Endoscopic Endonasal Approach for Urgent Decompression of Craniovertebral Junction in Syringobulbia

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Key words
- CVJ stability
- Endonasal approach
- Endoscopic
- Odontoidectomy
- Syringobulbia

Abbreviations and Acronyms
3D: 3-Dimensional
CSF: Cerebrospinal fluid
CT: Computed tomography
CVJ: Craniovertebral junction
MRI: Magnetic resonance imaging

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Citation: World Neurosurg. (2019) 130:499-505. https://doi.org/10.1016/j.wneu.2019.07.004

Journal homepage: www.journals.elsevier.com/world-neurosurgery

Available online: www.sciencedirect.com

BACKGROUND: **Syringobulbia is an uncommon lesion that occurs in the central nervous system; it is often defined as a pathologic cavitation in the brainstem. The cases with partial blockage of the cerebrospinal fluid pathways at the level of the foramen magnum are more common and the most important group. The most common treatment of syringobulbia is craniovertebral decompression.**

CASE DESCRIPTION: **This paper reports a case of a symptomatic syringobulbia in which an urgent endoscopic endonasal approach to the craniovertebral junction (CVJ) was done to limit bulbo-medullary compression and rapid neurologic deterioration. A 69-year-old man was admitted to the hospital because of acute onset of dysphonia, dysphagia, imbalance, and vomiting. Magnetic resonance imaging revealed a cystic lesion in the brainstem, suggestive of a syringobulbia in Klippel Feil syndrome with CVJ stenosis.**

CONCLUSIONS: **This case report details the successful use of endoscopic endonasal anterior decompression to treat syringobulbia, and adds to the growing literature in support of the endonasal endoscopic approach as a safe and feasible means for decompressing the craniocervical junction, even in the setting of urgency. However, prudent patient selection, combined with sound clinical judgment, access to instrumentation, and intraoperative imaging cannot be overemphasized.**

INTRODUCTION

Syringobulbia is an uncommon lesion that occurs in the central nervous system; it is often defined as a pathologic cavitation in the brainstem. There are several possibilities for the pathology and little agreement about what exactly is syringobulbia.

The cases with partial blockage of the cerebrospinal fluid (CSF) pathways at the level of the foramen magnum are the most common and most significant group.

The most common treatment of syringobulbia is craniovertebral junction (CVJ) decompression. It consists of a small craniectomy that decompresses the tonsils usually by removing the arachnoid and usually draws away part of the tonsils' parenchyma. In other situations, the syrinx is shunted into peritoneal, pleural, or atrial cavities if needed.

Nowadays, the endoscopic endonasal approach allows surgeons to have greater control of the CVJ, from the inferior third of the clivus to the middle-inferior third of the C2 vertebral body. The hard palate is the only aspect that deters the caudal limit of the approach.[1,2]

In this paper, the first case, to our knowledge, of symptomatic syringobulbia in which an urgent 3-dimensional (3D) endoscopic endonasal approach to the CVJ was done to limit bulbo-medullary compression is reported.

CASE DESCRIPTION

A 69-year-old man with a medical history of hypertension was admitted to the emergency department because of an acute onset of dysphonia, dysphagia, imbalance, and vomiting. Clinical examination showed hyperreflexia in his lower limbs, severe tetraparesis (strength 1 of 5 upper limbs and 2 of 5 lower limbs), bilateral positive cutaneous plantar reflex, dysphonia, and dysphagia. Magnetic resonance imaging (MRI) revealed a cystic lesion in the brainstem, suggestive of syringobulbia in Klippel Feil syndrome with CVJ stenosis (Figure 1). The patient was hospitalized and his dysphagia got worse in the following days causing ab ingestis pneumonia with consequent acute respiratory failure. He was transferred to our hospital as a referral center for skull base pathologies, where, considering the acute onset of symptoms and the sudden worsening of neurologic status, we decided to perform a 3D endoscopic approach to decompress the CVJ. The patient was maintained under intubation during the following days. Postoperative imaging demonstrated complete anterior resection of C1 and of the odontoid peg of C2 with brainstem decompression (Figure 2). After anterior decompression, a posterior C1-2 fixation was done with the aim of treating craniocervical instability because of wide decompression and the removal of the C1 arch (Figure 3).

The patient was extubated after posterior fixation; however, a tracheostomy was

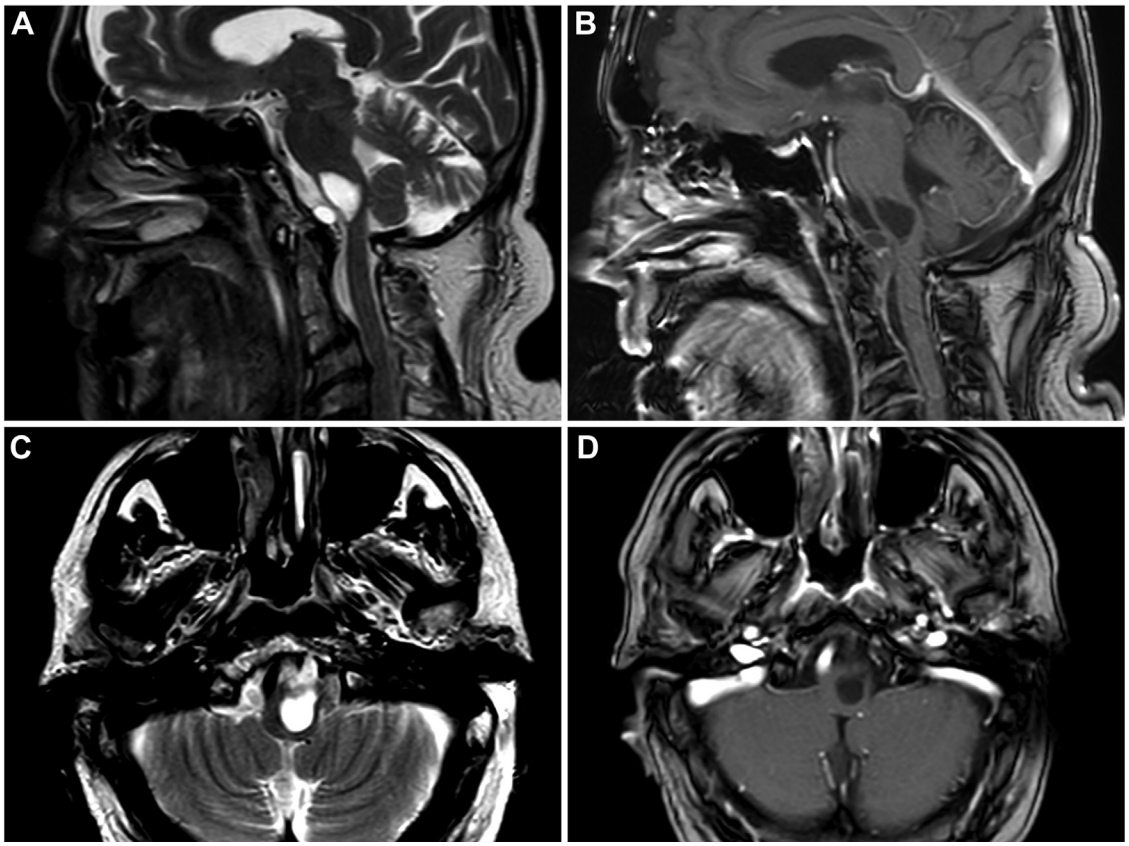

Figure 1. Preoperative magnetic resonance imaging showing syringobulbia because of craniovertebral junction stenosis: (**A**) T2 sagittal plane, (**B**) T1 sagittal plane, (**C**) T2 axial plane, and (**D**) T1 axial plane.

performed because of lower cranial nerve impairment. Postoperative somatosensory evoked potentials and motor evoked potentials, performed at 1 month after decompression, showed improvement of the pyramidal tracts electrical conduction, but damage on the right corticospinal tract was still present. Clinically, he showed very slow improvement in the strength of the 4 limbs, in particular in the lower limbs, reaching 3 of 5 after 1 month from surgery.

The patient started a rehabilitation program and speech therapy, and he began to show progressive neurologic improvement. In particular, he was able to move his arms and legs, but he had a motor impairment on his right side. Speech production was difficult to evaluate because of the tracheostomy. MRI performed 1 month after the operation showed a huge decompression of the CVJ and widening of the cisterna magna compared with preoperatory imaging. It was thought that there was an opening of the syringobulbia into the cisterna magna because of the effect of the decompression.

The last MRI, performed 3 months after the surgeries, showed a successful reduction of the syringobulbia (Figure 4). The patient showed a significant improvement in his neurologic status. At 3 months from surgery, the patient was able to walk and move with support from a walker, and speech production was regular and fluent.

Surgical Procedure

The patient underwent an endonasal image-guided full 3D endoscopic approach. A preoperative computed tomography (CT) scan which used the hard palate appropriately suggested the achieved surgical access via an endonasal approach.[1-5] A CT angiography study was used to assess the parapharyngeal carotid course.

The procedure was performed under the assistance of neurophysiologic monitoring. Data of somatosensory evoked potentials and motor evoked potentials were collected for the patient. In particular, the data showed baseline alterations of the pyramidal tract.

The surgical team was composed of a neurosurgeon and an ear, nose, and throat surgeon. All surgeries were done with a 3D endoscope (Visionsense III Ltd., Petach-Tikva, Israel). Everyone in the surgical theater wore passive polarized glasses to obtain a stereoscopic view.

The patient was placed in a supine position with his head fixed in a 3-pin headholder (Mayfield System [Integra LifeSciences Corporation, Cincinnati, Ohio, USA] or Infinity Skull Clamp System [Integra LifeSciences Corporation]). We positioned his head slightly tilted to the left on the coronal plane and slightly flexed to slide up the dens. Optic neuronavigation was acquired. The patient's

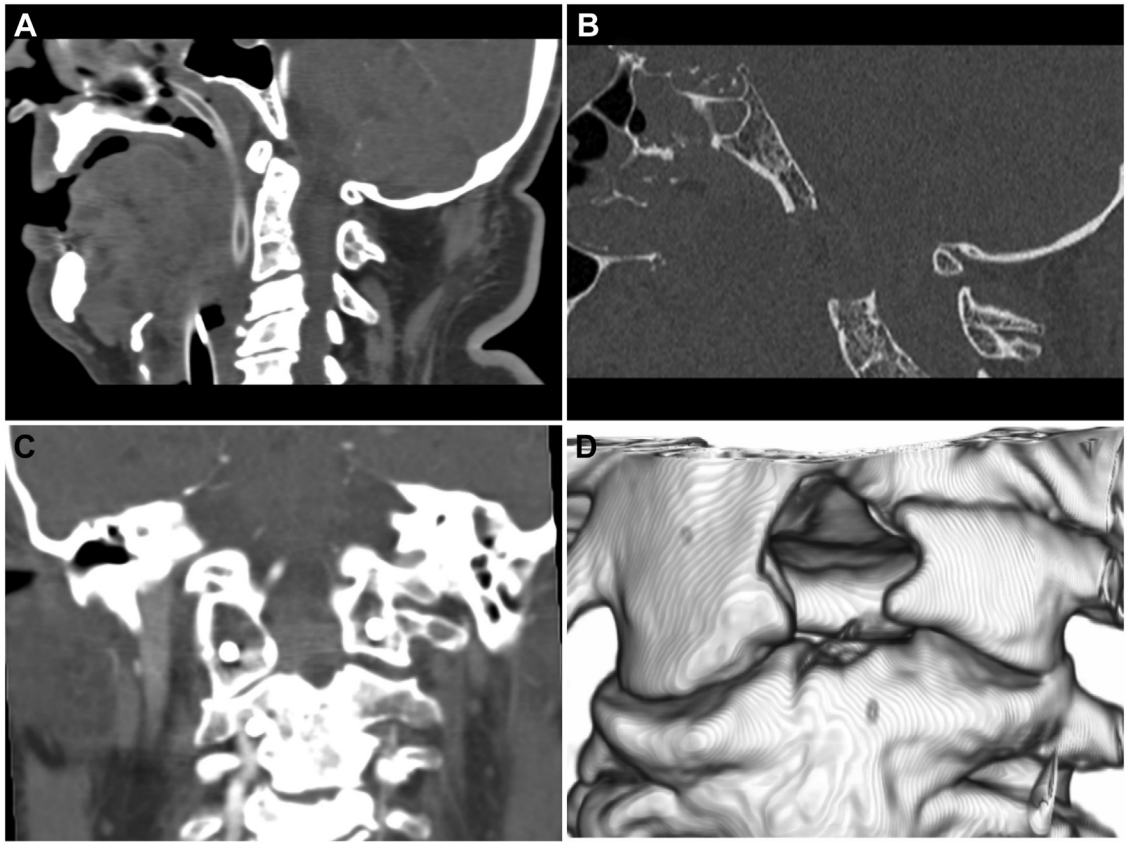

Figure 2. (**A**) Preoperative computed tomography (CT) scan. (**B** and **C**) Postoperative CT scan showing the decompression obtained by the removal of the odontoid peg and of the anterior arch of C1. (**D**) Vertebral 3-dimensional reconstruction of the postoperative CT scan shows the odontoidectomy.

head position enabled surgeons to stand comfortably while maintaining a good view of the stereoscopic information presented on the 3D monitor. Nasal cavities were prepared preoperatively with 2 mL of 10% mepivacaine with adrenaline, which was applied topically with patties. A binostril 4-hand technique was used.[6]

The inferior margin of the middle turbinate, the nasopharynx, and the eustachian tubes were the surgical landmarks that led surgeons to the craniocervical junction. Once the surgeons entered the right nostril, the inferior and middle turbinate were displaced laterally to widen the surgical corridor. Turbinates are not usually removed to preserve a physiologic airflow. The inferior margin of the middle turbinate was then followed until the nasopharyngeal cavity was entered. The posterior third of the nasal septum was removed to obtain a wider operative field using both nostrils.

The junction between the clivus and the atlas was grossly defined by a line connecting the eustachian tubes and checked with the neuronavigation system. The anatomic boundaries of the surgical field were defined by the floor of the sphenoidal sinus superiorly, the upper part of the oropharynx inferiorly, and the eustachian tubes and Rosenmuller fossa laterally. The stealth system provided significant help identifying the patient's anatomy at this point, especially concerning the vertical tracts of the internal carotid arteries.

An inverted, U-shaped flap of nasopharyngeal mucosa and muscular layers was harvested with the laser (CH fiber laser [Dornier MedTech, Munich, Germany]). The eustachian tubes had to be considered as the most lateral margin of the incision because they identified the parapharyngeal segment of the carotid artery. The flap was harvested from the inferior third of the clivus superiorly to the inferior edge of the C2 vertebral body inferiorly. The lateral exposure involved the lateral masses of the C1 vertebra bilaterally that were medial to the eustachian tubes.

The skeletonization of the anterior arch of C1 and of the odontoid process was then carried out in a subperiosteal fashion. We resected the anterior longitudinal ligament to complete the exposure of the atlantoaxial articulation; therefore, the articulation was skeletonized.

After we checked the correct surgical position with the navigation system, a 3-mm coarse diamond burr was used to remove the anterior arch of the C1 vertebra and then to enter the anterior cortex of the odontoid. An ultrasonic bone curette was then used to remove the tip of the odontoid and the base, and finally, the residual shell of the odontoid could be removed by sectioning apical and alar ligaments and

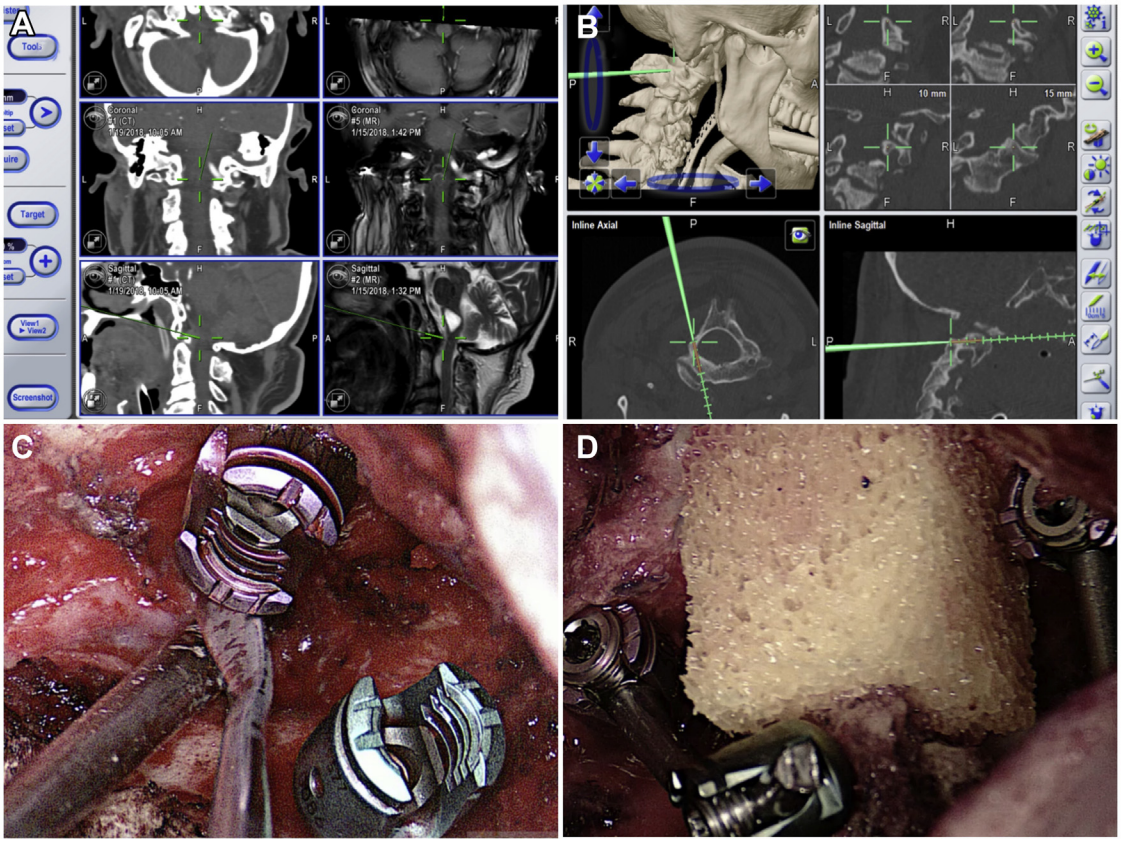

Figure 3. (**A**) Intraoperative navigation system showing the caudal edge of the bone removal on computed tomography scan. (**B**) Intraoperative navigation system use to place the screw. (**C**) Screw on C1-2. (**D**) Cube-shaped cancellous cadaveric bone graft was placed in the translaminar C1-2 space.

separating the process from adhesions to surrounding tissues (Figure 5).

At the end of the procedure, image guidance was helpful to assess the effectiveness of decompression by showing the extent of bone removal and the visualization of the transverse ligament (Figure 5).

Finally, the reverse U mucosal nasopharyngeal flap harvested at the beginning of surgery was repositioned and fixed with fibrin glue. A Foley catheter was held in place for 3 days to compress the mucosal flap.

A posterior approach was performed after the anterior phase. A C1-2 Harms-type fixation was planned. In particular, this included bilateral C1 lateral mass screws, right C2 pedicle screw, and left C2 laminar screw. The C2 left screw was planned with the laminar approach because the narrow corresponding pedicle was too narrow.

The patient was placed in a prone position with a Mayfield head clamp. Intraoperative monitoring of motor evoked potentials and sensory evoked potential was available throughout surgery. Then, an intraoperative CT scan was acquired to use a spine navigation system (Brainlab, Munich, Germany). All the screws were placed with the help of the aforementioned navigation system to maximize bone purchase and pursue safety in the degenerate bone (Figure 3). Screw size was 3.5 mm diameter and 30 mm length for the C1 screws and the left C2 screw, and 3.5 × 20 mm for the right C2 screw. After the rod was placed, cube-shaped cancellous cadaveric bone grafts were set in the translaminar C1-2 space to increase fusion chances (Figure 3).

DISCUSSION

Syringobulbia was first reviewed in 1932 by Jonesco Sisesti, and his work was republished in 1986. He restricted his account to syringobulbia clefts and described such syringobulbia as normally associated with syringomyelia. However, syringobulbia in isolation has not been described. There is a long-established tradition regarding syringobulbia as a developmental lesion with the inference that it may be related to dysraphism. This idea was concluded by pathologists working in an era before modern imaging and surgery. More recently, a hydrodynamic approach has proved helpful in understanding and managing syringomyelia, syringobulbia, and related diseases.[7]

Syringomyelia is not a disease, but is a condition with many possible causes. There are 2 main groups of cases, those in which the etiology is at the foramen magnum level and those primarily with spinal abnormality. The cases with partial blockage of the CSF pathways at the level of the foramen magnum are the most common and the most important group. These may be called hindbrain-related cases. In fact, syringomyelia is

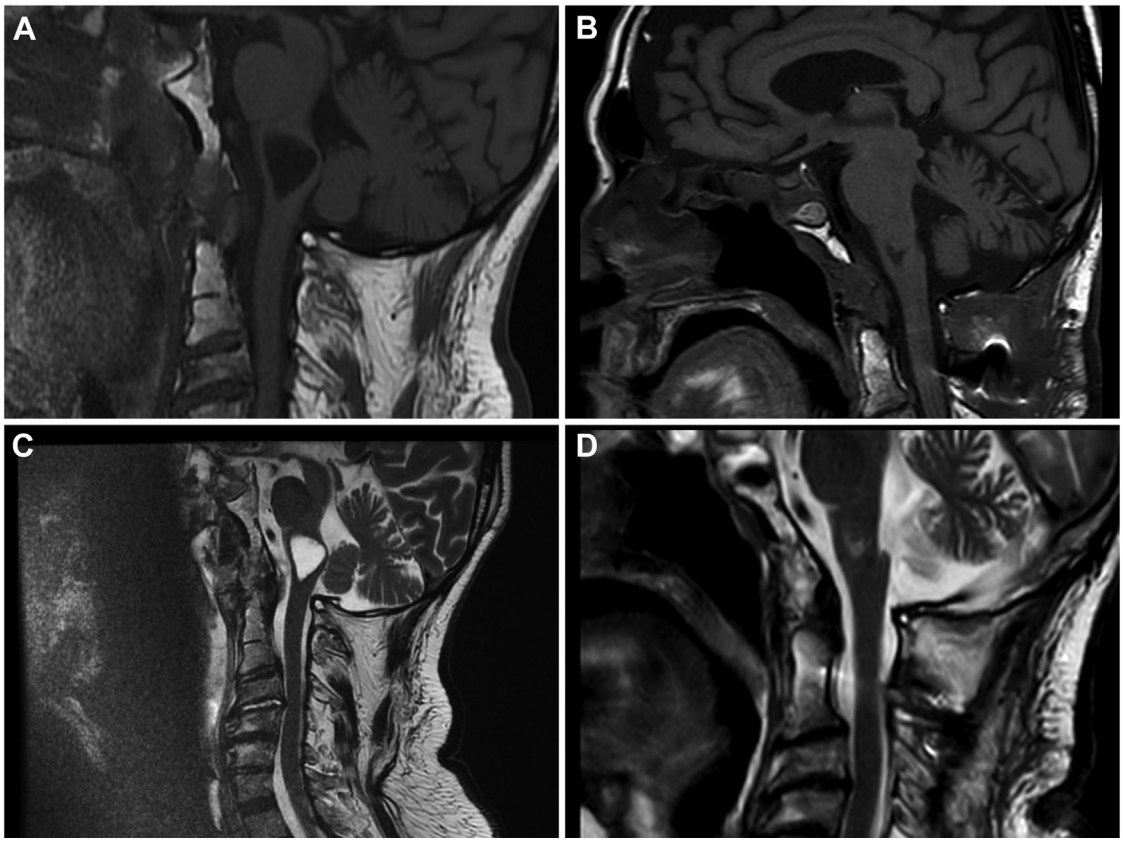

Figure 4. (**A** and **C**) One-month postoperative magnetic resonance imaging (MRI) control scan. (**B** and **D**) Three-month postoperative MRI control scan showing great reduction of the syrinx.

frequently accompanied with an extramedullary lesion at the foramen magnum, particularly a Chiari I malformation. Although syringomyelia is associated with foramen magnum obstruction, it has clinical, radiologic, and neuropathologic characteristics, and its pathogenesis remains unclear. Currently, prevalent hydrodynamical theories assert that obstruction of the subarachnoid space at the foramen magnum interferes with flow of CSF between the spinal and intracranial subarachnoid compartments. As a result, spinal CSF is driven into the spinal cord through the perivascular spaces to form the syrinx.[8]

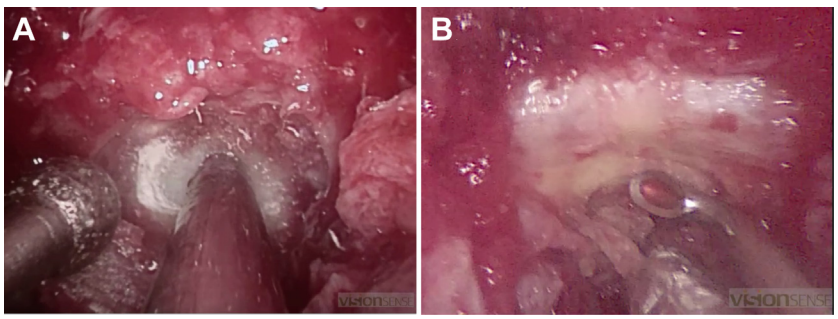

Figure 5. Intraoperative endoscopic views: (**A**) drilling of the odontoid peg after removing the C1 anterior arch upper portion, and (**B**) final endoscopic view after the odontoidectomy.

However, Levine[9] in 2004 proposed a new theory of pathogenesis of the syrinx because of obstruction of the foramen magnum. In the presence of subarachnoid obstruction at the foramen magnum, a variety of activities, such as assuming erect posture, coughing or straining, and pulsatile fluctuations of CSF pressure during the cardiac cycle, produce transiently higher CSF pressure above the block rather than below it. There are corresponding changes in transmural venous and capillary pressure favoring dilation of vessels below the block and collapse of vessels above the block. The spatially uneven change of vessel caliber produces mechanical stress on the spinal cord, particularly caudal to the block. The mechanical stress, coupled with venous and capillary dilation, partially disrupts the blood–spinal cord barrier, allowing ultrafiltration of crystalloids and accumulation of protein-poor fluid. The proposed theory is

consistent with the neuropathologic findings in syringomyelia and with the pressure and composition of syrinx fluid. It also accounts for the prolonged course of syringomyelia, and it is aggravated by cough, strain, and assumption of an erect posture.

The proposed theory also explains the relatively low incidence of syringobulbia. In the series of Klekamp et al.,[10] 73% of patients with Chiari I malformation had syringomyelia, whereas only 3% had syringobulbia.

The most common treatment of syringobulbia is craniovertebral decompression. It consists of a small craniectomy that decompresses the tonsils usually by removing the arachnoid and often by extracting away part of the tonsil tissue. Sometimes shunting the syrinx into peritoneal, pleural, or atrial cavities is needed.

In the patient described, it was thought that the syrinx resolution should have been provided with an anterior decompression of the CVJ.

In fact, the partial blockage of the CSF pathways at the level of the foramen magnum, which is described as the most important pathogenetic mechanism of syringobulbia development, in this case was because of CVJ stenosis at the level of C1-2.

Transoral decompression has been the criterion standard procedure for anterior decompression of C1 and C2; however, there may be some disadvantages to this approach.[3,11] These include a long surgical corridor with limited visibility and restricted manual and digital access beyond the oral cavity given dental, occlusal, and palatal anatomic constraints. After a transoral approach, there is exposure of the surgical bed to saliva and oral contents with each swallow raising concern for impaired healing or infection, especially in the event of CSF leakage. Postoperative oropharyngeal swelling increases concern for safe extubation and often dictates consideration of tracheostomy and feeding tube placement.

The presented case along with others have previously described the endonasal endoscopic approach to the craniocervical junction for several etiologies.[6,12-14] Advantages to the transnasal approach include outstanding endoscopic full-length visualization of the deep surgical corridor that is created, favorable nasal mucosal wound healing, early advancement to an oral diet, and the reduced likelihood of prolonged postoperative intubation because of concerns of surgical site swelling. Disadvantages are primarily anatomic, because the hard palate of the oral cavity limits endoscopic instrumentation below the C2 vertebral body, and technical, given the need for multidisciplinary surgical expertise with endonasal neurosurgery and for specialized equipment required to access this distant skull base location.[15] In addition to acute compression because of syringobulbia, an endonasal approach has also been previously used in decompression of patients with cervicomedullary tumor, rheumatoid arthritis, and degenerative disease.[14]

In our department, we have been performing endoscopic endonasal odontoidectomy from 2011. In our series, we have performed 16 procedures. In this series of patients that underwent endoscopic endonasal odontoidectomy for irreducible ventral brainstem compression in our division, the minimal follow-up was 6 months. Out of 16 patients, 12 have a follow-up longer than 24 months. Seven of them have been followed for over 5 years.

For all patients, a successful decompression was achieved at the first surgery. In 7 patients it was not possible to preserve the C1 anterior arch. In 6 cases there was neurologic symptoms improvement. Two patients referred preoperative dysphagia resolution in the few weeks after surgery; however, the neck pain did not completely resolve. Two patients of 12 were asymptomatic after surgery.

In particular, in this case, the procedure was performed in an emergency situation. There is also another description of endoscopic endonasal CVJ decompression performed in emergency. However, in that case, there was compression because of an infectious disease.[16] Indeed, this is the first report of a 3D endoscopic endonasal odontoidectomy performed to achieve CVJ decompression in syringobulbia.

Complete removal of the C1 arch and ligaments led to instability of the CVJ. For this reason, the patient went through a second surgery with posterior stabilization.[13,17-20] In other cases, a 2-stage operation was performed with C1-2 fixation. However, there are groups that strongly advocate that posterior stabilization should precede all considerations of anterior endonasal surgery.[1]

CONCLUSIONS

This case report details the successful use of endoscopic endonasal anterior decompression to treat syringobulbia, and adds to the growing literature in support of the endonasal endoscopic approach as a safe and feasible means for decompressing the craniocervical junction, now also in an emergency setting. Nevertheless, careful patient selection, combined with sound clinical judgment, access to instrumentation, and intraoperative imaging cannot be overemphasized.

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Conflict of interest statement: The authors declare that the article content was composed in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

Received 24 March 2019; accepted 1 July 2019

Citation: World Neurosurg. (2019) 130:499-505.
https://doi.org/10.1016/j.wneu.2019.07.004

Journal homepage: ***www.journals.elsevier.com/world-neurosurgery***

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