

Endoscopic Management of a Lower Clival Chondroid Chordoma: Case Report

Alt Klivus Yerleşimli Kordomanın Endoskopik Yol ile Tedavisi: Olgu Sunumu

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ABSTRACT

We report herein a case of lower clival chondroid chordoma, focusing on the surgical procedure of endoscopic endonasal surgery. A 36-year-old woman presented with progressive headache, right shoulder pain, and right hypoglossal nerve palsy. Computed tomography (CT) and magnetic resonance (MR) imaging revealed an extradural tumor located in the lower clivus, including the anterior aspect of the foramen magnum, deeply compressing the medulla and upper cervical spinal cord. Endoscopic endonasal surgery was performed via two nostrils. Since the basiocciput was destroyed by the tumor, removal of the tumor allowed identification of the middle clivus superiorly, the anterior arch of the atlas inferiorly, and anteromedial parts of occipital condyles bilaterally without drilling the basiocciput. The tumor was removed except for laterally and inferiorly extended lesions. Pathological diagnosis was chondroid chordoma. Postoperative course was uneventful, and the patient was discharged without further neurological deterioration. Endonasal endoscopic surgery provided safe and reliable tumor resection for a lower clival lesion. We believe that this minimally invasive procedure should be considered as an alternative to traditional surgical treatment.

KEYWORDS: Endoscopic endonasal surgery, Chondroid chordoma, Lower clivus, Foramen magnum, Upper cervical

ÖZ

Bir olguda, alt klivusa yerleşmiş kondroid kordomanın endoskopik endonasal yol ile cerrahi tedavisi ele alınmıştır. Otuz altı yaşında kadın hasta, giderek artan baş ağrısı, sağ omuz ağrısı ve sağ hipoglossal sinir felci semptom ve bulguları ile hastanemize kabul edildi. Bilgisayarlı tomografi ve manyetik rezonans görüntüleme çalışmasında alt klivus ve foramen magnumun anterior kenarını ektradural olarak doldurmuş medulla ve üst servikal spinal korda bası yapan kitle lezyonu görülmüştür. Her iki nostril kullanılarak nasal endoskopik cerrahi yapıldı. Basioksiput tümör tarafından harap edildiği için, tümörün çıkarılması ile klivusun orta kesimine üstten, atlasın anterior arkına alttan ve oksipital kondillerin anteromedial kesimlerine iki taraflı olarak cerrahi diril kullanılmadan ulaşıldı. Alt dışa uzanım gösteren tümöral yapı dışındaki tümör dokuları çıkarıldı. Patolojik tanı kondroid kordoma olarak geldi. Ameliyat sonrası dönemde ek sorunu olmayan hasta ek nörolojik defisit oluşmadan taburcu edildi. Endoskopik nasal cerrahi yöntem ile alt klival lezyonlar güvenli bir şekilde çıkarılabilmektedir. Bizim düşüncemize göre; minimal invaziv girişimler geleneksel cerrahi yöntemlere bir seçenek olarak düşünülmelidir.

ANAHTAR SÖZCÜKLER: Endoskopik nasal cerrahi, Kondroid kordoma, Alt klivus, Foramen magnum, Üst servikal

INTRODUCTION

Chondroid chordoma is a variant of chordoma with a cartilaginous component. This tumor commonly occurs as a clival lesion including the lower clivus, which is still a challenging lesion to approach. Traditionally, transcranial-transbasal, extended frontal, transmaxillary, and transoral approaches are performed for lower clival lesions (2), but recent advances in endoscopic endonasal surgery have contributed to the application of midline skull base surgery. Some clival chordoma series by extended endoscopic endonasal approaches have been reported (5, 7, 12, 17), but have not sufficiently described the surgical procedure and management of the lower clival tumor. We report

herein a case of lower clival chondroid chordoma, focusing on the surgical procedure and management by endoscopic endonasal surgery.

CASE REPORT

A 36-year-old woman presented with a 5-year history of progressive headache and right shoulder pain. On admission, neurological examination revealed right hypoglossal nerve palsy. Magnetic resonance (MR) imaging after gadolinium injection demonstrated an enhanced tumor in the lower clival region, including the anterior aspect of the foramen magnum. The tumor deeply compressed the medulla and upper cervical spinal cord. The tumor extended to the right side and was surrounding the right occipital condyle,

and also extended inferiorly behind the odontoid process (Figure 1A, B). Computed tomography (CT) showed that the tumor had destroyed the lower clivus and anteromedial part of the right condyle. The right hypoglossal canal was filled with tumor. The tumor was localized in the extradural space and contained some calcified components (Figure 2A, B). Endoscopic endonasal surgery was performed via two nostrils. The patient was anesthetized by endotracheal intubation without tracheostomy. The procedure was performed with a 4-mm, 0° rigid endoscopy (Endoarm; Olympus, Tokyo, Japan). The endoscope was inserted into the right nostril and advanced to the inferior meatus. Subsequently, the nasopharynx and bilateral Eustachian tubes were identified. A nasal flap was then prepared with the right septal mucosa in case of cerebrospinal fluid (CSF)

leakage. The vomer was resected and the superior aspect of the hard palate was flattened sufficiently, providing a wide surgical corridor without opening the sphenoid sinus (Figure 3A). Mucosa of the nasopharynx and the longus capitis muscle were cut concurrently with a monopolar coagulator (Colorad needle; Stryke, Michigan, USA) in a reverse U shape, and the flap was reflected downward, exposing the tumor (Figure 3B). The tumor was safely debulked, and bleeding from the tumor was easily controlled because of poor vascularization. Since the basiocciput had been destroyed by the tumor, removal of the tumor allowed identification of the middle clivus superiorly, the anterior arch of the atlas inferiorly, and anteromedial parts of occipital condyles bilaterally without drilling the basiocciput. After tumor removal, the dura mater was preserved without CSF leakage (Figure 3C). Tumor

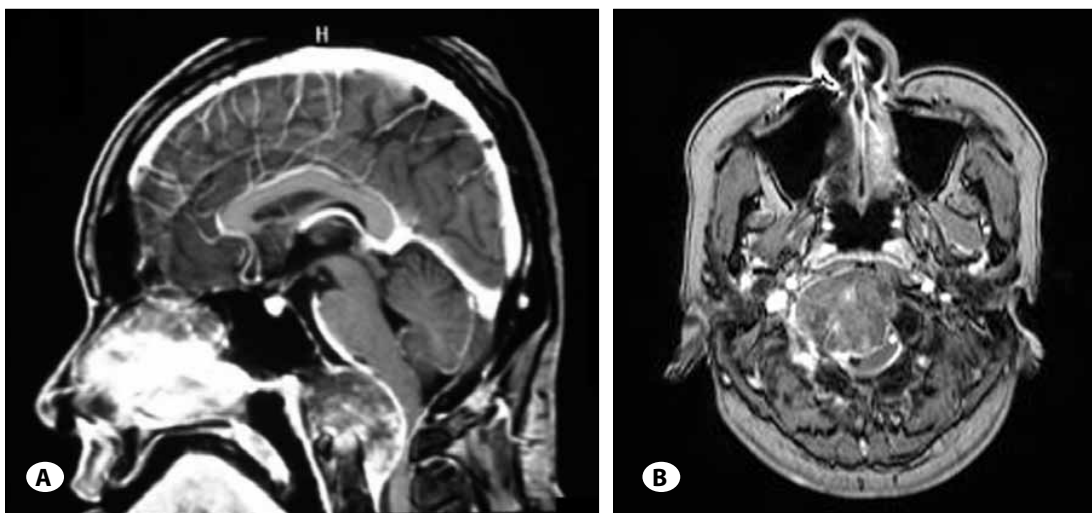


Figure 1: T1-Gd weighted sagittal (A) and coronal (B) MR imaging revealing a lower clival tumor including the anterior aspect of the foramen magnum. The tumor extends to the right side and surrounds the right occipital condyle, and also extends inferiorly behind the odontoid process. The tumor is deeply compressing the medulla and upper cervical spinal cord.

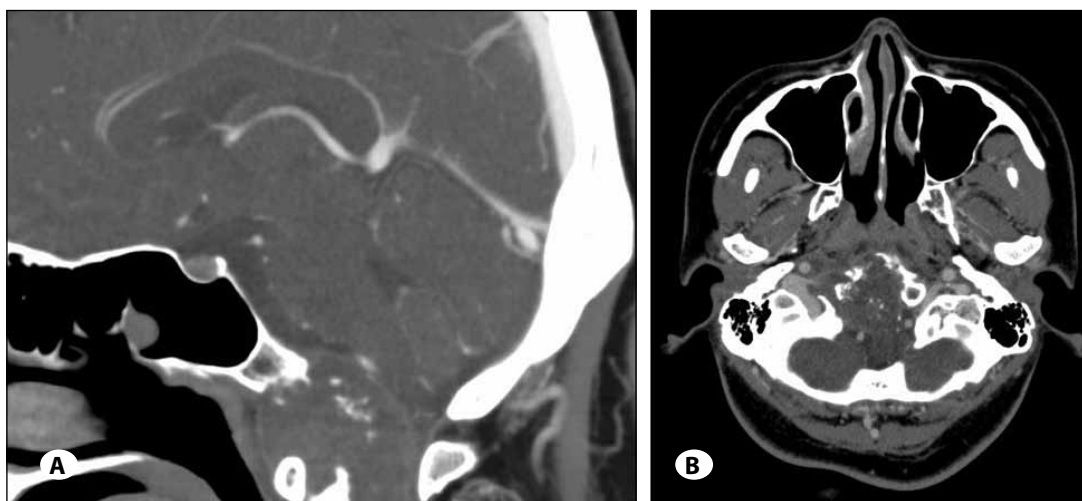


Figure 2: Sagittal (A) and coronal (B) enhanced CT showing that the tumor has destroyed the lower clivus, anteromedial part of the right occipital condyle and tip of the odontoid process. The right hypoglossal canal is filled with tumor. The tumor is localized in the extradural space, and contains some calcified components.

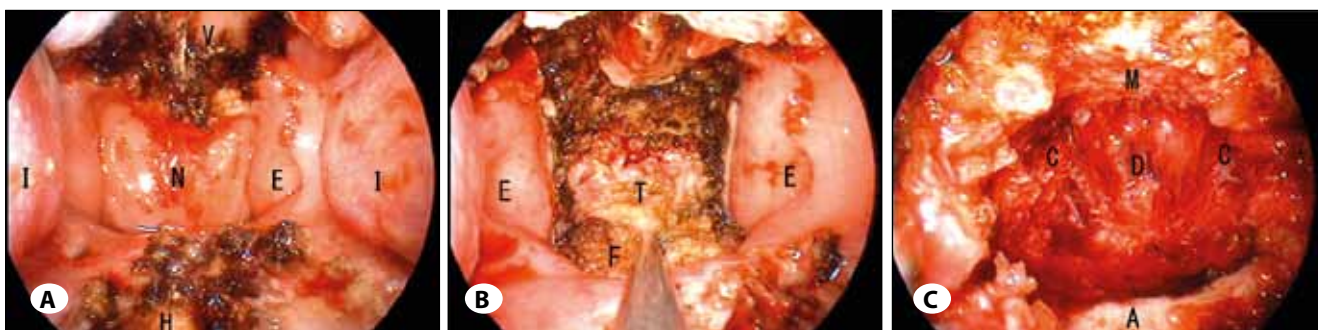


Figure 3: **A)** Intraoperative view after creating the surgical corridor. N, Nasopharynx; E, Eustachian tube; V, Vomer; I, Inferior turbinate; H, Hard palate. **B)** Intraoperative view after tumor exposure. T, Tumor; E, Eustachian tube; F, flap of the nasopharyngeal mucosa and longus capitis muscles. **C)** Intraoperative view after tumor removal. D, Dura mater; M, Middle clivus; A, Anterior arch of the atlas; C, Anteromedial part of the occipital condyle.



Figure 4: Postoperative T2-weighted sagittal **(A)** and coronal **(B)** MR imaging, showing sufficient decompression by tumor resection. Arrowheads indicate residual tumor located outside the right occipital condyle **(B)** and behind the odontoid process **(A)**.

extending laterally beyond the right occipital condyle and inferiorly behind odontoid process was left intact. Free fat and fibrin glue were placed in the surgical defect. Pathological diagnosis was chondroid chordoma. Postoperative course was uneventful. The day after surgery, the patient was allowed to walk independently. Postoperative MR imaging showed sufficient decompression with tumor resection (Figure 4A,B). The patient was discharged without further neurological deterioration.

DISCUSSION

Chordomas are tumors arising from rest of primitive notochord along the craniospinal axis. The most common site is the sacrococcygeal region (50%), followed by the skull base (35%) and vertebral bodies (15%) (6). Chordoma account for approximately 1% of all intracranial tumors and the retroclival region is the most common site (1).

Chondroid chordoma, first described by Heffelfinger in 1973, is a variant of chordoma that straddles the bridge between conventional chordoma and chondrosarcoma (8). About 28-34% of skull-base chordomas are of the chondroid variety (8). Chordoma usually occurs between the fourth and fifth decades of life, whereas chondroid chordoma present at an earlier age and has a female predilection (1, 15). Chondroid chordoma has a lower recurrence rate and better survival as compared to conventional chordoma (8). However, the location is challenging to operate, it commonly occur the clival region. Safe, sufficient decompression is the most important issue for lower clival tumors, particularly in cases involving the foramen magnum. Traditionally transcranial-transbasal, extended frontal, transmaxillary, and transoral approaches are performed for anterior approaches to the foramen magnum (2). However, such approaches carry a high risk of morbidity involving neurovascular structures. These

traditional approaches reportedly offer a total resection rate of 44-83%, neurological morbidity in 0-80% of cases, vascular injury in 9-12%, and CSF leakage in 8.3-30% (11-13, 17).

Conversely, recent advances in endoscopic endonasal surgery have contributed to development of midline skull base surgery. Some reports have described clival chordoma series via extended endoscopic endonasal approaches (5, 7, 9-10, 12, 17). In a recent report of clival chordoma, Stippler et al. reported in their series that nearly 50% of clival chordomas included lower clival lesions, and described a high resection rate for newly diagnosed chordoma (total resection, 66.7%; near-total resection, 16.7%) and a low morbidity rate (neurological complications, 5%; vascular complications, 3.8%) (17). The CSF leakage, as the biggest concern in endonasal endoscopic approaches, was limited to 5.4% with the use of a nasal flap. In addition, other reports have described 9 cases (5, 10, 14, 16) of lower clival chordoma, including 2 cases involving the foramen magnum (10, 14). In those 9 cases, CSF leakage occurred in 1 case (12%) that was successfully repaired, and with complications of permanent hemiparesis in 1 case (12%). Although the number of reported cases is still low, these data suggest that an endonasal endoscopic approach to lower clival chordoma is safe and reliable.

Furthermore, traditional transoral approaches to lower clival lesions require tracheostomy before surgery and incision of the soft palate, burdening patients with postoperative velopharyngeal incompetence, hypernasal speech, swallowing disturbances, and temporomandibular joint syndrome (2). An endoscopic endonasal approach both avoids such unpleasant procedures and improves patient satisfaction after the surgery.

However, endoscopic endonasal surgery has lateral limitations at the level of the foramen magnum. Cavallo et al. reported the lateral limitation of the foramen magnum is the anterior third of the occipital condyle, as the hypoglossal canal is located anterior and in the middle third of each condyle (3-4). Laterally extended lesions must be treated by additional surgical approaches or radiotherapy.

In conclusion, endonasal endoscopic surgery provides safe and reliable tumor resection of lower clival lesions. We believe that this minimally invasive procedure should be considered as an alternative for traditional surgical treatment.

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