Chordoid meningioma: A case report and review of the literature*

*Kordoid meningiom: Olgu sunumu ve literatürün gözden geçirilmesi**

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Özet

Amaç: Kordoid meningiom çok az olarak görülen bir meningiom varyantı olup, miksoid bir matriks içinde kordonlar ve trabekülalar oluşturan eozinofilik, vakuollü hücrelerden oluşmaktadır. Burada, kordoid meningiom tanısı alan bir olgu sunulmuş ve kordoid meningiomlarla ilgili kaynak bilgileri gözden geçirilmiştir.

Olgu Sunumu: 39 yaşındaki erkek hasta başağrısı ve giderek belirginleşen görme bozukluğu ile başvurdu. Bilgisayarlı tomografide sağ frontoparyetal bölgede yerleşim gösteren, durayla ilişkili, iyi sınırlı, 10 cm çapında tümöral kitle saptandı. Total olarak çıkarılan kitlenin mikroskopik incelemesinde, tümörün miksoid bir stroma içinde kordonlar ya da trabekülalar oluşturmuş eozinofilik sitoplazmalı, epitelioid görünümde hücrelerden meydana geldiği görüldü. İmmün dokukimyasal incelemede, tümör hücrelerinin EMA ve vimentin ile kuvvetli boyanma gösterdiği, buna karşın S-100 protein ve GFAP ile boyanmadığı görüldü. Olguda operasyondan sonraki 42 aylık dönemde rekürrens izlenmedi.

Sonuç: Kordoid meningiom az görülen bir meningiom variantıdır ve sık görülen diğer meningiom türlerinden daha olumsuz bir klinik gidiş gösterir..

Anahtar sözcükler: Kordoid meningioma, kordoma, prognoz

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Summary

Purpose: *Chordoid meningioma is a rare histologic variant of meningiomas and shows cords and trabeculae of eosinophilic, vacuolated cells in a myxoid matrix. In this paper, a case of chordoid meningioma is presented and the literature about chordoid meningiomas is reviewed.*

Case report: *A 39-year-old man was admitted to our hospital for evaluation of headache and gradual visual deterioration. The computed tomography scan demonstrated an extra-axial, well circumscribed, homogeneous enhancing mass lesion, 10 cm in diameter, adhering to the dura in the right frontoparietal region. The tumor was completely removed and histological examination of the tumor revealed cords or trabeculae of eosinophilic, vacuolated epithelioid cells in a myxoid matrix. A meningothelial pattern was noted focally. Immunohistochemical studies revealed that the tumor cells were strongly positive for EMA and vimentin, but negative for S-100 protein, cytokeratin and glial fibrillary acidic protein. Ki-67 was positive in a few tumor cells. The patient is alive with no recurrence after 42 months.*

Conclusion: *Chordoid meningioma is a rare morphologic variant of meningioma and is associated with a less favourable clinical outcome than ordinary meningiomas.*

Key words: *Chordoid meningioma, chordoma, prognosis*

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Meningiomas are generally slowly growing benign tumors derived from leptomeningeal arachnoid cells and account for approximately 15% of all primary tumors of central nervous system (1,2). Meningiomas have a wide range of histopathological appearances due to the capacity of arachnoid cells to exhibit divergent differentiation. The latest World Health Organization (WHO) classification of tumours of the central nervous system lists 15 histopathological variants of meningioma (Table I) (3). In contrast to the most common types of meningiomas, such as meningothelial, fibrous and transitional, the chordoid meningiomas are rare tumors, and in most instances, only solitary cases are reported. Literature review of this tumor is summarized in Table II (2,4-16). The majority of these reports suggest that chordoid meningiomas are frequently associated with hematological abnormalities or systemic manifestations of Castleman syndrome (2,5,9,10). However, recent reports of chordoid meningioma do not show any association with hematological abnormalities or systemic manifestations of Castleman syndrome (4,6,7,12-16).

We present herein a case of chordoid meningioma that occurred in a 39-year old man and discuss the clinico-pathologic features of such tumors.

Table I. Variants of meningioma.

Meningothelial
Fibrous (fibroblastic)
Transitional (mixed)
Psammomatous
Angiomatous
Microcystic
Secretory
Lymphoplasmacyte-rich
Metaplastic
Atypical
Clear cell
Chordoid
Rhabdoid
Papillary
Anaplastic (malignant)

Case report

A 39-year old man was admitted to our hospital for evaluation of headache and gradual visual deterioration. His medical and family history were unremarkable. Best corrected visual acuity was 10/200 in OD and 15/200 in OS at the presentation. Visuals fields were concentrically constricted. Funduscopic examination showed evidence of

Table II. Literature review of chordoid meningiomas.

Authors	No of cases	Age	Systemic effects (No of patients)	Resection (No of patients)	Postoperative course (No of patients)
Kepes et al.[9]	7	8-19	Anemia (7), Dg* (1)	Subtotal (2)	A&W† 5 yrs (1) Recurrence 2 yrs (1)
				Total	A&W 6 mos-3 yrs (4) Recurrence 20 mos (1)
Glassier et al.[1]	1	15	No	Total	Not known
Zuppan et al.[15]	1	10	No	Total	A&W 1 yr
Kumar et al.[11]	1	5	No	Total	Recurrence, DOD‡ 6 mos
Civit et al.[5]	1	22	Anemia	Total	A&W 3 mos
Kobata et al.[10]	1	15	Anemia	Total	A&W 2 yrs
Kajiwara et al.[8]	1	52	No	Total	A&W 2 yrs
Couce et al.[6]	42	12-77	No	Total (29)	A&W 1 mo-12 yrs (20) Recurrence 6 yrs (1) DOD 6 days, 1 mo (2) LFU§ (6)
				Subtotal (13)	Recurrence 1-16 yrs (13)
Yano et al.[13]	1	44	No	Total	Not known
Lee et al.[2]	1	55	Dg	Total	A&W 6 mos
Mori et al.[12]	1	62	No	Total	A&W 40 mos
Ayadi-Kaddour et al.[4]	1	47	No	Total	A&W 8 mos
Yeon et al.[14]	1	33	No	Total	A&W 7 mos
De Tella Jr et al.[7]	2	19	No	Total (1)	DOD 7 mos
		52	No	Partial (1)	A&W 3 yrs
Varma et al.[16]	2	20	No	Total (1)	A&W 1 yr
		33	No	Total (1)	Not known

* Dg, dysgammaglobulinemia; †A&W, alive and well; ‡DOD, dead of disease; §LFU, lost to follow up;

bilateral papilledema. Neurological examination revealed minimal left hemiparesis

Laboratory findings included a hematocrit level of 43.1% (reference range 42.6%-52.6%), hemoglobin count of 14.3 g/dl (reference range, 13.5-17.5 gr/dl), mean corpuscular volume of 87.1 fl (reference range, 82-98 fl), mean corpuscular hemoglobin of 29.5 pg (reference range 27-34 pg), mean corpuscular hemoglobin concentration of 34.7 g/dl (reference range 31-36 gr/dl). The patient's serum immunoglobulin level was normal.

The T1-weighted postgadolinium image demonstrates an extra-axial, well circumscribed, homogeneous enhancing mass lesion, 10 cm in diameter, adhering to the dura in the right frontoparietal region. The tumor was isointense on both T1 and T2-weighted images.

The tumor was completely excised via a right frontoparietal craniotomy. Intraoperatively, the tumor was found to be well circumscribed, gray-purple colored, rich in vascularity with a dural attachment. There was no gross invasion to brain tissue. The postoperative period was uneventful and the patient was discharged on the 8th postoperative day. The patient remains free of disease after 42 months of follow-up, however, during this period, progressive visual deterioration resulted in loss of visual function. Funduscopic examination showed a secondary optic atrophy.

Pathological Findings

The gross specimen consisted of tumoral fragments of variable size, ranging from 0.5x0.5x0.5 cm to 6x5x5 cm. The tumor was rather soft and moderately vascularized. The cut surface was grayish-tan and somewhat myxoid. On histologic examination, the larger portion of the tumor (more than 95% of the tumor) was composed of cords or trabeculae of eosinophilic, vacuolated epithelioid cells in a myxoid background (Fig 1). A meningothelial pattern was noted focally. There were no cellular atypia, mitotic figures or necrotic foci. Mild lymphoplasmacytic infiltrate was focally present. Masson-trichrome stain demonstrated a collagen deposition between the cords of tumor cells. Immunohistochemical studies revealed that the tumor cells were strongly positive for EMA (Fig 2) and vimentin, but negative for S-100 protein, cytokeratin and glial fibrillary acidic protein. Ki-67 was positive in a few tumor cells (less than 1% of tumor cells). On the basis of these histopathologic and immunohistochemical findings, a diagnosis of chordoid meningioma was established.

Discussion

The term "chordoid meningioma" was first used by Kepes et al.(9), in 1988, to describe a meningeal tumor displaying histological characteristics closely imitating those of chordoma and that arise in young patients who had iron-refractory hypochromic microcytic anemia and/or dysgammaglobulinemia. The first known case with similar clinical and histopathological features, was reported by Connors (17) in 1980 and later reviewed by Dimand et al. (18) in 1985. Such tumors were composed of spindle or epithelioid, partly vacuolated cells assuming a chordoma-like pattern with nests and cords in a myxoid matrix. In addition, peritumoral reactive lymphoplasmacellular infiltrates often showing a number of follicles with well-formed germinal centers were a regular features. Kepes et al. (9) suggested that the systemic effects occurring in cases of chordoid meningiomas, may be related to the lymphoplasmacellular response induced by some chemical or antigenic stimulants within the tumor. In these cases, surgical resection of the tumor results in normalization of the blood picture and improvement of the systemic disease.

The largest series of chordoid meningiomas was reported by Couce et al. (6) in 2000. Their study included 42 chordoid meningiomas representing 0.5% of all meningiomas operated in Mayo Clinic from 1975 to 1997. In this study, the clinical features of the patients were found to be significantly different from those observed in previous studies. The patient age ranged from 12 to 77 years (mean, 44 yrs) and only two tumors (5.2%) occurred in children. In contrast to previous reports, no significant association with systemic or hematologic abnormalities were found. The authors suggested that the reported associations are limited to the childhood period. Similarly, Kepes et al. (9) stated that younger individuals seem to be more susceptible to systemic effects of the lymphoplasmacellular infiltrate. The clinical features in our case are concordant with those observed by Couce et al. (6). The patient age was 39 and no systemic and hematologic abnormalities were observed.

Histologically, chordoid meningiomas are characterized by the presence of cords and nests of epithelioid or spindle cells in a myxoid matrix. Chordoid areas may occupy the whole tumor or are interspersed with typical regions of meningioma. Lymphoplasmacellular infiltrates are present in the majority of tumors with varying intensity, however, unlike what was previously thought, they are not a requisite for the diagnosis (6).

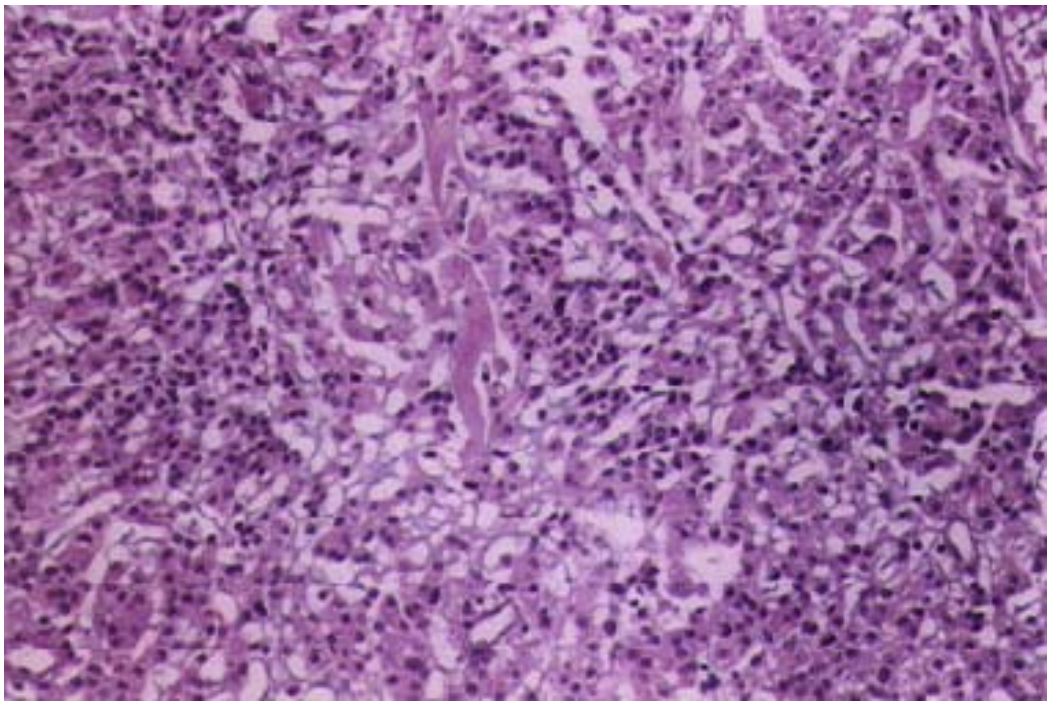

Figure 1. Larger portion of the tumor consists of cords of eosinophilic, vacuolated epithelioid cells in a myxoid background (H+E stain; original magnification X100).

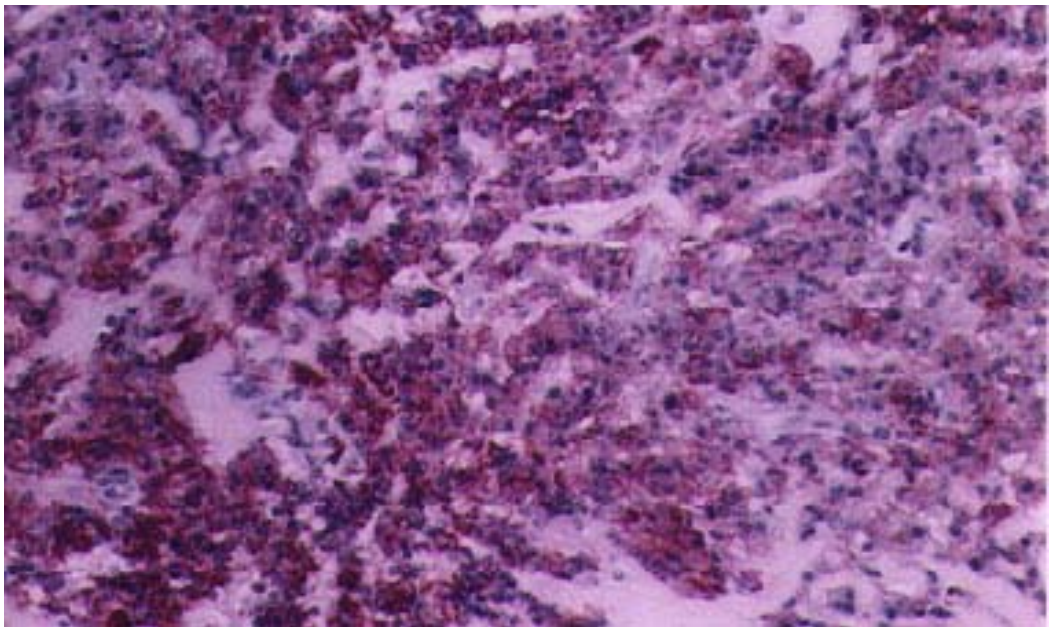

Figure 2. Tumor cells are diffusely and strongly positive for EMA (original magnification X200).

The differential diagnosis of chordoid meningiomas includes chordoma, extraskeletal myxoid chondrosarcoma, chordoid glioma, and metastatic mucinous carcinoma. Immunohistochemical features along with the location of the tumor are helpful in the differential diagnosis between chordoid meningiomas and chordomas. In chordoid meningiomas, tumor cells are strongly positive for EMA and vimentin, but negative for S-100 protein and cytokeratin. In contrast, the latter two antibodies are characteristically expressed in chordomas. Additionally, chordomas usually locate in the midline structures such as the clivus or sellar area. Extraskeletal myxoid chondrosarcomas are rare and have not been described in the central nervous system. In contrast to chordoid meningioma, they are positive for S-100 protein (19). Chordoid glioma is a recently described entity consisting of cords and clusters of epithelioid cells with abundant eosinophilic cytoplasm in a mucoid matrix, simulating chordoid meningioma. In contrast to chordoid meningioma, the tumor cells in chordoid glioma are strongly glial fibrillary acidic protein-positive (20). Metastatic mucinous carcinoma differs from chordoid meningioma by its strong and uniform Cam 5.2 positivity and by the presence of intracellular mucin in tumor cells (6). In our case, the presence of focal areas of meningothelial pattern, positive immunostaining for vimentin and EMA, and negative immunostaining for S-100 protein, cytokeratin and glial fibrillary acidic protein facilitated the differential diagnosis.

The major clinical parameter for predicting recurrence in chordoid meningiomas is the extent of resection (6), like in other meningioma variants (21). Chordoid meningiomas exhibit a high rate of recurrence following subtotal resection. In the series of Couce et al. (6), one or more recurrences were noted in 14 (42%) of the 33 cases with available follow up, and of those, 13 (92%) had been subtotally resected. The tumor grade is the most important histological parameter to determine the risk of recurrence in meningiomas (22). Although most meningiomas are benign and are graded into WHO grade I, the chordoid variant is associated with a less favourable clinical outcome and is graded into WHO grade II. The extent of chordoid pattern is also important in predicting prognosis. Couce et al. (6) observed that in the majority (85.7%) of recurred cases, the primary tumors show chordoid pattern in more than 50% of the tumor tissue. Proliferation indices have also been used to predict recurrence in meningiomas. Ki-67 proliferation index show a highly significant increase from benign, to atypical, and anaplastic meningioma (23). In our case, in spite of the extensive chordoid pattern, no recurrence was observed after the follow-up of 42 months. This may be explained by the total excision of the tumor and low Ki-67 labeling index.

In conclusion, chordoid meningioma is a rare morphologic variant of meningioma and may not be associated with systemic or hematologic abnormalities. The potential histopathologic misdiagnosis can be avoided by establishment of histologic features, supported by positive immunostaining for EMA and vimentin.

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