Clivus chordoma case series

Ceylan et al. retrospectively analyzed the endoscopic endonasal surgeries in the Pituitary Research Center and Neurosurgery Department of the Kocaeli University Faculty of Medicine, Turkey between January 2004 and December 2019. We retrospectively reviewed the medical data, radiological images, and surgical videos of patients, and 72 patients with chordoma and chondrosarcoma were included in the study.

Results: Based on pathology reports, 72 patients (seven pediatric) were identified, to whom 91 endoscopic operations were performed. We determined the surgical limitations for each clival segment as superior, middle, and inferior. Then, we divided these into three subgroups according to whether the tumor shows dural invasion (extradural chordoma, large extradural - minimal intradural component, and minimal extradural - large intradural component). The tumors of 19 (26.4%), 25 (26.4%), and nine (12.5%) patients originated from the superior, middle, and inferior clivus, respectively. Nineteen (26.4%) patients had panclival involvement. GTR was performed in 47 (65.3%) patients. The GTR rate in patients with panclival tumors was 47.3% (9/19). The experience, lateralization, dural involvement, and origin of the clivus affecting GTR were analyzed. Extradural - intradural extensions were verified as negative predictor factors for GTR, whereas tumors located in the superior (OR: 16.710, p=0.030) and middle (OR: 11.154, p=0.023) segments were positive predictive factors for GTR.

Conclusion: An increasing experience in endoscopic surgery significantly increases the GTR rates by widening the surgical limitations. Due to dense bone infiltration and adhesion to critical neurovascular structures, recurrence rates are high despite performing GTR. Although surgery and adjuvant treatments improve the 5-year survival of patients, the mortality rates remain high. Therefore, surgery of these tumors should be performed by experienced centers. In addition to surgical and adjuvant therapies, targeted molecular and translational biological therapies are also needed for chordomas and chondrosarcomas in the future [1)]

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present an alternative minimally invasive approach and reconstruction technique for selected clival chordomas.

Methods: Three cases of clival chordomas illustrating this technique are provided, together with an operative video.

Results: The mucosa of the rostrum is incised and elevated from the underlying bone, as first step of surgery. Following tumor resection with angled scope and instruments, the mucosa of the sphenoid sinus (SS) is removed and the tumor cavity and SS are filled with abdominal fat. The mucosal incision of the rostrum is then sutured. A hangman knot is prepared outside the nasal cavity and tightened after the first stitch and a running suture is performed.

Conclusion: We propose, in this preliminary report, a new targeted approach and reconstruction strategy, applying to EEAs the classic concept of skin incision and closure for transcranial approaches. With further development in the instrumentations and visualization tools, this technique may become a valuable minimally invasive endonasal approach for selected lesions [2)].

Data for 12 consecutive patients underwent GKS for post-operative residual histologically verified clival chordoma at the Department of Neurosurgery and Gamma Knife Center, International Medical Center (IMC), 42km. Ismailia Desert Road, Cairo, Egypt from 2006 through end of 2017 were retrospectively reviewed and analyzed with mean follow-up period of 45 months (range12-120 months).

In the last follow up MR, tumor growth control was achieved in 33.3% of patients (mean treated tumors volume was 2.7cc with mean peripheral prescription dose of 16 Gy), and 66.7% of patients reported lost tumor growth control (mean treated tumor volume was 9.2 cc with mean peripheral dose was 13.5 Gy). The overall tumor free progression with mean follow up period of 45mos was 33.3%. The Actuarial 2, 3 and 5 year tumor control rates after initial GKS was 35%, 30% and 25% respectively.

Without satisfactory maximum tumor reduction and sufficient high peripheral prescription radiation tumor dose, it should not be expected that GKS could efficiently control the progression of residual clival chordoma, especially for long term [3)].

A retrospective study evaluated patients treated at a single center from 1992 to 2017. During the study period, 24 patients underwent 24 surgeries. Twenty-two surgical resections (including 1 initial surgery and 1 surgery for recurrence) were deemed maximally safe cytoreductive resections (92%); the remaining 2 surgeries were deemed incomplete (8%), which were histologically confirmed in all but in 1 case (which involved radionecrosis). The complications were divided into endocrinologic, neurologic, and other complications. In 1 case (4%), surgery led to immediate dyspnea followed by death on the following day; in another case (4%), ischemic infarction led to sudden death. In 3 cases (13%), patients exhibited improvements of neurologic (visual or oculomotor) deficits that had been observed prior to surgery. The following new postoperative neurologic deficits were observed: oculomotor deficits in 4 cases, dizziness in 2 cases, and cranial nerve-attributed dysphagia in 3 cases. About 19 patients underwent adjuvant postoperative radiotherapy following the initial surgery (dose: 54.5 Gy in all cases). The mean and median follow-up durations were 50 ± 53 and 48.5 months, respectively. A Kaplan-Meier analysis estimated a median survival duration of 50.2 months (95% confidence interval 27.9-72.4 months). These findings highlight the importance of interdisciplinary treatment strategies, particularly those combining maximally safe cytoreductive tumor resection and adjusted radiotherapy and other treatment options, for patients with relatively good conditions [4)].

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14 consecutive clivus chordoma cases undergoing maximum surgical resection followed by intensity-modulated radiotherapy with simultaneous integrated boost (IMRT-SIB), using the institutional protocol from the Severance Hospital, Seoul, Korea, between 2005 and 2013. Total and near-total resections were achieved in 11 patients (78.6 %), partial in 2 patients (14.3 %), and 1 patient (7.1 %) received RT for recurrent tumor after total resection. Gross residual or the high-risk area defined the planning target volume (PTV)1; PTV2 was the postoperative tumor bed plus a 3-5-mm margin, and PTV3 was PTV2 plus a 5-10 mm margin. A moderate hypofractionation schedule was used: doses to PTV1, PTV2 and PTV3 were 3.9 Gy, 3.15 Gy and 2.8 Gy through 15 fractions for the first two patients, and the rest received 2.5 Gy, 2.2 Gy and 1.8 Gy through 25 fractions. The biologically equivalent dose in 2-Gy fractions (EQD2) was 65-68 Gy for PTV1, 52-56 Gy for PTV2, and 44.3-44.8 Gy for PTV3.

Median follow-up was 41 months. Eight patients were free of disease for median 42.5 months (range 23-91 months), four patients had stable disease for median 60.5 months (range 39-113 months), and

1 patient showed partial response for 38 months after RT. Local progression was seen in one patient who received EQD2 67.8 Gy after partial resection. Estimated 5-year progression-free and overall survival rates were 92.9 %. Surgery improved the neurologic deficit in six patients, and IMRT-SIB was well tolerated without lasting toxicity.

The experience suggests that maximum resection and high-dose IMRT-SIB can achieve local control without significant morbidities [5].

Twelve patients with histologically confirmed clivus chordoma were treated at the Johns Hopkins Hospital between 1971 and 1989. Eight of the patients were men and four were women. The mean age at first operation was 51 years (range, 10 to 80). The most common presenting symptoms were headache, diplopia, dysphagia and dysarthria, and facial sensory changes. Computed tomography, with and without contrast enhancement, proved adequate for tumor identification and localization. Magnetic resonance imaging and angiography were occasionally employed to localize the tumors further and to define tumor vascular supply and proximity to vascular structures. Twenty-two resections were performed in 11 patients, and another patient underwent biopsy only. Seven were also treated with radiation therapy. Tumors recurred in eight patients, six of whom underwent further operations. The mean time to first recurrence was 22 months (range 8 to 36 months). Six of the patients are still alive, with a mean follow-up of 31 months (range, 3 to 89 months) from first surgical resection. The mean survival time from first treatment was 31 months (range, 4 to 62 months) among those patients who died. There was no operative mortality. The 5-year cumulative survival rate was 20%. Six patients with long follow-up have had fair to good results, being free of recurrences for at least a year. However, none of the patients returned to their premorbid baseline of activities. Five of the patients had tumors with the histologic diagnosis of chondroid chordoma. Three of these patients are still alive. The mean age at first treatment was 44 (compared with 62 for typical chordomas). The mean time from symptoms to diagnosis was 29 months (typical chordomas, 18 months). The mean length of survival and time to tumor recurrence were not significantly different between chondroid and typical chordomas [6].

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