Presentation, Treatment, and Long-Term Outcome of Intrasellar Chordoma

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Introduction

Chordoma is a locally aggressive tumor arising from notochord remnants. Little is known about the characteristics and long-term prognosis of intrasellar chordoma. The purpose of this study was to determine common features, treatment approaches and survival characteristics of this rare clinical entity.

Methods

Institutional databases, the Surveillance, Epidemiology, and End Results (SEER) database, and PubMed/EMBASE were queried for chordoma with a primarily intrasellar component (**Figure 1**). The SEER database was also queried for adult skull base chordoma. Patient-level data were extracted where available. Kaplan-Meier survival analyses were conducted.

Results

Among 80 cases, the mean age at presentation was 55.6 (SD: 15.9), with a female predominance (1.16:1.00). Patients experienced symptoms for a mean duration of 19.0 months, including cranial nerve deficits, hypopituitarism, and hyperprolactinemia. Among patients receiving treatment, 77.5% underwent surgery. Additionally, less than half of the patients (34, 47.3%) received adjuvant radiation therapy. The 5-year overall survival (OS) of intrasellar chordoma was 60.0% (SE: 6.9). Patients aged 40 and younger had a 5-year OS of 80.8% (SE: 12.2), compared to patients older than 40, who had a OS of 55.4% (SE: 7.7) (Mantel-Cox, p = 0.044) (**Figure 2**). Males experienced a lower 5-year OS (44.0, SE: 9.7) than females (76.8, SE: 8.5), (Mantel-Cox, p = 0.003). Median OS was greater in patients with skull base chordoma than in patients with intrasellar chordoma (Mantel-Cox, p = 0.046) (**Figure 3**).

Figure 1.

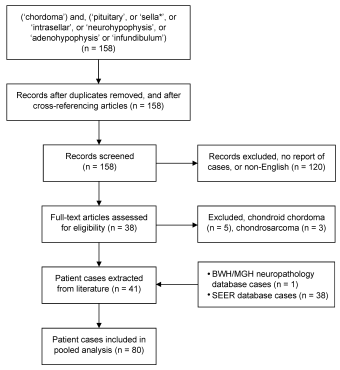
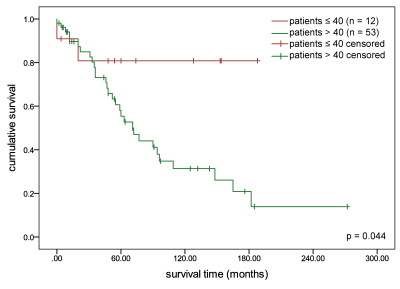
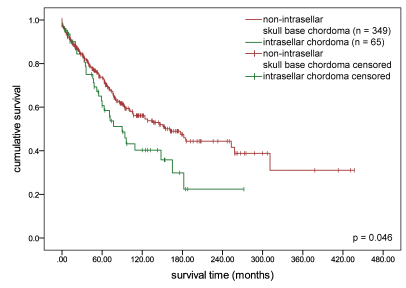


Figure 2.



A sequential comparison by 5-year increments revealed a significant age cut-off. Patients aged 40 and younger had a 5-year overall survival of 80.8% (SE: 12.2), compared to patients older than 40, who had a survival of 55.4% (SE: 7.7) (Mantel-Cox, p = 0.044)

Figure 3.



In patients with follow-up data, an even proportion of patients with intrasellar chordoma were male and female. Median overall survival was greater in patients with skull base chordoma than in patients with intrasellar chordoma (Mantel-Cox, p = 0.046)

Conclusions

This study represents the most comprehensive clinical understanding of intrasellar chordoma. This tumor has a unique clinical presentation and outcome characteristics among skull base chordomas. Young age and female sex predict a better prognosis. Although the relationship between extent of surgical resection and overall survival could not be determined, expert consensus for skull base chordomas is to achieve maximal surgical resection and administer proton beam radiation therapy. Overall, intrasellar chordoma is associated with a poorer prognosis than non-intrasellar skull base chordoma.

Learning Objectives

By the conclusion of this session, participants should be able to:

1) Describe the presentation of intrasellar chordoma.

2) Identify the prognostic indicators of overall survival of intrasellar chordoma.