Clivus chordoma outcome

Clivus chordoma shows poor patient prognosis because of its high recurrence rate. The oOverall 5-year survival is approximately 50% [1].

Even though many clinical factors and biomarkers are reported to be associated with prognosis, no prediction model has been applied clinically. Thus, Zhai et al. aimed to derive and validate a prognostic nomogram to predict progression-free survival (PFS) of chordoma.

A total of 201 patients were randomly divided into a derivation group (151 cases) and a validation group (50 cases). The expression levels of biomarkers were quantified using tissue microarray analysis. A nomogram was established via univariate and multivariate Cox regression analysis in the derivation group. The predictive performance of the nomogram was then tested in the validation group.

The mean follow-up interval was 57 months (range 26-107 months). One clinical factor and 3 biomarkers were confirmed to be associated with PFS, including degree of resection, E-cadherin, Ki-67, and VEGFA. The nomogram with these prognostic factors had areas under the receiver operating characteristic curve of 0.87 and 0.95 in the derivation group at 3 years and 5 years, respectively, compared with 0.87 and 0.84 in the validation group. Calibration and score-stratified survival curve were good in the derivation group and validation group, respectively.

The established nomogram performs well for predicting the PFS of chordoma and for risk stratification, which could facilitate prognostic evaluation and follow-up [2].

They show a strong tendency for local recurrence even after combined surgical and radiosurgical treatment. The possibility of spreading to distant locations of the neuraxis may further complicate the treatment and causes additional morbidity.

A Retrospective review of clival chordomas treated from 1993 to 2013.

Fifty patients (56% male) with median age of 59 years (range, 8-76) were newly diagnosed with clival chordoma of mean diameter 3.3 cm (range, 1.5-6.7). Symptoms included headaches (38%), diplopia (36%), and dysphagia (14%). Procedures included transsphenoidal (n = 34), transoral (n = 4), craniotomy (n = 5), and staged approaches (n = 7). Gross total resection (GTR) rate was 52%, with 83% mean volumetric reduction, values that improved over time. While the lower third of the clivus was the least likely superoinferior zone to contain tumor (upper third = 72%/middle third = 82%/lower third = 42%), it most frequently contained residual tumor (upper third = 33%/middle third = 38%/lower third = 63%; P < .05). Symptom improvement rates were 61% (diplopia) and 53% (headache). Postoperative radiation included proton beam (n = 19), cyberknife (n = 7), intensity-modulated radiation therapy (n = 6), external beam (n = 10), and none (n = 4). At last follow-up of 47 patients, 23 (49%) remain disease-free or have stable residual tumor. Lower third of clivus progressed most after GTR (upper/mid/lower third = 32%/41%/75%). In a multivariate Cox proportional hazards model, male gender (hazard ratio [HR] = 1.2/P = .03), subtotal resection (HR = 5.0/P = .02), and the preoperative presence of tumor in the middle third (HR = 1.2/P = .02) and lower third (HR = 1.8/P = .02) of the clivus increased further growth or regrowth, while radiation modality did not.

The findings underscore long-standing support for GTR as reducing chordoma recurrence. The lower

third of the clivus frequently harbored residual or recurrent tumor, despite staged approaches providing mediolateral (transcranial + endonasal) or superoinferior (endonasal + transoral) breadth. There was no benefit of proton-based over photon-based radiation, contradicting conventional presumptions [3].

Recurrence of clival chordoma due to seeding along the surgical pathway is an infrequent mechanism of treatment failure, with only rare cases documented in the literature. When deciding on the appropriate surgical approach, the surgeon must consider the risk of septal seeding during a transseptal approach. The emergence of transnasal endoscopic skull base approaches may reduce the likelihood of surgical pathway tumor seeding [4].

1)
Tenny S, Varacallo M. Chordoma. 2022 Feb 12. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2022 Jan–. PMID: 28613596.
2)
Zhai Y, Bai J, Li M, Wang S, Li C, Wei X, Zhang Y. A nomogram to predict the progression-free survival of clival chordoma. J Neurosurg. 2019 Dec 27:1-9. doi: 10.3171/2019.10.JNS192414. [Epub ahead of print] PubMed PMID: 31881545.
3)
Jahangiri A, Chin AT, Wagner JR, Kunwar S, Ames C, Chou D, Barani I, Parsa AT, McDermott MW, Benet A, El-Sayed IH, Aghi MK. Factors predicting recurrence after resection of clival chordoma using variable surgical approaches and radiation modalities. Neurosurgery. 2015 Feb;76(2):179-86. doi: 10.1227/NEU.0000000000000611. PubMed PMID: 25594191.
4)
Hines JP, Ashmead MG, Stringer SP. Clival chordoma of the nasal septum secondary to surgical pathway seeding. Am J Otolaryngol. 2014 Jan 2. pii:S0196-0709(13)00301-3. doi: 10.1016/j.amjoto.2013.12.018. [Epub ahead of print]PubMed PMID: 24480512.