Endonasal endoscopic resection of the odontoid process in a nonachondroplastic dwarf with juvenile rheumatoid arthritis: feasibility of the approach and utility of the intraoperative Iso-C three-dimensional navigation

Case report

Illya Laufer, M.D.,1 Jeffrey P. Greenfield, M.D., Ph.D.,1 Vijay K. Anand, M.D.,2 Roger Hartl, M.D.,1 and Theodore H. Schwartz, M.D.1

Departments of 1Neurological Surgery and 2Otolaryngology, Weill Cornell Medical College, New York Presbyterian Hospital, New York, New York

The authors report a case of a nonachondroplastic dwarf with severe basilar invagination and compression of the cervicomedullary junction (CMJ) due to juvenile rheumatoid arthritis. Initially excellent reduction of the invagination and decompression of the CMJ was achieved using posterior fixation. However, 1 month postoperatively symptoms recurred and the authors found imaging evidence of recurrence as well. The patient subsequently underwent an endoscopic transnasal resection of the dens with assistance of Iso-C navigation. He recovered well and tolerated regular diet on postoperative Day 2. (DOI: 10.3171/SPI/2008/8/4/376)

Key Words • basilar invagination • endoscopy • odontoidectomy • rheumatoid arthritis

**Abbreviations used in this paper:** CMJ = cervicomedullary junction; RA = rheumatoid arthritis.

**Case Report**

**Examination.** This 25-year-old man with a history of non-achondroplastic dwarfism and juvenile RA presented with progressive occipital headache, neck pain, and intermittent paresthesias in bilateral upper extremities. Chronic steroid therapy for his arthritis caused secondary osteoporosis. In the past he had undergone bilateral hip arthroplasties and was being evaluated for knee arthroplasty. On examination he had full motor strength and no evidence of myelopathy. Cervical magnetic resonance imaging revealed pronounced basilar invagination with compression of the CMJ (Fig. 1A).

**First Operation.** Initially, the patient was placed in traction and experienced significant reduction of the invagination...
tion (Fig. 1B and C). He subsequently underwent occipitocervical fixation and arthrodesis. Intraoperatively, his bone was noted to be severely osteoporotic and was judged to be too brittle for stable screw purchase. Thus, sublaminar wires at C-2 through C-6 were attached to bilateral titanium rods, connecting the construct to an occipital plate held in place with bicortical screws. The fusion was enhanced with allograft bone and bone morphogenetic protein. The patient was placed in halo fixation. Postoperative imaging revealed significant reduction of the invagination (Fig. 2A).

Second Operation. At the 1-month follow-up the patient complained of recurrent neck pain, and imaging revealed increased angulation of the cranial base in relationship to the neck with recurrent basilar invagination, but stable posterior instrumentation (Fig. 2B). The halo was readjusted and he was taken for an endoscopic transnasal odontoidectomy. The Iso-C system (Siremobil Iso-C Three-Dimensional, Siemens Medical Solutions) with 3D fluoroscopic reconstruction was used for frameless stereotactic navigation (BrainLAB) and for postresection intraoperative evaluation of the anterior decompression.

Postoperative Course. The patient was extubated the morning after surgery and was eating a regular diet within 2 days. Postoperative computed tomography scanning confirmed complete resection of the odontoid process and decompression of the CMJ (Fig. 2C).

Surgical Procedure

The details of the anatomical dissection and surgery have been reported elsewhere.\textsuperscript{1,6,11,16} We have incorporated small modifications to this previously described technique, which will be highlighted. In brief, following oropharyngeal intubation with the halo in place, antibiotics and dexamethasone were administered. A submucosal resection of the sepal cartilage was performed to facilitate the lateral-to-medial endonasal movement of the instruments. The inferior turbinates were outfractured bilaterally and the posterior 2 cm of the nasal septum was removed using a tissue shaver and a high-speed drill to enlarge the choana for a wider exposure. The sphenopalatine artery and the posterior septal artery were cauterized bilaterally. We did not remove the middle turbinates, and the sphenoid sinus was only opened slightly as a landmark. A 30-cm, 4-mm rigid 0° endoscope (Karl Storz) was held in the left nostril with an endoscope holder and surgery was performed using a bimanual technique either via the right nostril or both nostrils as required.
A red rubber catheter was placed through the nasal cavity into the oral cavity for downward retraction of the soft palate to facilitate exposure (Fig. 3). An inverted U-shaped incision was made in the basopharyngeal fascia with its superior extent just below the sphenoid sinus, passing down the clivus with its lateral limits just medial to the eustachian tube and its base at the level of the C-2 vertebral body (Fig. 4A). The flap was pushed inferiorly into the oropharynx and the longus colli and capitis muscles were pushed laterally with the bovie to expose the ring of C-1. The arch of C-1 was removed with a high-speed drill (Fig. 4B) to expose the lateral margins of the dens, which was transected across its base. Because the bone was demineralized from chronic steroid use, the odontoid was resected in a piecemeal fashion using an angled curette and a pituitary rongeur. The base of C-2 was then removed with the aid of a 30° 18-cm, 4-mm endoscope (Karl Storz) angled downward to facilitate inferior visualization. The alar and apical ligaments were transected. The soft-tissue pannus, transverse ligament, and tectorial membrane were then removed with a cavitron until dural pulsations were seen. The Iso-C was brought into the field to create a triplanar tomograph of the CMJ. The residual odontoid was identified laterally and it still compressed the thecal sac; the images were used for navigation to remove the remaining odontoid bone. A final image with the Iso-C demonstrated a complete resection. The basopharyngeal fascial flap was reapproximated using a fibrin sealant (Tisseel VH, Baxter).

Discussion

Basilar invagination is found in up to 11% of patients with RA and often manifests as occipital or neck pain, frequently progressing to disabling myelopathy. Once conservative management fails, occipitocervical fixation with or without anterior decompression is a viable treatment option. Posterior fixation can often be complicated in the RA population by poor bone quality, long-standing steroid use, and extensive ligamentous laxity.5,9,13

The transoral approach provides the most direct route to the ventral craniovertebral junction and is currently the standard of care for anterior decompression.5,14 Transmaxillary and transmandibular extensions of this approach may be used to provide wider rostral or caudal working fields.5 However, these approaches often require a postoperative tracheostomy or gastrostomy and cause significant tongue and tracheal swelling. Hypernasal speech and nasal regurgitation can occur. Mandibular splitting is cosmetically unappealing, and prolonged recovery and hospitalization are often needed. The wound is constantly bathed in saliva and oropharyngeal flora, which increases the risk of infection. Use of the operative microscope limits the field of view, and in patients with micrognathia and/or dwarfism the transoral microscope-assisted resection of the odontoid is contraindicated.

Endoscopic illumination brings the lens and light source closer to the operative field, thereby reducing the need for a wide operative approach.2 Endoscopic anterior cervical,19 transoral,7,8 and transnasal11 approaches have been previously described. The advantage of the transnasal approach is that the incision lies above the oropharynx. Thus, the transnasal approach should decrease the risk of tongue and tracheal swelling, hypernasal and nasal speech and wound infection. Our patient was extubated within 12 hours of completion of the operation and his diet was returned to normal within 2 days, certainly quicker than would have...
been possible after a transoral procedure. This led to an expeditious recovery and hospital discharge. In addition, patients with dwarfism or micrognathia and children whose natural orifices are too small for the microscope-based transoral approach can be treated using a transnasal endoscopic approach. The main limitation of the approach is the inferior extent, which can only reach the bottom of C-2 whereas the transoral approach can reach C-3.

We wished to acknowledge the excellent work in cadavers and the prior case in the literature in which the authors initially described the endoscopic transnasal approach to the CMJ.\textsuperscript{1,6,11,16} The purpose of our report was to validate the approach and demonstrate its reproducibility and success at another institution. In addition we wished to describe several operative nuances that differentiate our approach from prior reports. 1) Although the surgery benefits from collaboration between an otolaryngologist and a neurosurgeon, during the resection of the odontoid process, the operative field is relatively stable, allowing us to place the endoscope on a scope holder and obviating the need for 2 surgeons at all times. 2) We did not remove the middle turbinate and found the space adequate for surgery. We believe that preservation of both middle turbinates is helpful in restoring normal laminar flow of air through the nose and that it minimizes postoperative crusting. 3) We did not widely open the sphenoid sinus and drill down the clivus, as previously described.\textsuperscript{11} The top of the inverted U-shaped incision can sit just below the sphenoid sinus, which makes the operation quicker and even less invasive. 4) We performed a submucosal resection of the septal cartilage. This maneuver permits the scope and instruments to be moved medially without resistance. 5) The surgery in our case was performed in a dwarf with micrognathia, indicating that the size of the nasal passages is not a limitation, further validating the minimal-access philosophy of this approach. 6) Because the operative access for the transnasal endoscope approach requires exposure only of the nostrils, we were able to perform the approach while the patient was still fixed in the halo vest. This would not be possible using the transcervical endoscope approach.\textsuperscript{19} 7) We used the Iso-C system both for navigation and for intraoperative quality assurance that the odontoid had been adequately decompressed. The addition of triplanar fluoroscopy and real-time imaging of surgical results is extremely helpful to guarantee reliable results.

Conclusions

We report a case where the transnasal endoscopic approach to the CMJ provided a safe and effective minimal access alternative to the transoral approach. Patients with small oral and nasal cavities are still candidates for surgery and the morbidity and hospital length of stay should theoretically be decreased compared with those features in patients undergoing the traditional transoral surgery. The advent of this approach is particularly significant for patients whose anatomy precludes the standard transoral approach and requires the more morbidity-prone alternative approaches. Intraoperative Iso-C–based 3D navigation significantly contributed to a complete, accurate, and safe resection of the osseous mass. Larger case series will be required to further validate this approach.

References


Manuscript submitted August 23, 2007. Accepted December 4, 2007. Address correspondence to: Theodore H. Schwartz, M.D., Department of Neurological Surgery, Weill Cornell Medical College, New York Presbyterian Hospital, 525 East 68th Street, Box #99, New York, New York 10021. email: schwarh@med.cornell.edu.