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Two-Stage Surgical Management of a Giant Pediatric Chordoma of theCraniocervical Junction (C0-C3): A Case Report

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Case Report

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Abstract

Background

The surgical management of skull base and craniocervical junction (CCJ) chordomas remains a formidable challenge in modern neurosurgery. Various anterior and posterior approaches are currently employed for the resection of these tumors.

Case Presentation:

We present a rare clinical case of a three-year-old patient with a three-month history of headaches, progressive weakness in all extremities, and gait disturbance caused by giant chordoma extending from the clivus to the C1–C3 vertebrae. The surgical strategy involved a two-stage procedure: initial posterior craniocervical stabilization followed by tumor resection via a transoral approach.

Results

Subtotal resection of the tumor was achieved. Postoperatively, the patient's presenting tetraparesis demonstrated near-complete regression. The patient was discharged on the 14th postoperative day.

Conclusion

This case illustrates that advancements in neurosurgical techniques and accumulated clinical experience now enable the surgical treatment of extremely complex pathologies of the craniocervical junction, even in very young pediatric patients, and can ensure a high quality of life. This report details the surgical strategy and discusses the rationale for a staged approach in the management of giant pediatric chordomas.

Introduction

At the current level of neurosurgical advancement, the treatment of patients with chordomas of the craniocervical junction remains a complex challenge, feasible only for multidisciplinary teams in high-volume tertiary centers with extensive experience. The infiltrative and destructive growth pattern of chordomas, with their propensity to involve the craniocervical junction and upper cervical spine, creates significant difficulties in selecting an optimal surgical approach and achieving gross-total resection, which is critical for ensuring long-term progression-free survival. For ventrally located tumors, approaches through natural anatomical corridors (transnasal or transoral) are often the most viable options [1–4]. However, the C1-C2 vertebral complex located along this path can obstruct adequate exposure of the tumor, and its resection inevitably necessitates subsequent stabilization of the craniocervical junction via occipitospondylodesis or C0/1-C2/3 fixation (anterior or posterior) [5, 6].

While such procedures are well-established in the management of adult patients, the surgical treatment of children with craniocervical junction chordomas presents unique and distinct challenges.

Objective

To present a clinical case of a three-year-old patient with a giant chordoma of the craniocervical junction and C1–C3 vertebrae, managed with a two-stage surgical strategy.

Case Presentation

Patient Information

A 3-year, 11-month-old male presented with a three-month history of headaches, progressive weakness in all extremities, and gait disturbance. Past medical history was unremarkable, with no family history of similar conditions.

Clinical Findings

Neurological examination revealed left-sided tongue deviation, tetraparesis (muscle strength 1–2/5 on the MRC scale), and neurogenic bladder dysfunction.

Diagnostic Assessment

Magnetic resonance imaging (MRI) identified a giant tumor involving the clivus, craniocervical junction, and the C1-C3 vertebrae, causing significant compression of the brainstem (Fig. 1). Based on imaging findings, the presumptive diagnosis was chordoma.

Therapeutic Intervention

Therapeutic Intervention
Given the tumor's ventral location relative to the brainstem, an anteroposterior surgical trajectory was deemed optimal. As the tumor had also destroyed the ventral C1-C2 complex, its resection was necessary to access the intraspinal tumor components, which would inevitably result in craniocervical instability. A two-stage procedure was planned.

Stage 1: Posterior Stabilization. Occipitospondylodesis was performed using a patient-specific titanium implant (Fig. 2). Due to the patient's young age and the consequent thinness of the occipital bone, bicortical screws were utilized. Computational modeling of the custom stabilization construct is shown in Fig. 2.

Stage 2: Tumor Resection. Two weeks after the first stage, the patient underwent a transoral subtotal resection of the tumor. A tracheostomy was performed prior to the procedure. The diagnosis of chordoma was confirmed by histopathological examination.

Follow-up and Outcomes

The postoperative course was uneventful. Enteral feeding was initiated on postoperative day 5, and the tracheostomy was removed on day 7. Neurological examination showed significant improvement, with tetraparesis regressing to a muscle strength of 4/5. The patient was discharged in satisfactory condition on the 14th postoperative day. At the follow-up examination 5 months after surgery, further positive dynamics were noted, with limb strength improving to 5/5. There are no signs of continued tumor growth on the control MRI scan.

Discussion

Pediatric patients with chordomas of the craniocervical junction and upper cervical spine represent a particularly severe cohort. Due to the plasticity of neural structures, pronounced clinical symptoms develop relatively quickly, but typically only when the tumor has already reached large or giant sizes. This significantly complicates radical resection and diminishes the chances of patient recovery. In such situations, the primary goals become ensuring brainstem decompression and achieving the maximal safe tumor resection. These objectives are hindered by the tumor's propensity for infiltrative growth among major vessels and cranial nerves in close proximity to vital brainstem structures.

An anatomically justified approach in cases like the one we present is the transoral route. Despite its long history in neurosurgery, it is routinely employed only in high-volume tertiary centers due to the technical complexity of such procedures and the necessity for specific expertise not only from the surgeon but also from intensivists and anesthesiologists. Consequently, the management strategy for these patients varies significantly between institutions.

We accumulated extensive experience, having treated 896 patients with chordomas of various locations between 1985 and 2025. Of these, 522 patients had skull base tumors operated via transnasal, transoral, or combined approaches. Extracranial (transoral, n = 43; combined transoral and transnasal, n = 8) tumor resection with preliminary craniocervical stabilization was performed in 51 cases; however, the current three-year-old patient is the youngest to undergo this procedure. The literature describes no more than 250 similar operations, with the largest published series authored by David Choi and Alan Crockard [1, 2].

Currently, our standard protocol for such patients involves a two-stage management strategy: initial posterior decompression and stabilization, followed by transoral tumor resection after an interval of 2–3 weeks. This strategy has replaced single-session procedures due to their excessive duration (up to 11–14 hours) and the need for intraoperative patient repositioning [7, 8].

Given the tumor's anatomical location, gross-total or near-total resection using various approaches is achieved in no more than 80% of cases [9–13]. In our presented case, the tumor was also removed subtotally. Nevertheless, the primary surgical objective was met: effective decompression of the brainstem and upper cervical spinal cord. This resulted in a markedly positive impact on the patient's neurological status, with near-complete regression of tetraparesis and no postoperative complications, the reported frequency of which reaches 30% in the literature [9].

In treating patients of such a young age, the choice of surgical approach is paramount. Based on our institutional experience and the tumor's topographical anatomy, we selected the transoral approach. Its application in children is associated with increased technical difficulties due to the smaller surgical field and consequently reduced space for instrumentation. However, this anterior midline approach provides optimal exposure of ventrally located tumors, allowing for effective brainstem decompression. Another critical consideration is the extent of bone resection (and thus, growth plate involvement) and the selection of a craniocervical stabilization method that accounts for the patient's existing anatomy and future growth potential. Numerous stabilization techniques for the craniocervical junction have been developed and are applied for various pathologies in this region [6]. Many of these systems can be personalized. In our clinical case, a patient-specific implant for occipitospondylodesis was manufactured, tailored to the individual anatomy and the thin occipital bone, which necessitated the use of bicortical screw fixation.

Conclusion

Advancements in neurosurgical techniques, technology, and the accumulation of clinical experience now enable the surgical treatment of extremely complex pathologies of the craniocervical junction, even in very young pediatric patients, and can ensure a high quality of life. The primary remaining challenges concern the long-term consequences on spinal growth and child development, which necessitate lifelong dynamic follow-up and the continued accumulation of clinical experience.

Abbreviations

CCJ
Craniocervical junction
MRI
Magnetic resonance imaging

Declarations

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Competing Interests

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Declarations of interest: none

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The approval of the Burdenko neurosurgery center local ethics committee has been received.

Consent to participate

Informed consent was obtained from all individual participants included in the study.

Written Consent for publication was obtained from all individual participants.

Research was conducted in accordance with the Declaration of Helsinki

The authors affirm that human research participants provided informed consent for publication of the images № 1 and №3

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Author Contributions

A.S. - final edit

A.K. - final edit

I.C. - wrote the main manuscript text

A.N. - final edit

V.P. - prepared literature review

I.L. - prepared literature review

M.K. - prepared parient data

O.R. prepared figures 1-3

All authors reviewed the manuscript.

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Figures

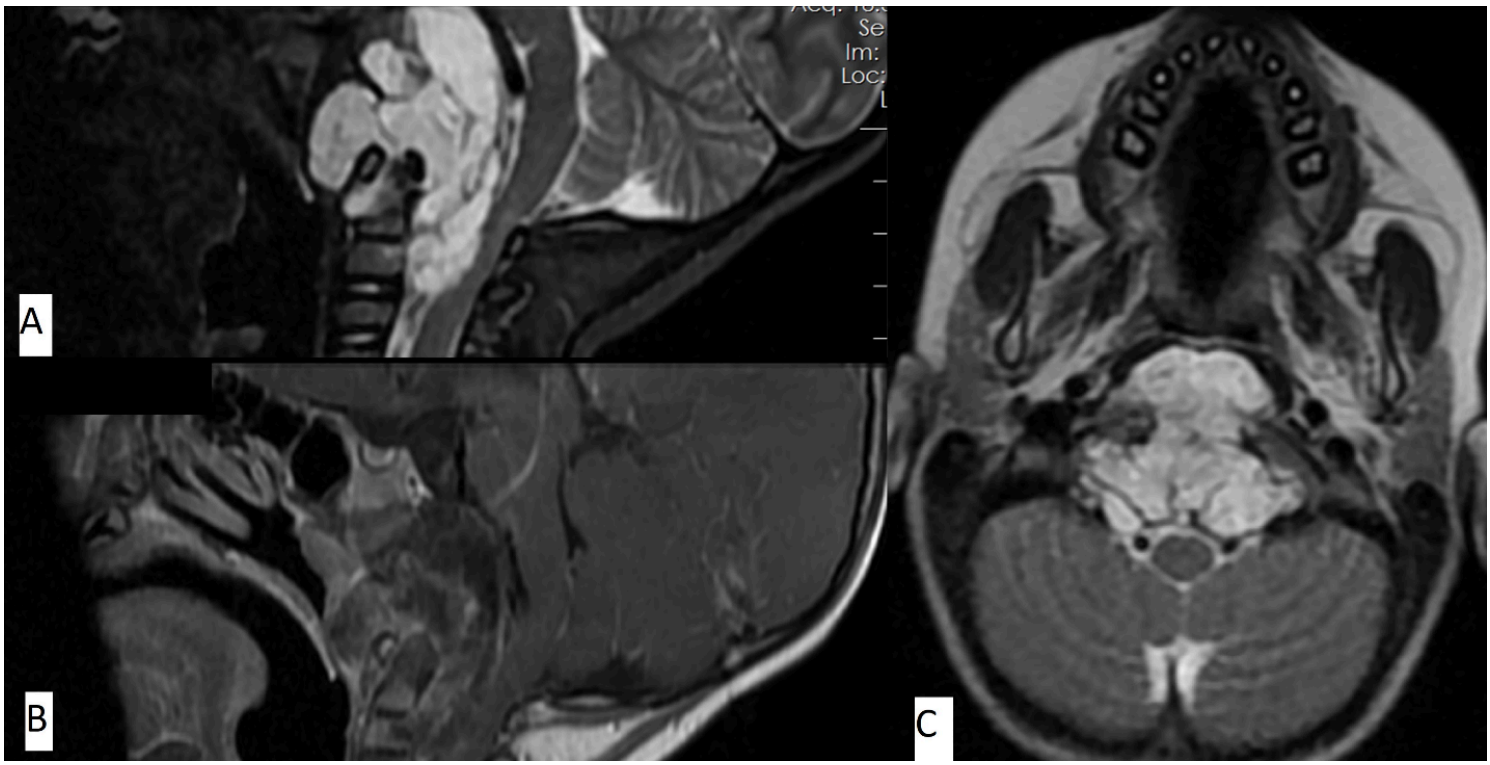


Figure 1

A – T2-weighted MRI, sagittal view. B – T2-weighted MRI, axial view. C – T1-weighted contrast-enhanced MRI, sagittal view. *The imaging reveals a giant chordoma causing severe compression of the brainstem structures.*

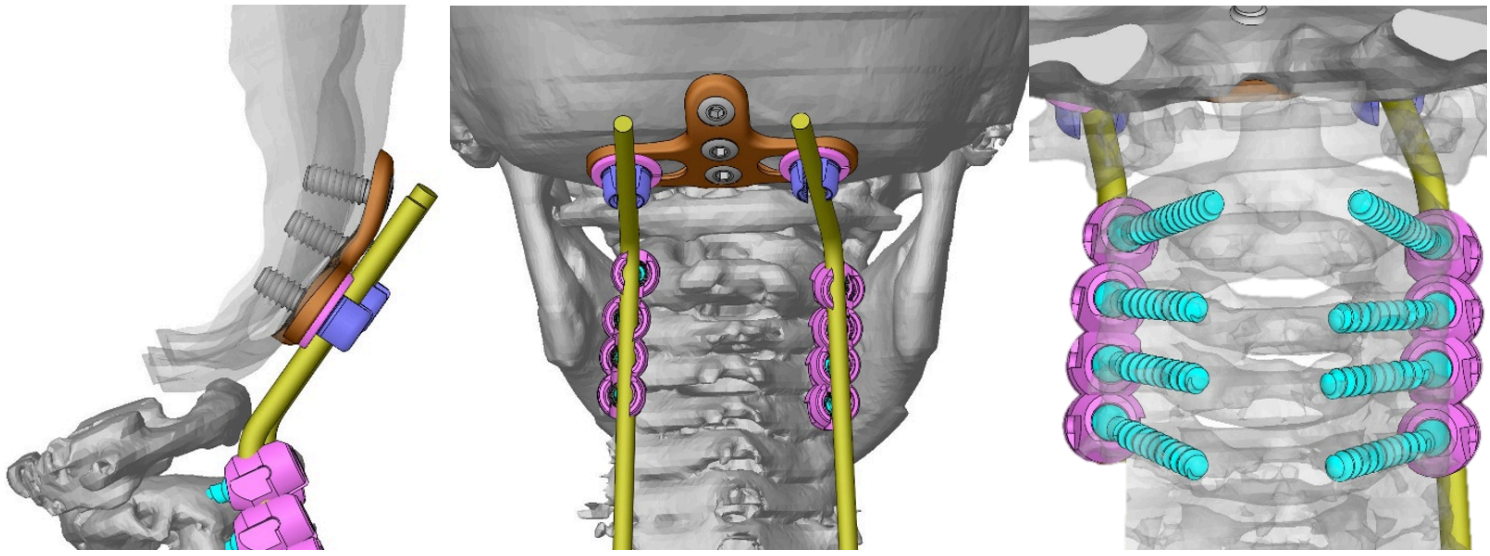


Figure 2

Patient-specific stabilization system in lateral, posterior and anterior view.

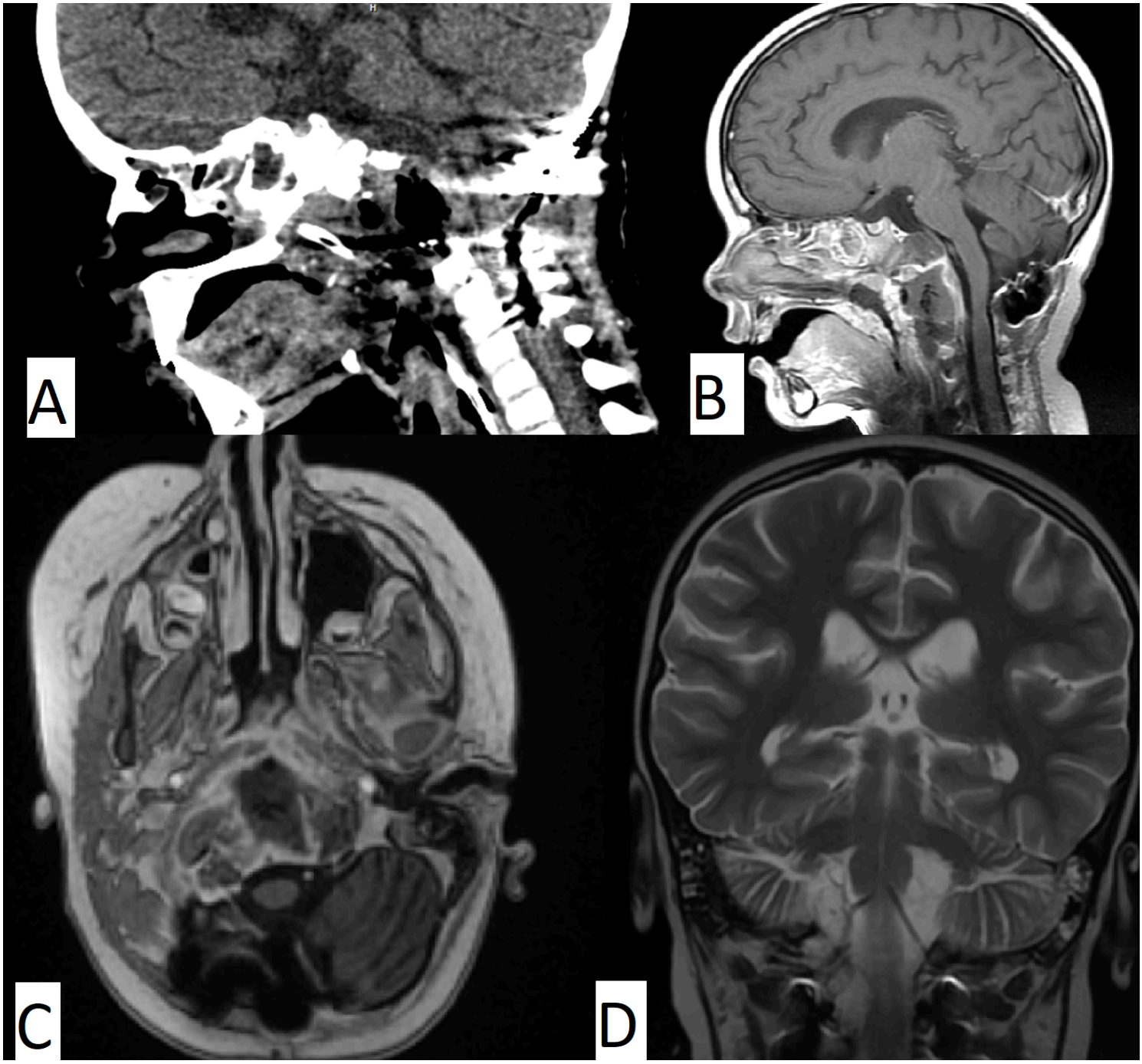


Figure 3

Postoperative CT and MRI on day 6. A – CT, sagittal view. B - T1-weighted contrast-enhanced MRI, sagittal view, C - T1-weighted contrast-enhanced MRI, axial view, D - T2-weighted MRI, frontal view. *The images demonstrate adequate brainstem decompression and residual tumor fragments in the parastem region.*