in the severely involved subset (e.g., the so-called mutilating type) showed severe progression of upper cervical lesions, whereas most patients in the less involved subset remained at the AAS stage. These findings suggest that prognosis of upper cervical lesions can be predicted by classification of the disease subset, and that such predictions can be used to choose the most appropriate treatment modality for individual patients.

Management of cervical lesions

Treatment for cervical lesions is comprised of conservative and surgical modalities. Conservative treatment includes medical treatment and orthosis. Medical treatment with nonsteroid anti-inflammatory drugs may be useful for pain control in some patients. However, theoretically, medical treatment cannot improve mechanical instability that has developed due to synovitis and enthesopathy. A simple, and therefore frequently chosen, therapy is orthotic treatment using a softplastic collar or a Philadelphia brace. A soft collar such as a plastic collar does not immobilize the atlantoaxial joints biomechanically,²⁰ and can be used for temporary immobilization when neck pain is severe. A rigid orthosis device such as a sternooccipitomandibular immobilization (SOMI) brace can immobilize the cervical spine more strongly, but compliance with rigid orthosis is generally very low, and therefore this is not a realistic alternative. Thus, surgery is the treatment of choice when neurological symptoms and/or intractable neck pain develops.

We now briefly discuss recent advances in spine surgery, focusing on techniques applicable to the rheumatoid cervical spine, details of which are described elsewhere. Atlantoaxial stabilization involves posterior wiring and bone grafting. Because this wire fixation is not mechanically

strong, the procedure requires prolonged rigid external fixation. The rate of nonunion is high for this procedure. Magerl and Seeman²¹ developed a technique for firm fixation of the atlantoaxial joint with a transarticular screw. This strong fixation allows the omission of postoperative external immobilization, and improves the rate of union. Recently, several systems comprised of a rod and screw or a hook and wire have been developed; these systems allow very firm fixation of the cervical spine from the occiput to the thoracic spine. These new techniques have solved some of the problems involved with surgery for rheumatoid cervical spine.

There are three major issues regarding surgical treatment: (1) the clarity of indications of surgery, choice of surgical techniques, and timing of surgery; (2) the ability of surgery to improve the activities of daily living (ADL) and quality of life (QOL), especially in patients with severe joint involvement; (3) the ability of surgery to change the natural course of cervical lesions.

Indications for surgery are (1) severe myelopathy, (2) developing or impending myelopathy, (3) vertebral artery insufficiency, and (4) severe neck pain that cannot be controlled by conservative treatment. In practical terms, it is very difficult to recommend surgery to patients who begin to develop myelopathy. Excessive conservatism can cause patients to miss a chance for timely surgery, but excessive intervention can result in unnecessary surgery.

Although no single factor has been shown to be a reliable indicator of surgery, two factors that are useful in decisions regarding surgical treatment are the space available for the spinal cord (SAC) and classification of the disease subset. The distance between the dorsal margin of the odontoid and ventral margin of the posterior arch of the atlas (SAC) is a roentgenographical parameter of myelopathy. When SAC becomes ≤ 14 mm, the possibility of myelopathy increases.²² As described above, in the severely involved subset, upper cervical lesions tend to progress rapidly and severely. For patients with these factors, surgical treatment may be beneficial, even at the early stage of an upper cervical lesion.

Can surgery improve ADL and QOL of rheumatoid patients?

To the best of my knowledge, no comparative study of conservative and surgical treatment has been conducted. A major issue in surgical treatment is the evaluation criteria of surgical results. A functional evaluation of a patient with multiple joint involvement is very difficult, and many authors used a relatively simple evaluation system.²³ These evaluation systems are sometimes not sufficiently sensitive to detect postoperative changes.

There have been many reports on the results of surgical treatment.²⁴⁻³⁸ The introduction of spinal instrumentation surgery has improved the success rate of (occipito-) atlantoaxial arthrodesis by 90%–95%. Therefore, the use of spinal instrumentation is generally recommended. However, spi-

nal instrumentation surgery entails risks such as injury of the vertebral artery by screws, and the strength of the fixation is not always guaranteed in rheumatoid spines because the bone quality is usually poor. Thus, surgeons must familiarize themselves with these techniques and choose a procedure that is appropriate to local bone quality.

Pain relief is generally achieved with surgery, but the neurological results are inconsistent. The neurological results correlate with the severity of preoperative neurological symptoms. As mentioned above, an evaluation of surgical results is very difficult because the function of patients with myelopathy is compromised by the involvement of the joints of the extremity. An accurate assessment of the real value of surgery for myelopathy due to cervical lesions requires the establishment of a better evaluation system.

The choice of a surgical procedure for subaxial subluxation is still controversial. Surgical techniques applicable to this condition are anterior fusion (with or without instrumentation) or posterior decompression (mostly laminoplasty; with or without fusion).²⁹

Can surgery change the natural course of cervical lesions?

The number of long-term follow-up studies of surgery for the rheumatoid spine has been increasing. Some studies include life prognoses as well as neurological and radiological results.^{39,40} Patients with (occipito-) atlantoaxial fusion often develop SS, usually at levels adjacent to fused segments. The more severely involved subset tends to develop SS, which requires additional surgery. However, one-third to one-quarter of patients who undergo surgical treatment for cervical lesions die within 5 years of the operation, and the significance of surgical treatment for these patients must be reevaluated.

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