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Prognostic factors for recurrence and complications in the surgical management of primary chordoid gliomas: A systematic review of literature

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ABSTRACT

Objective: Chordoid gliomas (CG) are rare neoplasms which frequently arise within the third ventricle. Surgery remains the mainstay treatment for CG. The present study comprehensively reviews all reported cases of CG within the literature in order to identify risk factors for surgical complications and tumor recurrence.
Methods: A comprehensive search on MEDLINE (OVID and PubMed), Scopus, Embase, and Web of Science was conducted following PRISMA guidelines to identify all reported cases of CG.
Results: A total of 81 patients met the study criteria which comprised of 33 males and 48 females. Median age at diagnosis was 48 years with a range from 5 to 72 years, and mean tumor size was 3.1 cm. Biopsy, subtotal resection (STR), and gross total resection (GTR) were achieved in 8, 34, and 33 patients, respectively, with six cases not reporting extent of resection (EOR). Thirteen patients underwent adjuvant radiotherapy. Postoperative complications were noted in 30 cases (37%), with new onset diabetes insipidus being the most common. Postoperative morbidity was not associated with age, tumor size, or extent of resection. A trans-lamina terminalis approach demonstrated a strong trend towards decreased overall rates of postoperative morbidity compared to other approaches ($p = 0.051$). GTR was associated with improved progression-free survival (PFS; $p = 0.028$), while adjuvant radiotherapy, age, tumor size and proliferative index were not predictive of patient outcomes.
Conclusion: GTR should be the primary goal for the management of CG, as it is associated with improved rates of tumor control without an increased rate of postoperative complications. Surgical approach was a stronger predictor of complication rates than extent of resection. Morbidity remains high, and future studies to further elaborate on factors predictive of postoperative complications are critical.
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1. Introduction

Intraventricular neoplasms can be highly challenging to treat due to an elevated morbidity profile associated with surgical approach to resection. Tumors that originate in the anterior portion of the 3rd ventricle include ependymomas, central neurocytomas, craniopharyngiomas, and suprasellar meningiomas. Chordoid gliomas (CG) are rare intracranial tumors characterized by the histological presence of both glial and chordoid elements that also occur primarily in this location. Initially described by Brat et al. [1], CG are classified as World Health Organization (WHO) grade II neuroepithelial tumors of uncertain histogenesis [2]. CG are typically found within the suprasellar region and the anterior third ventricle. These tumors present as solid, hyperdense lesions on computed tomography (CT), with uniform contrast enhancement on T1-weighted MRI, and mild hyperintensity on T2-weighted MRI. Cystic components have also been observed on rare occasions [3]. Histologically, CG are comprised of clusters and cords of epithelial cells in a mucinous matrix background with a strong presence of GFAP staining on immunohistochemistry [4].

Their specific location of origin results in a unique symptom profile at presentation consisting of intracranial hypertension symptoms associated with obstructive hydrocephalus, hypothalamic dysfunction, and/or visual impairment. Their close association,

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and sometimes tight adherence, to the hypothalamus makes *en bloc* resection difficult due to the increased risk of post-operative diabetes insipidus (DI) and other neuroendocrine dysfunction. Their growth is indolent and patients may present with persistent symptoms that originated months, or even years, previously. Surgeons have attempted to balance maximal cytoreduction with complication avoidance, and radiotherapy is often used as an adjuvant therapy in patients that undergo subtotal resection. Factors that govern morbidity, mortality, and recurrence in this disease have not been clearly elucidated and there is a lack of large retrospective studies on these tumors due to their rare nature. Various case reports and case series have attempted to illustrate some of these important points. The current systematic review accrued data from 81 patients presented in various case reports and smaller case series in the literature in order to elucidate factors that influence recurrence, morbidity, and mortality, of patients with chordoid gliomas.

2. Materials and methods

2.1. Literature search

Two researchers (LA, WC) each performed independent literature searches on MEDLINE (OVID and PubMed), Embase, Scopus, and Web of Science with the keywords “chordoid” and “glioma” to identify all published reports on CGs. Each database was searched on 3/9/2015 and no publication date restriction was placed on the study. The search was further refined by limiting to manuscripts published in English. Each reviewer constructed a data sheet for each searched database, which were utilized to compare and remove duplicate studies. Data sheets were also compared between researchers in order to agree and confirm the eligible studies included. The Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) guidelines was applied to this review. The protocol was not registered (Fig. 1). Our inclusion criteria were simplified by including all pathologically confirmed cases of intracranial CGs with disaggregated data available for each patient reported. Cases were excluded for studies that did not provide adequate treatment or clinical parameters for each individual patient.

2.2. Data collection

The following variables were collected from each study: age, gender, tumor location, size, duration of symptoms, surgical approach, extent of resection, Ki-67/MIB-1 proliferation index, post-operative complications, adjuvant radiotherapy, progression-free survival, recurrence status, follow-up time, patient clinical status on last follow-up, and overall survival. All components

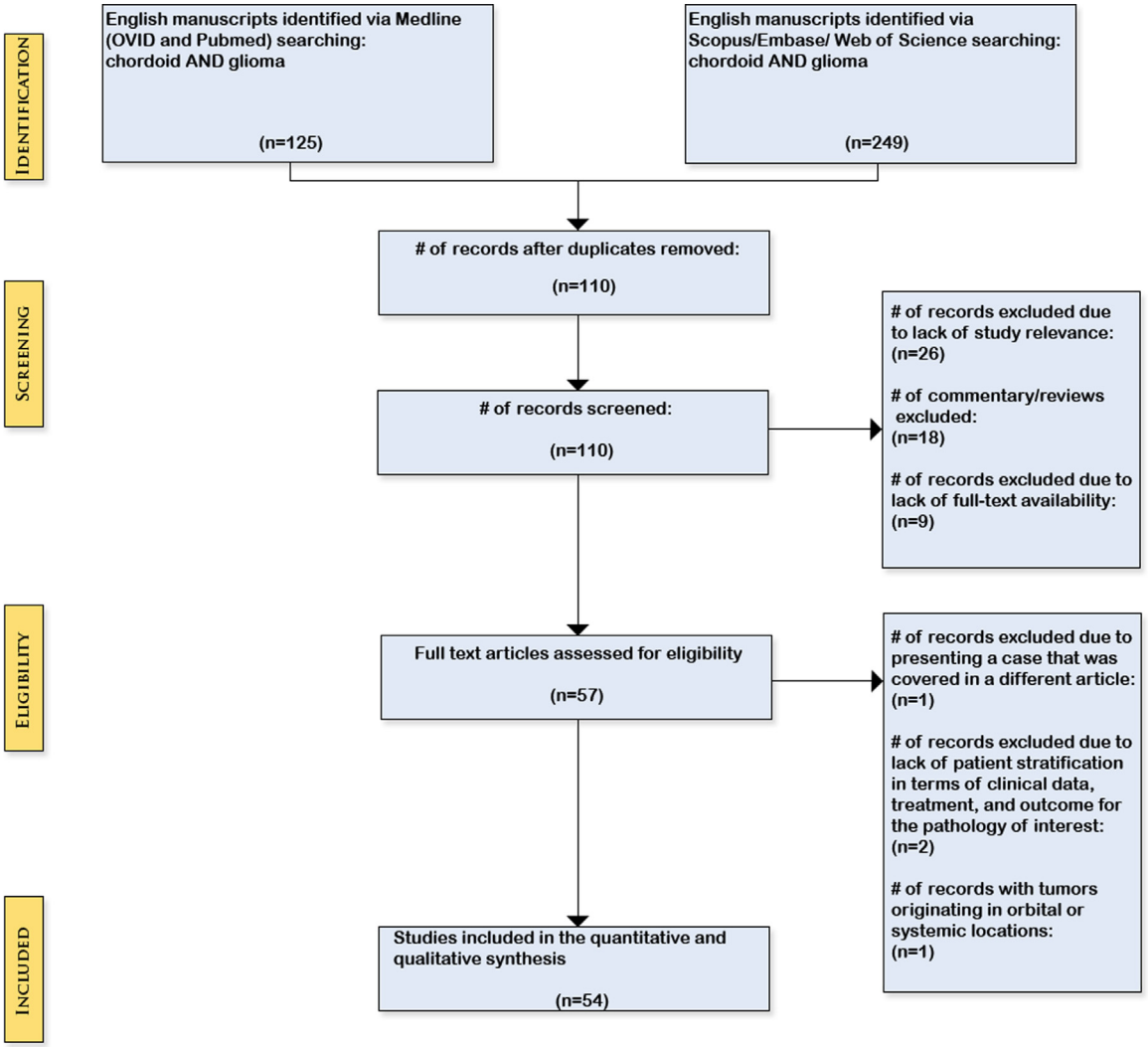

Fig. 1. Preferred reporting items for systematic reviews and meta-analyses (PRISMA).

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