Chordoid meningioma

The term chordoid meningiomas was first used by Kepes et al. in 1987 to describe a meningeal tumor in young patients associated with microcytic anemia and/of dysgammaglobulinemia. Such tumors were composed of spindle or epithelioid cells disposed in chordoma-like clusters and cords in a myxoid matrix and often featured a prominent lymphoplasmacellular infiltrate.

Chordoid meningioma, WHO grade II, is an uncommon variant of meningioma with a propensity for aggressive behavior and increased likelihood of recurrence.

Classified as Grade II/atypical meningioma, is a rare subtype, which represents only 0.5% of all meningiomas.

Morphologically, it can mimic other chondromas and myxoid tumors within the brain and its vicinity thus posing a diagnostic challenge. Accurate diagnosis, therefore, assumes importance as these tumors have an aggressive clinical course and propensity to recur compared to classical meningiomas. Furthermore, the prognosis and treatment strategies vary when compared to tumors with morphological overlap.

Tumour cells in CM contain epithelial membrane antigen (EMA), D2-40, and focal S100 protein.

In a study, Immunohistochemistry was performed with antibodies against D2-40, S100, pankeratin, epithelial membrane antigen (EMA), brachyury, and glial fibrillary acidic protein (GFAP) in 4 cases of chordoid glioma, 6 skeletal myxoid chondrosarcomas, 10 chordoid meningiomas, 16 extraskeletal myxoid chondrosarcoma, 18 chordomas, 22 low-grade chondrosarcomas, and 27 enchondromas. Staining extent and intensity were evaluated semiquantitatively and mean values for each parameter were calculated. Immunostaining with D2-40 showed positivity in 100% of skeletal myxoid chondrosarcomas, 96% of enchondromas, 95% of low-grade chondrosarcomas, 80% of chordoid meningiomas, and 75% of chordoid gliomas. Staining with S100 demonstrated diffuse, strong positivity in all (100%) chordoid gliomas, skeletal myxoid chondrosarcomas, low-grade chondrosarcomas, and enchondromas, 94% of chordomas, and 81% of extraskeletal myxoid chondrosarcomas, with focal, moderate staining in 40% of chordoid meningiomas. Pankeratin highlighted 100% of chordoid gliomas and chordomas, 38% of extraskeletal myxoid chondrosarcomas, and 20% of chordoid meningiomas. EMA staining was positive in 100% of chordoid gliomas, 94% of chordomas, 90% of chordoid meningiomas, and 25% of extraskeletal myxoid chondrosarcomas. Brachyury was positive only in the chordomas (100%), whereas GFAP was positive only in the chordoid gliomas (100%). EMA was the most effective antibody for differentiating chordoid meningioma from skeletal myxoid chondrosarcoma, low-grade chondrosarcoma, and enchondroma, whereas D2-40 was the most effective antibody for differentiating chordoid meningioma from extraskeletal myxoid chondrosarcoma and chordoma. Our findings demonstrate that in conjunction with clinical and radiographic findings, immunohistochemical evaluation with a panel of D2-40, EMA, brachyury, and GFAP is most useful in distinguishing chordoid meningioma from chordoid glioma, skeletal myxoid chondrosarcoma, extraskeletal myxoid chondrosarcoma, chordoma, low-grade chondrosarcoma, and enchondroma. A lack of strong, diffuse S100 reactivity may also be useful in excluding chordoid meningioma. Among the neoplasms evaluated, brachyury and GFAP proved to be both sensitive and specific markers for chordoma and chordoid glioma, respectively. Of note, this study is the first to characterize the D2-40 immunoprofile in extraskeletal myxoid chondrosarcoma, results that could be of utility in differential diagnostic assessment [1)].

Systematic review

2016

The largest systematic review of CMs to date was published 2016. A comprehensive search on MEDLINE (OVID and Pubmed), Scopus, Embase, and Web of Science utilizing the search terms “chordoid” AND “meningioma” was performed to identify all reports of pathologically confirmed intracranial CMs. A total of 221 patients were included, comprising 120 females and 101 males. Mean age, MIB-1 index/Ki67, and tumor size was 45.5 years, 4.3 % (range 0.1-26.6 %), and 4.1 cm (range 0.8-10 cm), respectively. 5-, and 10- year progression free survival was 67.5 and 54.4 %, respectively. Gross total resection (GTR) and subtotal resection was achieved in 172 and 48 patients, respectively. Adjuvant radiotherapy (RT) was given to 30 patients. Multivariate analysis found GTR was strongly correlated with decreased recurrence rates (HR 0.04, p = <0.0001), while higher MIB-1 labeling index (≥5 vs <5 %) was associated with increased recurrence (HR 7.08; p = 0.016). Adjuvant RT, age, gender, and tumor location were not associated with recurrence. GTR resection is the strongest predictor of tumor control, and should be the goal to minimize local progression. Additionally, higher MIB-1 labeling was associated with increased rates of tumor recurrence. Tumors that are subtotally resected or demonstrate higher MIB-1 are at greater recurrence and warrant consideration for RT and close long term follow up [2].

Case series

Chordoid meningioma case series.

Case reports

Chordoid meningioma case reports.

1)
Sangoi AR, Dulai MS, Beck AH, Brat DJ, Vogel H. Distinguishing chordoid meningiomas from their histologic mimics: an immunohistochemical evaluation. Am J Surg Pathol. 2009 May;33(5):669-81. doi: 10.1097/PAS.0b013e318194c566. PubMed PMID: 19194275.
2)
Choy W, Ampie L, Lamano JB, Kesavabhotla K, Mao Q, Parsa AT, Bloch O. Predictors of recurrence in the management of chordoid meningioma. J Neurooncol. 2016 Jan;126(1):107-16. doi: 10.1007/s11060-015-1940-9. Epub 2015 Sep 26. PubMed PMID: 26409888; PubMed Central PMCID: PMC4684776.