Endoscopic resection of odontoid process in Arnold Chiari malformation Type II

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Abstract. Introduction: Arnold Chiari Malformation Type II can be associated with basilar invagination through an elongated retroflexed odontoid process (dens axis). Traditionally, decompression surgery has been performed transorally under microscopic vision or via transcutaneous latero-cervical/posterior approaches. Endoscopic approaches were introduced a few years ago.

Case report: We report of an eleven-year-old girl with Arnold Chiari Malformation Type II who had undergone surgery eight years ago for posterior cranial fossa decompression at the department of neurosurgery. At that time, an external transcutaneous median approach was performed to resect the posterior arch of the atlas. The patient now presented with the initial symptoms of brainstem compression as a result of an elongated retroflexed odontoid process and cranio-cervical instability.

Surgical technique: An endoscopic transoral/transnasal approach was chosen for the resection of the dens.

Conclusion: Endoscopic surgery was successful and the complete resection of the dens was achieved without any complications. In a second intervention, orthopaedic surgeons performed cranio-cervical arthrodesis.

Introduction

Arnold Chiari originally described the malformation Type II in 1891 as the caudal displacement of the vermis, brainstem and fourth ventricle.\textsuperscript{1-3} Chiari malformation Type II is also associated with myelomeningocele of varying extent and hydrocephalus.\textsuperscript{1} An elongated retroflexed odontoid process of the axis (C II) can also be found. Patients younger than two years have a small cerebellum and small posterior cranial fossa with a large foramen magnum through which cerebellar structures protrude.\textsuperscript{3} Patients present with elevated intracranial pressure, cranial nerve dysfunction and consecutive vocal and respiratory defects like inspiratory stridor. Gastrointestinal dysfunction leads to malnutrition and aspiration. Other symptoms include para- or tetraparesis, hypotonia, nystagmus and opisthotonus, and mental and physical retardation.\textsuperscript{3} In patients older than two years, cervical myelopathy with plegia of the upper extremities, ataxia and headache and/or cranio-cervical pain prevail.\textsuperscript{3} Surgical interventions for hydrocephalus and the decompression of the cervical spine are part of the treatment algorithm.

The “gold standard” for cervical spine decompression and the resection of the odontoid process used to be transoral microscopic or external transcutaneous approaches.\textsuperscript{4} Recently, endoscopic endonasal surgery has been used for the resection of the odontoid process as a less invasive technique resulting in fewer intra- and post-operative complications and comorbidities.\textsuperscript{4-7}

Case report

An 11-year-old girl with Arnold Chiari Malformation Type II presented with initial symptoms of brainstem compression. Eleven years previously, she had undergone multiple ventriculo-peritoneal shunt surgeries for hydrocephalus and the closure of a lumbar meningomyelocele. Furthermore, the posterior arch of the atlas (C I) had been resected for posterior cranial fossa decompression in 2001 at the neurosurgical department. Two years later, the patient had developed syringomyelia as a result of the compression of the brain stem and consec-
uitive cerebrospinal fluid malcirculation.

Further diagnoses included left-sided thoraco-scoliosis and rightsided lumbar scoliosis and a neurogenic reflex bladder.

Recent clinical examination and imaging revealed an elongated, retroflexed odontoid process with basilar invagination and cranio-cervical instability (Figures 1a and 1b).

A two-step interdisciplinary approach was decided upon, with primary endoscopic transnasal/transoral odontoid process resection by ENT surgeons and neurosurgeons, and secondary cranio-cervical arthrodesis by orthopaedic surgeons.

Surgical technique

The procedure was performed using computer-assisted navigation with the CT/MR fusion technique (InstaTrak® 3500 Plus by GE Healthcare, Vienna, Austria). Firstly, a velottractor was used for the gentle elevation of the soft palate. Four-millimetre Karl Storz 0° and 45° endoscopes were used transnasally and transorally for visualisation. The transoral insertion of the angled endoscope provided more space for instruments in both nostrils since surgery was performed using a four-handed technique. C I and C II were identified (Figure 2). A T-shaped mucosal incision was then made, with the horizontal bar along the body of C I and the vertical portion down to the lower margin of C II. Preparation was continued until the periosteum was reached and the prevertebral muscles were detached laterally up to four to five millimetres on each side. The periosteum was dissected sharply and, after the identification of the anterior arch of C I, a diamond burr was used to resect the latter. After exposure, the odontoid process was cavitated under navigational control until the peripheral bone compacta was reached. This remaining bony layer was then resected carefully. Bony margins were smoothened with the burr. After identifying the dura of the posterior cranial fossa, the defect was closed with TachoSil® (Nycomed International Management GmbH, Zurich, Switzerland) and fibrin glue. Layers of prevertebral muscles and mucosa were sutured transorally. No complications occurred.

In a second intervention, orthopaedic surgeons successfully performed posterior cranio-cervical arthrodesis from the skull base to C V (Figure 3). Post-operative wound healing was uneventful and no naso-gastric feeding tube was needed because swallowing was normal. The patient was discharged eleven days after surgery.

Follow-up

No special nasal treatment (nasal douches, debridement) was required. One year after surgery the patient’s cervical spine was stable without any further signs of brain stem compression. The syringo-
myelia that was apparent preoperatively remained stationary and was not affected by the surgical approach.

**Discussion**

Traditionally, the resection of the odontoid process for brainstem decompression in patients with basilar invagination is performed transorally or via craniotomy. Recent anatomical studies described endoscopic endonsasal approaches and their feasibility. Indications for surgical intervention are rheumatoid arthritis, tumours, cysts, trauma, cranio-cervical malformations (Klippel-Feil Syndrome) and skeletal diseases (osteogenesis imperfecta, Paget Disease, Hurler Syndrome, osteomyelitis etc.). At present, the literature does not include any report of endoscopic odontoid process resection in Arnold Chiari Malformation Type II.

The advantages of the endoscopic approach are the enhanced visualisation of the surgical field.
and the less invasive nature of the procedure. Endoscopic surgery avoids opening a wide surgical corridor with palatal splitting, the resection of the posterior septum or even the middle turbinates, and the prolonged compression of the tongue as in microscopic approaches. In transmandibular approaches, the cosmetic outcome was unsatisfactory, with post-operative requirements like nasogastric feeding tubes, prolonged intubation or even tracheostomy, pain and prolonged hospitalisation. Complications such as velopharyngeal insufficiency, with nasal reflux and hypernasal speech, necrosis of the tongue, upper airway obstruction resulting from oedema, and wound dehiscence have been reported for external approaches. Furthermore, the incision in endoscopic approaches is above Passavant’s bar, preventing nasal regurgitation and regurgitation from the oropharynx.

In our case, the surgery was performed in a four-hand technique in partnership with a neurosurgeon. Access to the surgical field was therefore established by alternating the endoscope’s position – through the nostrils or through the mouth – depending on the best angle of vision. The rhino-neurosurgical team achieved the meticulous dissection of the bony odontoid shell with three hands manipulating instruments simultaneously and one hand guiding the endoscope, alternating routes and approaches on demand. Filling the surgical cavity with haemostyptic resorbable materials, and suturing the prevertebral musculature and nasopharyngeal mucosa, resulted in the adequate closure of the defect. As the incisions and surgical access remained above Passavant’s bar, not even a (naso-) gastric probe was required post-operatively. This report describes another extension of the indications for endoscopic transnasal surgery beyond the paranasal sinuses.

Conclusion

An endoscopic combined transnasal/transoral approach for the resection of the odontoid process in basilar invagination appears to be safe and is less invasive than traditional external approaches. Visualisation of the surgical field is excellent, requiring a smaller surgical corridor than traditional approaches. Navigation with CT/MRI fusion is of great help in preserving vital structures like vertebral arteries, especially when surgical manipulation is off midline. Given these advantages, endoscopic transnasal/transoral resection of the odontoid process should be considered in individual cases.

References

14. Nayak JV, Gardner PA, Vescan AD, Carrau RL, Kassam AB, Snyderman CH. Experience with the expanded endonasal approach for resection
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