Choroid plexus tumor

Choroid plexus tumors are rare intraventricular papillary neoplasms derived from choroid plexus epithelium.

Epidemiology

Choroid Plexus Tumor Epidemiology.

Classification

Choroid plexus papilloma

Atypical choroid plexus papilloma

Choroid plexus carcinoma

They include three histologies, choroid plexus papilloma (WHO grade I), atypical choroid plexus papilloma (WHO grade II) and choroid plexus carcinoma (WHO grade III). Altogether, they account for 0.4-0.6% of all brain tumors [1) 2)].

Results support the role of aggresome as a novel prognostic molecular marker for pediatric choroid plexus tumors (CPTs) that was comparable to the molecular classification in segregating samples into two distinct subgroups, and to the pathological stratification in the prediction of patients' outcomes. Moreover, the proteogenomic signature of CPTs displayed altered protein homeostasis, manifested by enrichment in processes related to protein quality control [3)].

Choroid plexus metastases

see Choroid plexus metastases.

Diagnosis

On CT, choroid plexus tumors appear heterogeneous and isodense with calcifications and necrosis.

Case series

Amer et al. examined the presence of aggresomes in 42 patient-derived tumor tissues by immunohistochemistry and we identified their impact on patients' outcomes. We then investigated the proteogenomics signature associated with aggresomes using whole-genome DNA methylation and proteomic analysis to define their role in the pathogenesis of pediatric CPTs.

Aggresomes were detected in 64.2% of samples and were distributed among different pathological and molecular subgroups. The presence of aggresomes with different percentages was correlated with patients' outcomes. The ≥ 25% cutoff had the most significant impact on overall and event-free survival (p-value < 0.001) compared to the pathological and the molecular stratifications.

These results support the role of aggresome as a novel prognostic molecular marker for pediatric CPTs that was comparable to the molecular classification in segregating samples into two distinct subgroups, and to the pathological stratification in the prediction of patients' outcomes. Moreover, the proteogenomic signature of CPTs displayed altered protein homeostasis, manifested by enrichment in processes related to protein quality control [4].

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A total of 349 patients with CPTs were identified (120 CPCs, 26 aCPPs, and 203 CPPs). Patients with CPC presented at a younger age (median 3, mean 14.8 years) relative to CPP (median 25, mean 28.4 years; p < 0.0001). Histology was a significant predictor of OS, with 5-year OS rates of 90, 77, and 58 % for CPP, aCPP, and CPC, respectively. Older age and male sex were prognostic for worse OS and CSS for CPP. Only extent of surgery had a significant impact on survival for CPC. The use of adjuvant RT in patients with CPC undergoing surgery was not associated with a significantly improved OS (p = 0.17). For patients undergoing GTR without RT as part of an initial course of therapy, estimated 5- and 10-year OS were 70 % (±7 %) and 67 % (±8 %), respectively. Prospective data are required to define the optimal combination of surgery with adjuvant therapies, including chemotherapy [5].

Seventeen childhood patients were recorded with CPT. Cases were distributed so that 9 cases were choroid plexus-papilloma (CPP) (52.9%), 2 cases atypical CPP (11.7%) and 6 cases choroid plexus-carcinoma (CPC) (35.2%). Age at diagnosis was less than 2 years in 14 of the 17 patients (82.3%) and the incidence was higher in males (82.3% of the cases). Gross total resection was performed in 16 patients (94.1%). Adjuvant treatment was used in 6 patients (all this cases with CPC) (35.2%). Two of the 17 patients died (11.7%), showing an incidence density of 0.01 deaths/year.

The case series is consistent with previous published in scientific literature regarding epidemiology, tumor grade, clinical presentation, radiological features and therapeutic approach. Gross total resection is considered the therapeutic gold standard for choroid plexus tumors. Chemotherapy and radiotherapy should be used as adjuvant treatment in CPC and recurrent or remaining atypical CPP [6].

References

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