Clivus Chordoma: Case Report and Current Considerations on Endoscopic Endonasal Trans-Sphenoid Surgery with Neurosurgeon-Otolaryngologist Collaboration

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Abstract

Chordomas, infrequent malignancies primarily located along the craniospinal axis, showcase gradual growth and localized bone destruction, with the clival region involved in approximately 25-35% of cases. Headaches accompanied by neurological deficits are the typical clinical presentations. Complete surgical resection is the mainstay, with recent collaborative efforts between otorhinolaryngologists and neurosurgeons leading to a positive shift from traditional craniotomic procedures to endoscopic endonasal approaches, fostering minimally invasive techniques and utilizing endoscopy for primary visualization across the neuraxis. Furthermore, the concept of team surgery has been introduced, involving simultaneous contributions from ENT surgeons and Neurosurgeons at all stages of the procedure, including the approach, resection, and reconstruction phases. This report presents a series of two successful cases of clival chordomas managed using the endoscopic endonasal approach at Siloam Hospital Lippo Village This indicates its potential as a viable surgical choice, particularly within medical centers that possess the necessary specialties. Successful implementation is notably enhanced through collaborative efforts between otolaryngologists and neurosurgeons, underscoring the significance of interdisciplinary teamwork.

Introduction

While a spectrum of immune and endocrine neoplasms can also occur, chordomas and chondrosarcomas are the most frequently encountered neoplastic pathologies involving the clivus.[1] Characterized as rare tumors (with an incidence of 0.08 per 100,000), chordomas originate from embryonic notochordal tissue and tend to exhibit slow growth, often forming within the bone. Although they can affect individuals of all ages, chordomas most commonly emerge during the third to the fifth decade of life, with a slight predominance in males.[2] The tumors in question display a low to intermediate malignancy and are characterized by their tendency to show local aggression.

Metastases are rare, usually occurring after recurrence. Primarily localized in the sacrum, clivus, or cervical vertebrae, these lesions often affect the distal and proximal ends of the spine. Caudal chordomas are observed in about 50% of cases, while a third of cases involve cranial chordomas affecting the clival region. These cranial chordomas typically appear as extradural midline masses and may involve cranial nerves, particularly the abducens nerve, during the initial presentation. [2] The incidence of chordomas varies depending on the anatomical location, with the sacrococcygeal region being the most common site (50-60%), followed by the spheno-occipital region (25-35%), and the vertebral column (10%).[3] Moreover, chordomas occur more frequently in males compared to females, highlighting a higher incidence in the male population. [4]

Chordomas in the cervical spine usually originate from parapharyngeal or paravertebral masses, accounting for approximately 3 to 7% of cases.[5] These chordomas develop from residual fetal or embryonic notochord remnants, which normally contribute to the formation of the nucleus pulposus in intervertebral discs. However, in the cephalic region of the notochord, these remnants differentiate into precursor cells responsible for forming the sella, the posterior body of the sphenoid, and the basiocciput bone.[6] The malignant transformation typically occurs in individuals aged between 50 and 60 years, with a relatively low incidence rate observed before the age of 40.[7] Chordomas in children tend to exhibit aggressive behavior. Surgical resection is the primary treatment objective, as it offers the most favorable prognosis. Patients who undergo complete tumor resection have a more optimistic outlook. The surgical challenge presented by the clivus arises from its intricate anatomy and its proximity to critical neurovascular structures.[8] Its anatomical structure is divided into three distinct regions: the upper clivus, mid clivus, and lower clivus. Traditionally, these regions have necessitated distinct surgical approaches, such as the orbito-zygomatic and transsphenoidal routes. [9]

The sphenoid sinus, positioned posterior to the ethmoid sinuses, carries significant anatomical importance for both neurosurgeons and ENT surgeons. [10] The evolution of the endoscopic endonasal approach (EEA) to the sphenoid sinus has taken distinct paths in neurosurgery and otolaryngology. Initially introduced in the early 1990s for pituitary lesion skull base surgeries, the endoscope emerged as a collaborative creation between otolaryngologists and neurosurgeons.[11] Collaboration between these two specialties proves vital for endoscopic endonasal transsphenoidal approaches due to the potential challenges and complications inherent in these procedures. This need for collaboration

arises from the structural variations, abnormalities, and complexities that neurosurgeons encounter during EEA to the sphenoid sinus.[12] Recent innovations, particularly within the realm of EEA, have revolutionized access to the clivus. This technique not only reduces complications but also enables local reconstructive flaps. EEA not only plays a pivotal role in managing chronic sinusitis and accessing the clival region but also empowers otolaryngologists to address a range of sinonasal disorders, even those involving complex sphenoid sinus lesions. The positive outcomes achieved through endoscopy further motivate neurosurgeons to embrace endoscopic techniques for clival procedures, drawn by the improved visualization they offer. [13,14] The triumph of endoscopic transsphenoidal approaches hinges on dynamic collaboration between neurosurgeons and ENT specialists.[12,15] Such collaboration proves indispensable when navigating through nasal variations, intricate skull base lesions, and multifaceted surgical interventions. In cases involving intrasellar lesions, neurosurgeons frequently take the lead in procedures at low-volume centers, with ENT surgeons playing a pivotal role in extended approaches.[12]

The emphasis on minimal invasiveness in the endoscopic approach not only facilitates the realization of "four hands" surgery but also confers notable advantages.[14] Collaborative endeavors between neurosurgeons and otorhinolaryngologists yield distinctive benefits, including heightened spatial orientation within the nasal cavity, simultaneous management of pathologies, and time savings in surgical procedures due to the specialized expertise each specialist contributes.[14] This has been further enhanced by the introduction of the team surgery concept, wherein ENT surgeons and neurosurgeons work in tandem across all stages of the procedure (approach, resection, and reconstruction).[16] This paradigm shift has also reshaped the surgical landscape, elevating the endonasal route to the primary approach for treating such lesions in carefully selected patients, relegating external approaches to a secondary option. The endonasal approach has indeed revolutionized skull base surgery, enabling less aggressive procedures that reach deep-seated structures without necessitating craniotomy and brain retraction. [16]

Case Report

We present two cases of clival chordomas that were observed between 2022 and 2023. In both cases, the surgical resection of the tumor was performed using an endoscopic endonasal approach, with collaboration between neurosurgeons and otolaryngologists.

First, a 20-year-old female who has been experiencing limited eye movement and double vision in the right eye for the past 3 years, accompanied by associated

pain. Initially, the double vision only occurred during lateral gaze, but gradually extended to straight-ahead vision. The left eye showed no complaints, and there was no history of prior illness. Neurologically, there was evidence of paresis in the right V1 cranial nerve and diplopia. Histopathological examination revealed multilayered columnar epithelial tissue, loose and dense connective tissue, atypical stellate-shaped nuclei, some with red cytoplasm, and myxoid stroma. There were also areas of necrosis, hemorrhage, trabecular bone, and granulation tissue, consistent with chordoma.

Second, a 21-year-old female patient who presented with blurred and unfocused vision in her left eye, accompanied by intermittent headaches over the past 2 years. The intensity of the headaches has been progressively increasing, along with worsening of her visual symptoms. Neurologically, there was evidence of paresis in the left VI cranial nerve and the right V cranial nerve. Other examination results were within normal limits. One year prior, the patient had been diagnosed with chordoma. MRI of the head revealed a contrast-enhancing, inhomogeneous, and destructive mass involving the clivus, intrasellar, suprasellar, and left parasella, extending into the sphenoid sinus and left prepontine cistern, measuring approximately +/- 4 x 3.1 x 2.6 cm, with a suggestion of chordoma.

The two patients were subsequently scheduled to undergo a resection procedure through collaborative efforts between an otolaryngologist and a neurosurgeon, using Endoscopic Endonasal Transsphenoidal Surgery (EETS). Both procedures followed the same approach, adhering to the principles of the binostril four-hand technique. The otolaryngologist initiated the procedure by creating a Hadad-Bassagasteguy flap. Subsequently, they established a path leading to the clivus through the sphenoid sinus. This involved performing a sphenoidotomy and a posterior septectomy to ensure more adequate access. Following this phase, the neurosurgeon proceeded with the surgical drilling towards the clivus, performing a subtotal removal of the chordoma. Upon its completion, the otolaryngologist took over to close the defect using a fibrin glue flap.

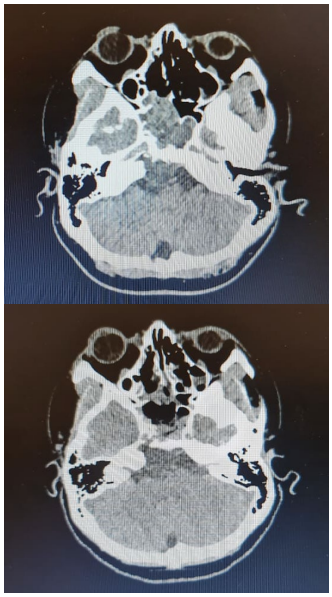

Figure 1. Image of chordoma size on pre- and post-operative non-contrast CT scans, showing a reduction in size on case 1

The CT scan (**figure 1.**) and contrast/non-contrast head MRI examinations in both cases revealed a significant decompression of the chordoma tumor mass following the endoscopic endonasal transsphenoidal surgery. In the first case, the dimensions were reduced to 3.3 x 5.9 x 2.8 cm, while in the second case, they were approximately +/- 2.6 x 1.5 x 1.8 cm.

After the operation, the first patient still complained of persistent shadowing vision and limited right eye movement, but some improvement was observed. Additionally, the patient had complaints of nasal congestion. 5 days later the patient was discharged and received prescribed medication upon returning home. For the second case, histopathological examination of the tissue obtained during the procedure showed a lobulated tumor mass composed of round nuclei resembling clustered epithelioid cells. These epithelioid cells exhibited pleomorphic nuclei with cytoplasm resembling physaliphorous cells. Necrosis and hemorrhage were also evident, consistent with a diagnosis of chordoma. Consequently, both patients were scheduled for multisession gamma knife treatment.

Discussion

Clival chordoma is a midline tumor known for its locally aggressive behavior at the spheno-occipital synchondrosis.[17] The concept of this tumor originating from remnants of the notochord was initially proposed by Muller, and later in 1894, Ribbert provided support for this theory after reviewing 5 cases.[18] Clival chordomas gained recognition as malignant tumors following the first reported mortality case in 1903.[18] In patients afflicted with clival chordomas, the typical presentation involves intractable headaches accompanied by neurological deficits, predominantly cranial nerve neuropathy.[19] Clival chordomas often manifest with abducens nerve involvement, resulting in diplopia, which is a common complaint. Additionally, rare symptoms and signs such as intracranial hemorrhage, epistaxis, and, in more uncommon cases, hearing loss, vertigo, and facial paralysis may present depending on the tumor's extent of growth.[20]

The narrow window of the clivus poses a challenging operative field for neurosurgeons. Adding to this complexity is the lack of clear protocols for managing clival chordomas, mainly due to its rarity. [17] Consequently, most reported cases exist in case series, resulting in limited literature and a scarcity of evidence-based treatment strategies for clival chordomas.[17] Nonetheless, it is established that the 5-year survival prognosis for treated clival chordomas falls between 50% and 85%, with the most favorable outcomes observed in cases where complete resection was achieved.[20] In the early

twentieth century, open approaches like transoral and transpalatal methods were the preferred means to access the clivus. [18] However, these approaches posed significant risks of morbidity, particularly cranial nerve injuries, as surgeons had to meticulously dissect around the delicate cranial nerves and vascular structures to reach the midline. [22] Furthermore, the open approaches showed high recurrence rates in clival chordoma surgeries due to the challenge of dealing with the lateral boundaries bounded by the cranial nerves and vascular system.

The advancement of endoscopic technology has significantly benefited surgeons in accessing the central part of the skull base, particularly the clivus. The direct anterior approach to the clivus offers several advantages, including the ability to avoid neurovascular structures at the lateral boundaries, resulting in a less invasive and lower morbidity procedure for patients. The continuous evolution of the endoscopic endonasal technique has enabled successful complete resection of clival chordomas. The integration of neuronavigation has become an essential tool in assisting surgeons to confirm vital landmark structures during dissection. [20] By combining MRI and CT scan data, the surgeon gains a comprehensive understanding of the operative field, including both bony structures and surrounding soft tissues.[23] The synergy between endoscopic endonasal methods and neuronavigation enhances safety and precision during surgery. Notably, virtual reality imaging has demonstrated an average shortest distance of 18.0 ± 1.8 mm between bilateral boundaries, with neuronavigation allowing for lateral extension of drilling in the superior and middle clivus, safeguarding the internal carotid artery from potential injury.[24] Nevertheless, some complications have been reported with this method, including persistent CSF leakage, hypopituitarism, temporary diabetes insipidus, and meningitis. Studies by Alessandro Paluzzi et al. have revealed promising results, recording a gross total resection rate of 83% for new chordomas and 44% for recurrent cases using the endoscopic transnasal method.[25] Following gross total or near-total resection of chordomas with endoscopic transnasal surgery, patients are typically recommended for radiochemotherapy to minimize the likelihood of recurrence and complete the comprehensive treatment approach.

In this case report, we present the successful management of 2 patients diagnosed with clival chordoma utilizing the endoscopic endonasal resection method with the collaborative efforts of neurosurgeons and otolaryngologists. Prior to the surgical procedure, prophylactic antibiotics were administered to both patients to prevent the occurrence of meningitis. Following the surgical intervention, both patients underwent

gamma knife therapy as an additional treatment to address the residual tumor and enhance local tumor control. The utilization of gamma knife therapy aimed to further reduce the tumor burden and improve the overall treatment outcome. In both of these cases, the chosen therapeutic approach involved the utilization of Endoscopic Endonasal Transsphenoidal Surgery (EETS) in a collaborative effort between otolaryngologists and neurosurgeons. This decision was made after carefully considering the manifold merits of skull base surgery in contrast to external surgical procedures such as craniotomy. The location of clival chordomas, situated within the midline of the skull base and surrounded by a multitude of intricate neurovascular structures, engenders a heightened risk of morbidity when managed via an external approach. EETS, on the other hand, offers a less invasive surgical intervention. Collaborative efforts between otolaryngologists and neurosurgeons span the entirety of the procedural spectrum, encompassing the approach, resection, and reconstruction phases.

Employing the "four-hand" concept, neurosurgeons in these cases benefit significantly in terms of enhanced visualization and orientation during tumor resection. This advantage is attributable to the prior assistance provided by otolaryngologists in facilitating improved access to the clivus. This is achieved through their extensive expertise in navigating the intricacies of nasal cavity structures and intervening in nasal cavity anatomy, particularly when confronted with variations or pathological features. This yields a distinct advantage, as the time required to access the sphenoid sinus is generally expedited when under the purview of otolaryngologists, with a concomitant reduction in endonasal complications. Neurosurgeons also enjoy greater procedural flexibility compared to undertaking EETS in isolation. The collaborative decision-making process fosters cross-fertilization of ideas, ultimately resulting in the selection of the optimal course of action. Additional advantages encompass a diminished incidence of morbidity, the absence of visible scarring, reduced surgical durations, and shorter postoperative convalescence periods.

In these specific cases, postoperative patients exhibited a swift recovery, necessitating only a brief 5-day hospital stay with minimal discomfort. Furthermore, patients were able to swiftly resume their daily activities. These outcomes underscore the inherent advantages of EETS over craniotomy, particularly when executed through direct collaboration between otolaryngologists and neurosurgeons.

Following the treatment, all patients underwent neuro-radiological imaging to

monitor their progress, considering the known high recurrence rate of clival chordoma. Both patients were kept under close follow-up, and subsequent MRI scans revealed a reduction in the size of the residual tumors, with the patients experiencing full restoration of their daily activities. These favorable results demonstrate the effectiveness of the endoscopic endonasal method in managing clival chordomas and achieving sustained remission in the long term. The successful outcomes further support the use of this approach as a viable treatment option for clival chordomas, emphasizing its potential for favorable long-term prognosis and improved patient quality of life.

Conclusion

Prior to the development of the endoscopic endonasal method, the resection of clival chordomas was exclusively performed by neurosurgical teams using open approaches. However, this approach was associated with a high incidence of recurrence and significant morbidity. With the introduction of endoscopic endonasal surgery, the quality of life for patients with clival chordomas can now be better maintained.

Extended Endonasal Transsphenoidal Surgery is a collaborative endeavor that goes beyond individual expertise. Collaboration between neurosurgeons and ENT surgeons is indispensable even for simpler approaches. The practical implications of endonasal structural variations underscore the importance of thorough preoperative assessments to minimize surgical complications. Additionally, the stepwise surgical approach to the sphenoid sinus highlights the significance of collaborative interventions in addressing complex anatomical challenges. EETS thrives on the unity of medical disciplines, utilizing collective expertise to optimize patient outcomes and surgical success.

Nevertheless, the limitation of this case series lies in its small sample size, which is a consequence of the rarity of this disease. Future innovations in this method should be pursued to further enhance and optimize the resection of clival chordomas, ensuring better outcomes for patients in the long run. Continued research and advancements in the field hold the promise of improving the management and prognosis of this challenging condition.

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