CASE REPORT

Acquired Chiari Malformation Secondary to Atlantoaxial Vertical Subluxation in a Patient With Rheumatoid Arthritis Combined With Atlanto-occipital Assimilation

—Case Report—

Yuiko KIMURA,1 Atsushi SEICHI,2 Akira GOMI,1 Masahiro KOJIMA,2 Hirokazu INOUE,2 and Atsushi KIMURA2

Departments of 1Neurosurgery and 2Orthopedics, Jichi Medical University, Shimotsuke, Tochigi

Abstract

A 65-year-old woman with a history of rheumatoid arthritis presented with a rare case of acquired Chiari malformation secondary to atlantoaxial vertical subluxation, associated with congenital atlanto-occipital assimilation. Syringomyelia and tetraparesis improved immediately after posterior fossa decompression and simultaneous occipito-cervical junction fusion. The progression of acquired Chiari malformation is not well known. We concluded that coexisting assimilation accelerated crowded foramen magnum following atlantoaxial vertical subluxation and induced acquired Chiari malformation over the course of a few years.

Key words: atlantoaxial subluxation, atlas assimilation, Chiari malformation, rheumatoid arthritis, syringomyelia

Introduction

Most spinal manifestations of progressive rheumatoid arthritis (RA) are observed at the craniocervical junction. Radiographic abnormalities of the cervical spine are observed in as many as half of patients with RA.4) However, few reports have described Chiari malformation with syringomyelia following progression of atlantoaxial subluxation.8,9) We describe a rare case of acquired Chiari malformation secondary to atlantoaxial vertical subluxation in a patient with RA combined with congenital occipitoatlantal assimilation.
Case Report

A 62-year-old woman with a 4-year history of RA presented with progressive neck pain. Magnetic resonance (MR) imaging revealed mild atlantoaxial subluxation without syringomyelia or tonsillar herniation at that time (Fig. 1). The patient underwent noninvasive treatment using a cervical brace. She was admitted to hospital 3 years later following progressive tetraparesis starting 6 months earlier. Repeat MR imaging identified progression of atlantoaxial vertical subluxation with syringomyelia at the C2–C5 levels, anterior cervico-medullary compression, and tonsillar herniation (Fig. 2). Reconstructed computed tomography showed occipitoatlantal assimilation and upward displacement of the odontoid process to 9 mm above Chamberlain's line (Fig. 3).

The patient underwent occipitocervical junction fusion with C2 pedicle screws and iliac bone grafting using Olerud Cervical® (NordOpedic, Uppsala, Sweden), along with foramen magnum decompression and C1 laminectomy with the aim of improving instability of the atlantoaxial joint and syringomyelia. Limited foramen magnum decompression up to the inferior nuchal line was required to keep the area of the occipital bone available for implant fixation using a loop rod with screws and/or cable instrumentation (Fig. 4). Ultrasonography confirmed pulsation of the tonsil under direct vision and opening of the dorsal subarachnoid space after minimum suboccipital craniotomy and durotomy. Occipitoaxial fixation in a position of mild extension was performed using Olerud Cervical® with two cranial screws and C2 pedicle screws (Fig. 5). After iliac bone grafting, duraplasty was performed with trapezius fascia.

Almost complete relief from tetraparesis was achieved postoperatively. Postoperative MR imaging demonstrated...
Chiari Malformation Associated With RA

685

improvement of syringomyelia and tonsillar herniation (Fig. 6A). As of 2 years postoperatively, no recurrence of symptoms or breakage of implants has occurred. Cervico-medullary angle on MR imaging was 140° before surgery, 151° soon after surgery, and 145° at 2 years after surgery (Figs. 2 and 6). Although slight loss of the cervico-medullary angle was apparent, only a small trace of syringomyelia was observed on follow-up MR imaging (Fig. 6B).

Discussion

Atlantoaxial subluxation is common in patients with progressive RA. However, syringomyelia complicated by RA of the spine is rare. In the few cases that have been described, a casual link to coexisting syringomyelia was concluded to exist. Chiari malformation is a disorder of uncertain origin that has been conventionally defined as downward herniation of the cerebellar tonsil through the foramen magnum, attributable to overcrowding of the hindbrain in a posterior cranial fossa. In some series of Chiari malformation, syringomyelia developed in 58–65% of cases. The most frequent types of Chiari malformation have been considered to result from mesodermal defects. Acquired forms of Chiari malformation are often seen after lumbar shunting procedures, such as lumbo-peritoneal shunts indicated for the treatment of certain forms of communicating hydrocephalus, or after drainage or serial spinal taps for the treatment of postoperative or post-traumatic cerebrospinal fluid leakage. In these cases, a normalized posterior fossa is seen with a “crowded” foramen magnum.

The present case illustrates acquired Chiari malformation following progression of atlantoaxial vertical subluxation. Acquired Chiari malformation is not usually induced by only atlantoaxial subluxation in RA patients, whereas craniocervical junction anomaly including occipitoatlantal assimilation is well known to carry a potential risk of Chiari malformation due to progressive basilar invagination and irreducibility. Comprehensive preoperative imaging is necessary to achieve an appropriate diagnosis and create a surgical plan. In this case, we believe that the coexisting congenital occipitoatlantal assimilation caused relatively rapid acceleration of crowding of the fossa following severe vertical subluxation and resulted in tonsillar herniation with syringomyelia.

As a technical note regarding the surgical treatment for this patient, foramen magnum decompression was planned for improvement of syringomyelia with Chiari malformation, but simultaneous stabilization was necessary to prevent worsening of atlantoaxial subluxation. Surgical treatment was performed for 5 cases of Chiari malformation associated with atlantoaxial dislocation, and the importance of performing posterior decompression followed by occipito-cervical junction fusion via plate and screw fixation was emphasized. However, an overly large craniotomy prevents implant fixation at the midline of the occipital bone. We performed craniotomy with a minimum area using ultrasonography to confirm appropriate decompression with opening of the dorsal subarachnoid space, achieving successful decompression and stabilization.

The necessity for reducing vertical subluxation in craniocervical junction fusion is controversial. We did not try to reduce upward migration of the odontoid process and decreases in cervico-medullary angle during follow up were minor (Figs. 2 and 6). Meticulous follow up is needed to observe progression of adjacent segment degeneration following fusion surgery, malalignment, and effects of residual anterior compression of the odontoid process.

Conflict of Interest

No benefits in any form have been or will be received from any commercial party related directly or indirectly to the subject of this article.
References


Address reprint requests to: Atsushi Seichi, MD, Department of Orthopedics, Jichi Medical University, 3311-1 Yakushiji, Shimotsuke, Tochigi 329-0498, Japan.

*e-mail: seichi-spine@jichi.ac.jp*