Basilar Invagination: Case Report and Literature Review

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Key Words
- Anterior odontoid decompression
- Basilar invagination
- Cervicomedullary junction compression
- Endoscopic endonasal odontoidectomy
- Posterior odontoid decompression

Abbreviations and Acronyms
BI: Basilar invagination
CVJ: Craniovertebral junction
RA: Rheumatoid arthritis

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BACKGROUND: **Basilar invagination is a rare clinical condition characterized by upward protrusion of the odontoid process into the intracranial space, leading to bulbomedullary compression. It is often encountered in adults with rheumatoid arthritis. Transoral microscopic or endonasal endoscopic decompression may be pursued, with or without posterior fixation. We present a case of basilar invagination with C1–C2 autofusion and discuss an algorithm for choice of anterior versus posterior approaches.**

CASE DESCRIPTION: **A 47-year-old woman with rheumatoid arthritis presented with severe occipital and cervical pain, dysphagia, hoarseness, and arm paresthesias. Findings on magnetic resonance imaging revealed moderate cranial settling with the odontoid indenting the ventral medulla but no posterior compression. Computed tomography demonstrated bony fusion at C1–C2 without lateral sag. Given autofusion of C1–C2 in proper occipitocervical alignment and the absence of posterior compression, the patient underwent endoscopic endonasal odontoidectomy without further posterior fusion, with satisfactory resolution of symptoms.**

CONCLUSION: **Endoscopic endonasal odontoidectomy offers a safe and effective method for anterior decompression of basilar invagination. Preoperative assessment for existing posterior fusion, absence of posterior compression, and preservation of the anterior C1 ring during operative decompression help stratify the need for lone anterior approach versus a combined anterior and posterior treatment.**

INTRODUCTION

Symptomatic anomalies of the craniovertebral junction (CVJ) present a unique challenge for neurosurgical management with regard to the selection of surgical approach and planning and the accompanying technical difficulty. Of the CVJ anomalies, basilar invagination (BI) confers a particular therapeutic challenge. BI often coexists with or is a result of other medical conditions (e.g., Chiari malformation, Down syndrome, Klippel Feil, osteogenesis imperfecta, and rheumatoid arthritis [RA]), which complicate the achievement of atlanto-axial stability and increase perioperative complications (23). In patients with BI, the ventral aspect of the brainstem or the craniocervical junction often is compressed, leading to neurologic symptoms necessitating decompression. Because of the mechanical and anatomical complexities, surgical treatment continues to center on varying combinations of anterior or posterior decompression with or without traction or fusion. Common routes for decompression include endonasal and transoral access ventrally or an occipital-cervical posterior decompression and fixation (15). We present a case of RA with odontoid basilar compression and autofusion of C1–C2, in whom an endoscopic endonasal decompression was pursued, without posterior fusion. We further discuss a systematic algorithm for selection of operative approaches to treatment of BI based on our experience and literature published.

CASE REPORT

A 47-year-old woman with long-standing RA presented with 6 months of increasing neck and occipital pain, occasional dysphagia, dysphonia, and difficulty breathing. Magnetic resonance imaging (MRI) and computed tomography (CT) revealed BI and moderate cranial settling (Figure 1). Review of prior cervical spine films revealed mild subluxation of C1 and C2 since 1998. Operative intervention was indicated given her symptomatic and radiographic progression of brainstem compression. Observation of autofusion at the C1–C2 joint in proper occipitocervical alignment prompted an endoscopic endonasal approach for odontoidectomy and decompression, deferring an accompanying posterior fusion. The odontoid was exposed following a small inferior posterior septectomy and lateralization of the buccopharyngeal fascia from the level of the Eustachian tubes to C2. The inferior aspect of the clivus was drilled to expose foramen magnum and the odontoid peg was resected in the setting of pannus and hypertrophied ligamentous tissue. Adequate decompression was achieved with preservation of the arch of C1. The patient tolerated the procedure well and reported near-resolution of

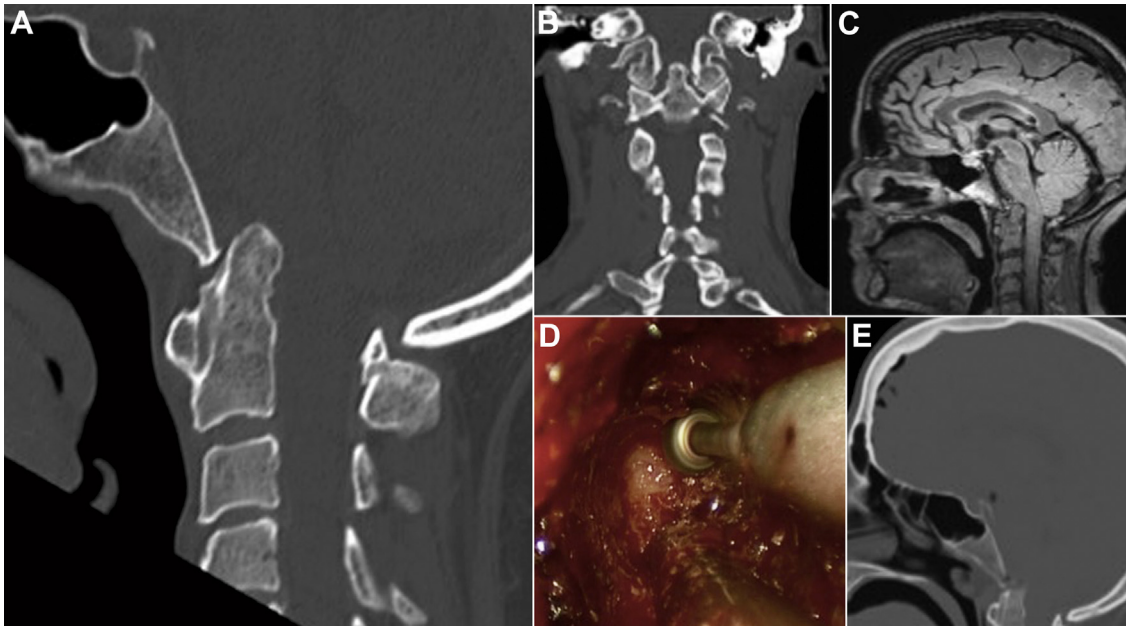

Figure 1. Preoperative and postoperative basilar invagination imaging. Preoperative cervical CT of the spine demonstrating (**A**) basilar compression of the medulla in mid-sagittal view and (**B**) cranial C1–C2 settling on coronal view. (**C**) Preoperative sagittal view of the cervicomedullary compression on T2-weighted fluid-attenuated inversion recovery MRI. (**D**) Intraoperative drilling of odontoid peg. (**E**) Postoperative CT illustrating successful anterior decompression after endoscopic endonasal odontoidectomy.

occipital and cervical pain, with only intermittent mild dysphagia, on follow-up.

DISCUSSION

Currently, no defined guidelines exist for the management of patients with BI. Each patient is different and complex because of frequently accompanying concomitant diseases. Decisions are based on imaging findings, often complicated by abnormalities such as cranial settling, autofusion of cervical vertebrae, platybasia, and syringomyelia. Management should account for both compression of the brain stem and cervical cord as well as for craniospinal instability. There is no clear consensus as to treatment approaches for this condition, with options ranging from cervical traction, lone anterior decompression, posterior sagittal realignment and fusion with or without bony decompression, as well as combined anterior/posterior approaches (9, 13-15, 22, 26).

Preoperative Considerations

Preoperative imaging of the connective tissue, musculature, and osseous alignment should be closely evaluated. In cases in which there is evidence of ventral tissue contraction, posterior reduction may be limited, thus necessitating an anterior approach. Identification of the relationship of the vertebral arteries to the C1-C2 lateral masses, posterior arch, and entry at the occiput is essential. The course of the vertebral arteries is also known to course irregularly along the paraesophageal tract and ventral body of C2, affecting the operative approach (20, 21, 29).

Considerations for Posterior Approaches

In cases of BI without cranial nerve palsies, suggesting low-level ventral compression, posterior realignment and fusion has shown to be successful when a mobile and reducible C1–C2 complex is present (1). This realignment can be achieved with preoperative traction, a Halo vest, distraction of C1–C2 by intra-articular spacers, or by intraoperative traction, followed by subsequent posterior instrumentation and fusion (1, 9, 28).

Considerations for Anterior Approaches

In cases of moderate-to-severe ventral compression, initial criteria to consider are presence of cranial nerve palsies, in which case, an anterior approach often is necessary, independent of posterior complex stability (17, 26). When C1–C2 is fused, unsafe to distract, or if distraction fails to alleviate the ventral compression, anterior approaches have been shown to be successful and necessary (8, 11, 26, 32). Autofusion of nearby bodies such as C2–C3 is insufficient if the C1 integrity is not preserved (12). In select cases, anterior decompression can be safely pursued without causing significant instability, thereby avoiding a posterior fusion (3, 8, 11, 27, 32) (**Table 1**). Deteriorating neurologic function any time after a basilar decompression without posterior fusion would need to be further investigated, because a posterior fusion eventually may be required.

If there is evidence of auto-fusion of the C1–C2 lateral masses, either transoral or endonasal odontoidectomy is effective. The approach is best decided in accordance to the patient's anatomy and surgeon's intuition regarding the exposure needed to successfully treat the patient. The endonasal approach is favored when the lesion is located 2 cm above the palate (6). The nasopalatine line can assist with

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