Clinical Study

Choroid plexus papillomas: a single institutional experience

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Summary

Objective: To determine the long-term outcome of resected choroid plexus papillomas (CPPs).

Methods: Medical records and histologic specimens were reviewed for 41 patients (19 male, 22 female; median age, 36 years; range, 6 months to 74 years) with CPP seen between 1974 and 2000. Tumor locations were as follows: 76%, fourth ventricle; 17%, lateral ventricle, and 7%, third ventricle. Fifty-six percent had a gross total resection (GTR) and 44% had a subtotal resection (STR). Median follow-up was 6.5 years.

Results: Five-year local control, distant brain control, and overall survival were 84%, 92%, and 97%, respectively. Comparison of GTR and STR at 5 years showed a significant increase in local control (100% vs. 68%; \( P = 0.04 \)) but not in overall survival (100% vs. 94%). Even after STR, only 50% of patients required a subsequent resection for recurrence. Addition of radiation therapy to initial STR did not seem to influence outcomes. At first relapse, GTR was accomplished in 1 patient, and only STR was accomplished in the others. Addition of radiation therapy to STR in our study led to disease control in half the patients treated, and STR alone led to disease control in only a quarter of the patients. Second relapses were treated palliatively with radiation therapy.

Conclusions: Surgical resection is the treatment of choice for CPPs. After initial STR, reoperations for recurrence are required only half the time. Therefore, there seems to be no role for radiation therapy after initial STR. For STRs at first relapse, local control outcome is poor.

Introduction

Choroid plexus papillomas (CPPs) are slow-growing, epithelial tumors of the choroid plexus that account for less than 1% of brain tumors in adults [1]. They usually arise within the ventricles. As a result of their deep intraventricular location, marked vascularity, and radiographic similarity to the normal choroid plexus, CPPs present diagnostic and therapeutic challenges. Many patients who have CPP present with signs of increased intracranial pressure and hydrocephalus. Tumoral calcification may be seen on plain radiographs, but the finding that usually prompts resection is a large, circumscribed, contrast-enhanced intraventricular tumor with associated hydrocephalus on computed tomography or magnetic resonance imaging (Figure 1). Ependymal or leptomeningeal spread is uncommon.

Total excision by surgical extirpation is expected to be curative, with infrequent recurrences [2]. However, a gross total resection (GTR) is often not feasible because of the deep intraventricular location of lateral ventricular tumors and the proximity of third and fourth ventricular tumors to critical brain-stem structures. Additionally, the role of adjuvant radiation therapy is unsettled [3–8]. We address these issues in our retrospective review of 41 patients with CPP who had operations at Mayo Clinic (Rochester, MN) between 1974 and 2000.

Materials and methods

From 1974 to 2000, 41 patients were seen at Mayo Clinic as part of their treatment evaluation for
CPP (17% were seen before 1980). The patients were identified through a search of the database of the Mayo Clinic Tissue Registry. The histologic diagnoses were confirmed in each case by a review of slides by one of the authors (BWS). Retrospective reviews indicated that each patient had undergone a full history and physical examination, routine blood tests, and radiographic studies as appropriate and available during the years encompassed by this review.

The study group included 19 male and 22 female patients (age range, 6 months to 74 years; mean, 36.6 years; median, 36 years). The distribution of tumor sites was as follows: fourth ventricle, 31 (76%); lateral ventricles, 7 (17%); and third ventricle, 3 (7%). Of the 7 lateral ventricular lesions, 6 (86%) were right-sided. Hydrocephalus was present at diagnosis in 14 patients (34%). Atypical histologic features (elevated mitotic and MIB-1 labeling indices; architectural complexity) were noted in 5 patients (12%).

For initial treatment, all patients underwent an operation, including GTR or subtotal resection (STR) and, where indicated, shunt placement (ventriculoatrial or ventriculoperitoneal). Ten patients underwent STR elsewhere and were subsequently referred to Mayo Clinic for evaluation for further treatment. Postoperative radiation therapy was administered to 4 patients who had undergone an initial STR. Radiation therapy was also combined with surgery at first relapse after STR in 2 patients and as palliative treatment at second relapse in 3 others, including 1 patient treated with combined modality therapy (chemoradiotherapy) and 1 treated by gamma knife. Three of the patients received external beam radiotherapy after 1985, and the others before 1985.

Follow-up time for all patients alive at last contact ranged from 4 months to 28 years (mean, 6.6 years; median, 6.5 years). All data regarding distant and local brain control were based on neuroimaging. Generally, patients who received a diagnosis and were followed up before 1985 were imaged with computed tomography; after 1985, with magnetic resonance imaging. Local control was defined as absence of tumor regrowth on radiographic imaging (computed tomography or magnetic resonance imaging). Distant brain control was defined as absence of tumor relapse in the brain at a site distant from the initial disease on radiographic imaging (computed tomography or magnetic resonance imaging). Actuarial curves for survival, brain control, and local control were calculated using Kaplan–Meier methods [9]. Tests of significance were based on the log-rank test [10]. Multivariate analysis used the proportional hazard model and the log-linear hazard function of Cox.