# Chordoid meningioma case series

## 2018

A series reporting 33 cases is the third largest series in published literature from a single Institution. They reviewed Clinico-pathological characteristics of 33 patients diagnosed with chordoid meningioma between 2001 and 2015 in our institution. Forty-one specimens were available for review of histopathological and immunohistochemical characteristics. There were 15 men and 18 women with mean age of 36.8 years (median 36 years, range 9-62 years) at diagnosis with three cases occurring in pediatric age group. The majority were supratentorial in location with 11 convexity, 1 falcine, 5 parasagittal, 1 intraventricular, skull base involvement in 12 with 4 being petroclival location and 3 had spinal lesions. Lymphoplasmacytic infiltrates were seen in 23 cases with majority being T cells. MIB index varied from 1 to 14%. Five patients received radiotherapy for residual lesion. Two patients died (recurrence-1, post-operative complication-1). Three patients were lost to follow up after surgery. The mean post-operative follow up period for the remaining was 55.3 months. Seven patients had recurrence of which three had it twice. This study adds to the pool of available data for better understanding of this variant of meningioma. These meningiomas occur in middle age; spinal lesions and pediatric cases are not uncommon. We did not find any association between surgery, post-operative radiotherapy and histopathological features with recurrence and survival. Small number of cases may be responsible for this statistical insignificance. [1)].

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Zhang et al.,retrospectively reviewed clinical data from 111 cases of grade II meningiomas, including 55 cases of CM and 56 cases of CCM, between January 2011 and December 2015.

RESULTS: The mean follow-up time of the WHO grade II meningiomas (n=111) was 45.3 months. In the CM group, 8 patients (14.5%) experienced recurrence, and 2 patients (3.6%) died. In the CCM group, 22 patients (39.3%) experienced recurrence, and 9 patients (16.1%) died. Significant differences were observed between the CM and CCM groups in tumour size (P=0.019), history of surgery (P=0.038) and peritumoural oedema (P=0.004). In the CM group, gross total resection (GTR) was associated with favourable progression-free survival (PFS) (hazard ratio (HR)=0.144, 95% confidence interval (CI)=0.029-0.714; P=0.018). In the CCM group, univariate analyses revealed that a preoperative Karnofsky performance scale (KPS) score <80 (P<0.001), a tumour size ≥5 cm (P=0.015), tumour size (per-centimetre increase) (P=0.022), bone invasion (P=0.004), a history of surgery (P<0.001) and subtotal resection (STR) (P=0.009) were associated with worse PFS. The male gender (P=0.039), tumour size (per-centimetre increase) (P=0.043), bone invasion (P=0.030), and a history of surgery (P=0.007) were associated with poor overall survival (OS).

CONCLUSION: This study revealed that GTR should be achieved in grade II meningiomas. Patients with larger tumour sizes and/or surgical histories had worse outcomes [2)].

### 2010

The age range was from 30 to 67 years old (mean, 34.2 years). Seven patients were female and three male. The duration of symptoms varied from 3.5 months to 5 years (mean, 14.1 months). No systemic symptoms were noted. The tumor was localized in eight cases in the supratentorial compartments.

Histologically, the tumors were characterized by strands and cords of meningothelial cells arranged in a mucinous stroma. Two of the ten tumors showed metaplasic changes, and seven showed brain invasion. Tumor cells demonstrated CK7, EMA and focal S-100 protein and Ep-CAM. Cytokeratin AE1/AE3, GFAP and synaptophysin were negative. The MIB-1 proliferative index was from 6 to 9% (mean 7.8). PCNA Li was 6 to 20% (mean, 14), and microvascular density was 6-16 (mean, 14.5). The mean rate of the MIB-1 labeling index in recurrences was 7.1% versus 6.33% for no tumor recurrence. Chordoid meningioma, World Health Organization grade II, is an uncommon variant of meningioma with a propensity for aggressive behavior and increased likelihood of recurrence. Chordoid meningiomas are predominantly tumors of young adults with a predilection for the supratentorial location. Intraventricular location and absence of systemic manifestations, despite the presence of abundant B-lymphocytes, mast cells and low MIB-1 LI, are some of the interesting findings in the present series that need further study. Hence, a larger number of cases with adequate follow-up data need to be studied further to establish the clinical relevance of this variant [3)].

**2006**

The age ranged from 12-67 years (mean 34.2 years) and three of them occurred in < 18 years. Male to female ratio was 1:1.4. The duration of symptoms varied from 3.5 months to 5 years (mean 14.1 months). No systemic symptoms were noted. The location of tumor in eight cases was in the supratentorial and rest four in the infratentorial compartments. Interestingly, two cases were in intraventricular location, one in the lateral ventricle and other in the fourth ventricle. Microscopic examination showed lobulation with chordoid elements constituting > 95% of the entire tumor area in 11 of the total 12 cases. In one case, chordoid pattern constituted about 30% of the total tumor area; the rest was predominant meningothelial (60%). Mild to severe lymphoplasmacytic cell infiltrate was present in all cases. The histochemical stains showed the pattern of acidic mucin and interestingly revealed the presence of mast cells both in connective tissue stroma and epithelial cell islands. The inflammatory infiltrate was B-cell predominant. MIB-1 labeling index was low (< 2%) in all cases except two, which showed LI of 6% and 8%. Strong diffuse immunoreactivity for vimentin and focal positivity for epithelial membrane antigen was noted in all cases.

Chordoid meningiomas are predominantly tumors of young adults with predilection for supratentorial location. Intraventricular location, absence of systemic manifestations despite the presence of abundant B-lymphocytes, presence of mast cells and low MIB-1 LI are some of the interesting findings in the present series, which need documentation. Hence, larger number of cases with adequate follow-up data need to be studied further to establish the clinical significance of this variant [4)].

**2000**

A study includes 42 chordoid meningiomas that represented 0.5% of all meningiomas operated at Mayo Clinic during the interval 1975 to 1997. The male to female ratio was 1:1 and the age range was 12 to 77 years (mean, 47.4 yrs). Only two (5.2%) occurred in children. The majority (88%) were large and supratentorial. No manifestation of systemic disease was noted. Chordoid elements comprised 10% to 100% of the tumors: 34 (81%) were more than 50% chordoid. Thirty-seven tumors (88%) were classified as typical and five as atypical. Lymphoplasmacytic infiltrates varied, being moderate in 10 cases (23.8%), mild in 15 (35.7%), and absent in 17 (40.5%). In 14 (42%) of the 33 cases with available follow up, one or more recurrences were noted. All but one recurrent tumor had been subtotally resected. In 86% of recurrent tumors, the primary lesion was more than 50% chordoid in pattern and contained little or no inflammatory infiltrate. In Couce et al experience, chordoid

meningiomas are primarily tumors of adults, lack sex predilection, are unassociated with systemic manifestations, and uniformly recur when subtotally excised [5)]

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