CASE REPORT doi: 10.2176/nmc.cr2012-0421
Neurol Med Chir (Tokyo) 53, 902–906, 2013 Online October 21, 2013

Intraoperative Computed Tomography for Cervicomedullary Decompression of Foramen Magnum Stenosis in Achondroplasia: Two Case Reports

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Abstract

The authors report two cases of cervicomedullary decompression of foramen magnum (FM) stenosis in children with achondroplasia using intraoperative computed tomography (iCT). A 14-month-old girl with myelopathy and retarded motor development, and a 10-year-old girl who had already undergone incomplete FM decompression was presented with myelopathy. Both patients underwent decompressive suboccipitalcraniectomy and C1 laminectomy without duraplasty using iCT. It clearly showed the extent of FM decompression during surgery, which finally enabled sufficient decompression. After the operation, their myelopathy improved. We think that iCT can provide useful information and guidance for sufficient decompression for FM stenosis in children with achondroplasia.

Key words: achondroplasia, cervicomedullary decompression, foramen magnum stenosis, intraoperative computed tomography

Introduction

Achondroplasia is an autosomal-dominant, inherited dwarfism syndrome characterized by short stature, macrocephaly, and shortening of the extremities, resulting from gene mutation in fibroblast growth factor receptor located on chromosome 4.[7] Its incidence ranges from 0.5 to 1.5 in 10,000 live births;[4,10] however, approximately 75% of cases are due to a new, sporadic mutation.[4] Patients with achondroplasia manifest cervicomedullary compression or hydrochephalus caused by foramen magnum (FM) stenosis, resulting in respiratory failure or neurological deficits, as well as sudden death in some cases.[1–3,6,11–14,20] Therefore mortality rates are relatively high, and can approach 7.5% in the 1st year of life and 2.5% between 1 and 4 years of age.[5] Patients with FM stenosis in achondroplasia have undergone FM decompression;[1,9,15,19–21] however, in some cases of duraplasty, cerebrospinal fluid (CSF) leakage occurred, which led to meningitis, and ventriculoperitonial shunting was eventually required.[1] Recent reports described that symptomatic FM stenosis can be successfully treated with FM decompression without duraplasty, with significant clinical benefit and minimal morbidity using intraoperative ultrasonography to assess residual compression.[2,21] In this report, we present two cases of FM decompression for children with achondroplasia using intraoperative computed tomography (iCT) to assess residual bone compression without duraplasty, and describe the usefulness of iCT for this surgery.

Received December 6, 2012; Accepted January 28, 2013

Case Report

Case 1: A 14-month-old girl with achondroplasia inherited from her mother presented with retarded motor development. Neurological examination showed severe myelopathy with positive ankle clonus and hyperreflexia. She could neither stand nor walk by herself. Head computed tomography (CT) demonstrated mild hydrocephalus and macrocrania. Cervical CT and magnetic resonance (MR) imaging showed the horizontal orientation of the posterior rim of the FM, causing compression of the medulla (Fig. 1A, B).

FM decompression and C1 laminectomy were performed using iCT. Just before the operation, iCT showed an overgrown opisthion, including the posterior rim of the FM (Fig. 2A). After drilling down the posterior rim of the FM using a diamond-tipped drill and removal of a thickened fibrous epidural band, iCT clearly showed incomplete decompression of the FM (Fig. 2B). We found incomplete osteotomy of the right side of FM as iCT had shown (Fig. 2C), and drilled down the residual bone to

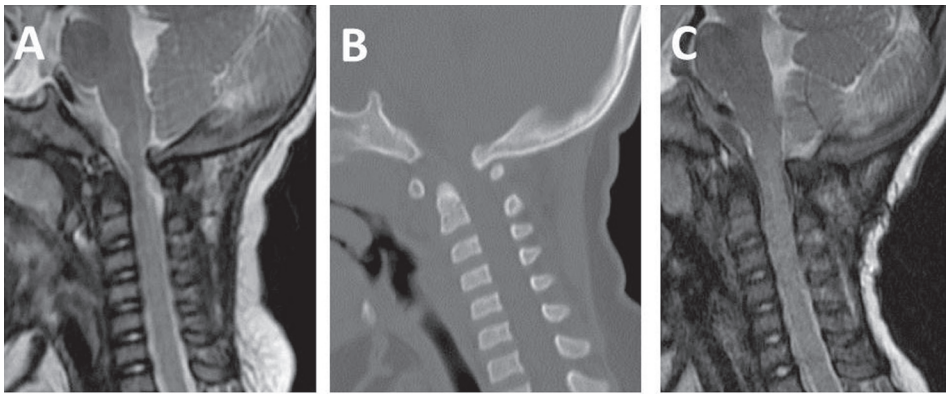

Fig. 1 A: Sagittal T_2-weighted magnetic resonance (MR) imaging before operation showing compression of the medulla at the foramen magnum. B: Computed tomography (CT) before operation showing the horizontal orientation of the posterior rim of the foramen magnum, causing kinking of the cervicomedullary junction. C: Sagitt T_2-weighted MR imaging after the operation showing decompression of the medulla at the foramen magnum.

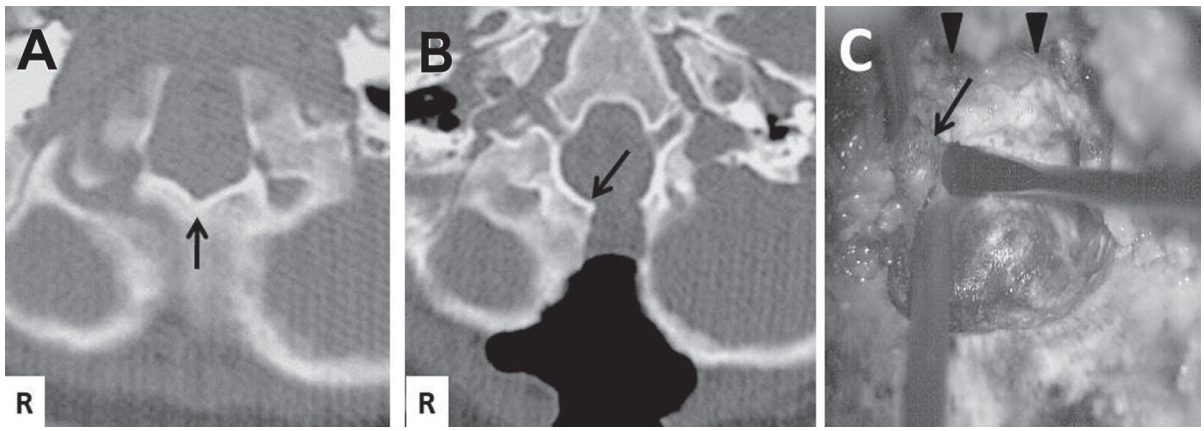

Fig. 2 A: Intraoperative computed tomography (CT) before surgery demonstrating the horizontal orientation of the posterior rim of the foramen magnum (*arrow*). B: Intraoperative CT demonstrating incomplete removal of the posterior rim of the foramen magnum, especially on the right side (*arrow*). C: Photograph just after intraoperative CT showing incomplete decompression of the foramen magnum on the right side (*arrow*). C-1 arch has been removed as well as the thick epidural band (*arrowheads*).

achieve sufficient decompression. After additional drilling, we could see good pulsation of CSF over the thin dura, and it was believed that duraplasty was unnecessary.

Her postoperative course was uneventful. Postoperative MR imaging demonstrated decompression of the medulla (Fig. 1C). Her myelopathy has improved and she can walk by herself. Hydrocephalus has been arrested for 2 years postoperatively.

Case 2: A 10-year-old girl with achondroplasia presented with numbness of her hands and feet. When she was 1 year old, she had undergone FM decompression at another hospital. On admission to our hospital, neurological examination revealed myelopathy with positive ankle clonus, hyperreflexia and spastic gait. Cervical MR imaging showed the horizontal orientation of the posterior rim of the FM, causing compression of the medulla (Fig. 3A). Head CT demonstrated macrocrania without hydrocephalus and incomplete removal of the posterior rim of FM (Fig. 3B, C).

In this case, FM decompression and C1 laminectomy were performed using iCT with a navigation system (Fig. 4A). Just before the operation, iCT showed an overgrown opisthion and incomplete decompression of FM at her first surgery (Fig. 4B). After drilling down the posterior rim of the FM and removing a thickened fibrous epidural band, we noted the edge of the occipital bone and checked the extent of decompression using the navigation system, as the posterior rim of the FM was extremely horizontal and deep. Therefore, we could not confirm the exact location using the pointer of the navigation system; however, iCT clearly showed sufficient removal of the occiput at FM (Fig. 4C). We were assured of cervicomedullary decompression by iCT and duraplasty was not performed.

Her postoperative course was uneventful. Postoperative MR imaging demonstrated sufficient decompression of the medulla (Fig. 3D). Her myelopathy has improved.

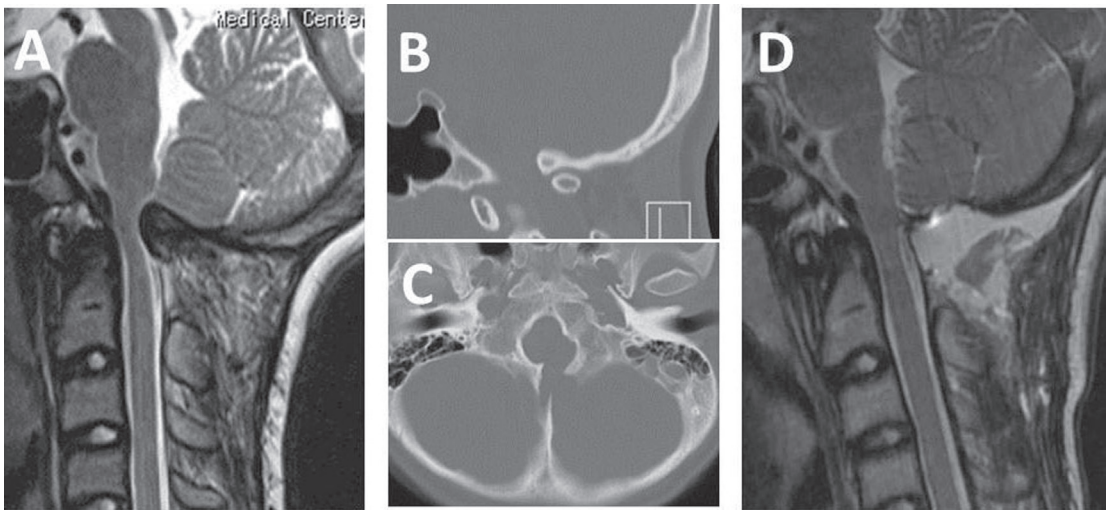

Fig. 3 A: Sagittal T_2-weighted magnetic resonance (MR) imaging before the operation showing severe compression of the medulla. B: Computed tomography (CT) before operation showing the horizontal orientation of the posterior rim of the foramen magnum, causing severe stenosis of the cervicomedullary junction. C: CT demonstrated macrocrania showing incomplete removal of the posterior rim of the foramen magnum. D: Sagittal T_2-weighted MR imaging after operation showing sufficient decompression of the medulla.

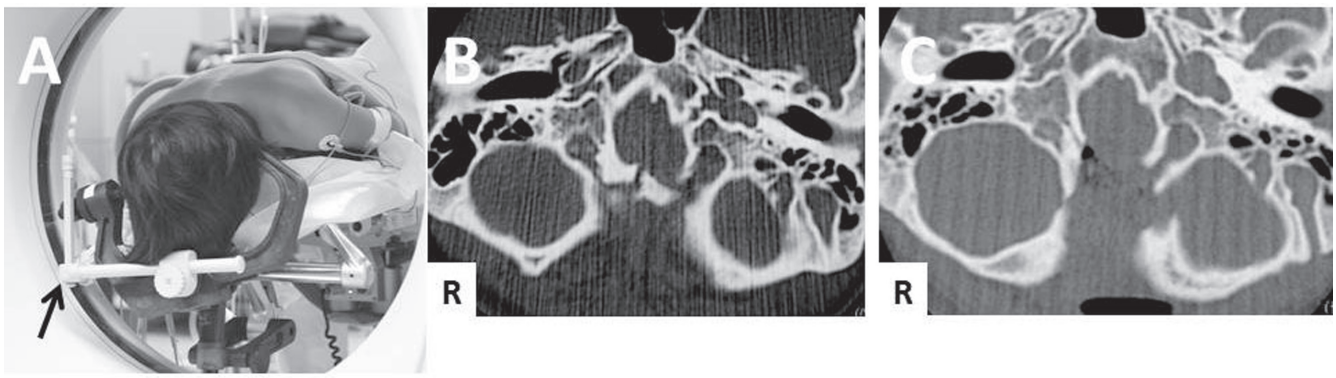

Fig. 4 A: Photograph showing preoperative setting of intraoperative computed tomography (CT) with the navigation system (*arrow*) just before surgery. B: Intraoperative CT just before surgery demonstrating the horizontal orientation of the right posterior rim of the foramen magnum, causing kinking of the cervicomedullary junction. C: Intraoperative CT demonstrating the removal of the posterior rim of the foramen magnum.

Discussion

Achondroplasia patients with clinically significant cervicomedullary compression have undergone FM decompression and upper cervical laminectomy.[1,9,15,19–21] Invariably, the posterior rim of the FM is extremely thickened, and its orientation is more horizontal than usual,[1,8–10,15,19–21] making adequate bone decompression difficult. Once bone removal using the high-speed drill is completed, the thickened fibrous tissue band should be carefully separated from the underlying dura mater.[1,9,15,19–21]

Duraplasty for achondroplasia has been controversial. Previously duraplasty was performed to release dural constriction and allow free CSF flow.[1,15] In a previous study from Johns Hopkins University, Baltimore, the first author routinely performed duraplasty for achondroplasia patients with cervicomedullary compression,[1] however, in a recent report from their institution, Bagley et al. described that duraplasty is unnecessary to decompress the suboccipital region adequately, and they have stopped routinely performing duraplasty considering CSF leakage and infection.[2] They suggested that the use of intraoperative ultrasonography to assess residual compression at the time of surgery aids in the decision

of whether to open the dura mater. Visualization of adequate CSF space on axial and sagittal ultrasound images can eliminate the need to open the dura and for duraplasty.

In FM decompression for achondroplasia, adequate bone removal of FM is one of the essential steps to improve cervicomedullary compression and CSF flow.[1,2] To ensure an adequate decompression of FM, accurate removal of the posterior lip of FM and C1 laminectomy should be performed; however, occipital bone removal and FM decompression with achondroplasia seem to be difficult for the following reasons. First, the posterior rim of the FM is extremely thickened and, moreover, its orientation is more horizontal and deeper than usual.[1,8–10,15,19–21] Second, most patients with FM stenosis in achondroplasia are babies or infants,[1–3,8–11,14,15,19–21] and their skeleton at the cervicomedullary junction is very small.

ICT is considered to be helpful in neurosurgery, and especially has advantages in skull base and spinal surgery because these procedures mainly involve osseous structures.[16–18,22] Usually, iCT can clearly demonstrate osseous structures without a constructive shift during surgery; therefore, we think that iCT is appropriate and useful in surgery for FM stenosis in achondroplasia. In our case 1, iCT clearly demonstrated the actual removal of occipital bone and C1 and incomplete decompression of FM during surgery, which could lead to sufficient decompression of FM. In our case 2, iCT was superior to the navigation system to assess residual bone compression. Although we could not confirm the sufficient removal of occipital bone by the navigation system, iCT clearly showed the actual extent of bone removal of FM. During surgery for FM stenosis in achondroplasia, it seemed to be difficult to locate the horizontal and deep position of the occipital bone using the pointer of the navigation system.

Radiation exposure from CT scans should be considered, because most patients with achondroplasia are babies or infants who are more radiosensitive than adults. Therefore, we think iCT should not be a routine process for this surgery. We recommend iCT when surgeons cannot confirm dural pulsation macroscopically or adequate CSF flow by ultrasonography. Once will be enough to check the bone removal of FM.

After the removal of bone structures, the thickened fibrous tissue band should be certainly removed, and we agree that intraoperative ultrasonography is useful to assess residual compression during surgery.[2] In addition, we think that iCT can provide surgical benefits and help neurosurgeons who are inexperienced in cervicomedullary decompression of achondroplasia to perform adequate decompression of FM. This might be a valuable option and has the potential to reduce repeat operations for FM decompression in achondroplasia.

Conflicts of Interest Disclosure

The authors report no conflict of interest concerning the materials and methods used in this study or the findings specified in this article. All authors who are members of The Japan Neurosurgical Society (JNS) have registered online Self-reported COI Disclosure Statement Forms though the website for JNS members.

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