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CLINICAL STUDY – PATIENT STUDY

Chordoid meningioma: a clinicopathologic study of 11 cases at a single institution

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Abstract Chordoid meningioma is an uncommon variant of meningioma, which histologically bears a great resemblance to chordoma and often follows an aggressive clinical course. We examine clinicopathologic features of 11 cases of this rare tumor to further elucidate its behavior. Thirteen specimens of chordoid meningioma belonging to 11 patients were obtained at a single institution from 1995 to 2009. Correlations of histologic parameters, immunohistochemical study, and clinical features were assessed. This series included six men and five women with a mean age of 60.8 years at first surgery. Aside from one patient (case 5) who died of disease immediately after the first operation, the mean postoperative follow-up period for the other 10 patients was 41.4 months. Two patients each had a local tumor recurrence. The mean time to recurrence was 10.4 years. No systemic manifestations of Castleman syndrome, such as iron-refractory hypochromic/microcytic anemia and dysgammaglobulinemia, were found. Six tumors (46%) were classified as benign (grade I) and seven tumors (54%) atypical (grade II), if based solely on histologic grading irrespective of chordoid or clear cell components in our cases. Lymphoplasmacytic infiltrate was moderate in one tumor (7%), mild in eight tumors (62%), and absent in four tumors (31%). The inflammatory cells were predominantly T cells (CD3+), with only scarce B cells (CD20+). There was a wide range of MIB-1 labeling indices (0.3–25.8%, mean 7.5%), which increased following tumor recurrence. Our study demonstrates that chordoid meningiomas are not always associated with Castleman's Syndrome, and that this histologic category can be seen in the elderly as opposed to only in younger age groups.

Keywords Chordoid meningioma · Cords · Cribriforms · Nests · Immunohistochemistry

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Introduction

Meningiomas are common intracranial tumors that originate from the arachnoidal cap cell of the meninges and have a large variety of histopathologic appearances. Most of the variants are merely descriptive and carry no prognostic significance. However, some subtypes, such as chordoid and clear cell, have unique clinical associations or prognostic implications. Chordoid meningioma (CM) was classified separately for the first time in the 1993 World Health Organization (WHO) classification of tumors of the central nervous system (CNS) [1]. Its acceptance as a variant of meningioma came following its initial description by Kepes et al. [2]. Their observations of its occurrence in young individuals, chordoma-like histologic appearance, peritumoral company of lymphoplasmacellular infiltrates, and association with systemic manifestations of hematologic conditions, including iron-refractory hypochromic/microcytic anemia and bone marrow plasmacytosis with dysgammaglobulinemia (Castleman syndrome), inspired reporting of similar findings [3–23]. The published articles to date were largely in the form of isolated case reports, with only four series [2–5]. In the 2007 revision of

WHO classification of tumors of CNS, CM joined clear cell meningioma and atypical meningioma in a grade II category [24], due to a high rate of recurrence, particularly following subtotal resection.

Histopathologically, CM is composed of epithelioid or spindle cells, forming cords or nests, in a pale, basophilic mucoid matrix. The tumor cells bear great resemblance to physaliferous cells of chordoma, featuring vacuolated cytoplasm. Chordoid morphology often dominates the primary tumors, and the proportion of chordoid elements in pathological specimens almost always increases with recurrences. Pure CM cases from the outset are not infrequent [3, 4].

The present study was undertaken to document the clinicopathologic correlation of 11 cases of this uncommon tumor, operated at the Chang Gung Memorial Hospital–Kaohsiung Medical Center during 1995–2009.

Materials and methods

All the cases shown in this study represented materials from the surgical pathology files of the Chang Gung Memorial Hospital–Kaohsiung Medical Center from 1995 to 2009. Clinical data, which included age, sex, presenting symptoms, tumor size, location of involvement, treatment, and clinical follow-up, were obtained from the medical records. Surgical specimens were promptly fixed in neutral buffered formalin, paraffin-embedded, and routinely processed. Routine hematoxylin-eosin-stained sections were generated from formalin-fixed, paraffin-embedded tissue, which was cut to 3 μm thick. Special stains for periodic acid-Schiff (PAS) with and without diastase, mucicarmine, and alcian blue at pH 2.5 were applied to all cases.

Immunohistochemical stains were performed using standard reagents and techniques on an i6000 Automated Staining System (BioGenex, San Ramon, CA). The following antibodies were used for immunohistochemistry: epithelial membrane antigen (EMA) (clone GP1.4, Novocastra; steam in citrate buffer, 1:400), cytokeratin high molecular weight (HMW) (clone 34βE12, Neomarkers; steam in citrate buffer, 1:100), cytokeratin low molecular weight (LMW) (clone AE1, Neomarkers; steam in citrate buffer, 1:100), podoplanin (Santa Cruz; steam in citrate buffer, 1:50), D2-40 (DAKO; steam in citrate buffer, 1:100), vimentin (clone V9, Neomarkers; steam in citrate buffer, 1:400), CD10 (clone 56C6, Novocastra; steam in citrate buffer, 1:100), glial fibrillary acidic protein (GFAP) (clone ZCG29, Invitrogen; steam in citrate buffer, 1:150), S-100 (polyclonal, DAKO; 1:1,000), MIB-1 antigen (clone MIB-1, DAKO; steam in citrate buffer, 1:100), Leukocyte common antigen (T200) (2B11 + PD7/26, Dako; steam in citrate buffer, 1:200), CD3 (clone PS1, Novocastra; steam in citrate buffer, 1:300), CD20 (clone 1FB, DAKO; steam in citrate buffer, 1:400), and CD68 (clone KP1, Novocastra; steam in citrate buffer, 1:100). Inactivation of endogenous peroxidase activity was obtained by incubating sections in 3% H_2O_2 for 15 min. Localization of bound antibodies was performed with a non-biotin polymeric technology (Super Sensitive^{TM} Polymer-HRP Detection System; BioGenex). Immunoreactions were visualized using 3,3′-diaminobenzidine tetrahydrochloride (ZYMED®; Invitrogen, Carlsbad, CA). Appropriate positive controls for each antibody were run in parallel. The immunostained slides were then assessed for both extent and intensity of staining. The percentage of tumor with positive staining was estimated first. Next, the intensity of staining was semiquantitatively scored from 0 to +++. The rough percentage of tumor with positive staining and the semiquantitative measure of staining intensity were then tabulated for each specimen (see Table 3, below).

In addition to the mitotic index, cell proliferation was assessed by immunohistochemical staining for MIB-1 antigen. The MIB-1 labeling index (MI) was calculated in regions of maximal activity and expressed as percent nuclear area of staining. Based on the 2007 revision of new WHO classification of tumors of CNS [24], histologic features of atypia include a mitotic index ≥4/10 high-power fields (HPF) or the presence of at least three of the following variables: increased cellularity, small cell with a high nuclear: cytoplasmic ratio, prominent nucleoli, uninterrupted patternless or sheet-like growth, and foci of spontaneous or geographic necrosis. The mitotic index was defined as the maximal number of mitoses observed in any 10 consecutive HPF (1 HPF = 0.16 mm^2). Histologic malignancy was defined by the presence of obviously malignant cytology or an exceptionally high mitotic index (≥20/10 HPF).

Results

Clinical features

The essential clinical information of all 11 cases is summarized in Table 1. This series included six men and five women with a mean age of 60.8 years (range 43–77 years) at first surgery. There was no pediatric case. These tumors were distributed in sites where common meningiomas normally occur, including one at convexity, eight at parasagittal area, one at skull base, and one at cerebello-pontine angle. The tumors ranged from 2.0 to 8.0 cm in diameter, and the mean diameter was 5.5 cm. The common initial symptoms included headache, hemiparesis, conscious change, aphasia, and facial numbness. No systemic manifestations of Castleman syndrome, such as iron-refractory hypochromic/microcytic anemia and dysgammaglobulinemia, were found.

Table 1 Summary of clinical data

Case no.	Age (years)/sex	Tumor size (cm)/location	Extent of surgery	Clinical outcome
1	68/M	8.0/parasagittal	STR	Alive, AWT, 9 mos[a]
2	44/F	7.0/parasagittal	TR	Alive, ANT, RD (14 yrs), 14.9 yrs[a]
3	43/M	8.0/parasagittal	TR	Alive, AWT, RD (6.8 yrs), 8.7 yrs[a]
4	79/M	6.1/parasagittal	TR	Alive, AWT, 3.2 yrs[a]
5	46/F	NK/parasagittal	TR	Died of disease immediately after operation
6	77/M	6.0/parasagittal	TR	Alive, ANT, 1.8 yrs[a]
7	76/F	2.2/C–P angle	TR	Died of aspiration pneumonia, 8 mos[a]
8	57/F	7.5/parasagittal	TR	Alive, ANT, 7 mos[a]
9	53/M	2.0/parasagittal	TR	Alive, ANT, 3.5 yrs[a]
10	71/F	5.0/convexity	TR	Alive, ANT, 3 mos[a]
11	55/M	3.0/sella turcica	TR	Alive, ANT, 1 mos[a]

F Female, *M* male, *C–P* cerebello-pontine, *NK* not known, *TR* total resection, *STR* subtotal resection, *AWT* alive with tumor, *ANT* alive without tumor, *RD* recurrent disease, *yr(s)* year(s), *mo(s)* month(s)

[a] Time from first surgery at our hospital to last visit

Follow-up information was available for all patients. One patient (case 5) died of disease immediately after the first operation. For the other 10 patients, the mean postoperative follow-up period was 41.4 months (range 1–207 months). Nine patients had gross total resection at the first surgery. Among them, 2 patients (cases 2 and 3) each experienced a tumor recurrence. The mean time to recurrence was 10.4 years. The pattern of recurrence was local recurrence without CSF seeding or extracranial metastasis. They were both re-operated on; one (case 2) received gross total resection and was alive without tumor, the other (case 3) received subtotal resection and was alive with tumor. Two patients (cases 1 and 4) underwent subtotal resection at the first surgery and were alive with tumor. The three tumors (from cases 1, 3 and 4) partially removed with subtotal resection were all located at parasagittal area involving posterior aspect of the superior sagittal sinus. One patient (case 7) died of aspiration pneumonia 8 months after surgery. Among the nine patients still alive, six were free of disease, and three were alive with tumor. No other therapies, such as radiation and chemotherapy, were given as further treatment, even for the two patients (cases 1 and 4) undergoing subtotal resection at the first surgery and one patient (case 3) receiving subtotal resection at the second surgery.

Light microscopy and immunohistochemistry

There were in all 13 primary and recurrent intracranial CM specimens. The diagnostic parameters, histological grades and percentage of chordoid morphology are summarized in Table 2. The immunoprofile is summarized in Table 3. The patterns of chordoid morphology and clear cell characteristics, and immunohistochemical findings are illustrated in Figs. 1, 2 and 3. On microscopic examination, the tumors were composed of epithelioid cells (8/13) or plump to spindle cells (5/13), forming cords, cribriforms or nests, in a mucoid matrix. The chordoid elements constituted 30–98% of the tumor areas, with 9 of 13 (69%) tumors where over 50% of the components belonged to chordoid patterns. Sweeping, uninterrupted sheeting appearance was appreciated in four specimens where chordoid patterns were over 95% of the tumor areas; otherwise, they were alternating or intermingled with other subtypes (second subtype). The other meningioma subtypes included were three clear cell, three transitional, two metaplastic (lipomatous and xanthomatous, respectively), two meningothelial, two fibrous, and one psammomatous. Through thorough sampling of the specimens, we were able to identify typical meningothelial differentiation evidenced by whorl formation or intranuclear inclusions in all of them. Four specimens exhibited brain invasion while two exhibited skull invasion. Six tumors (46%) were classified as benign (grade I) and seven tumors (54%) atypical (grade II), if based solely on histologic grading irrespective of chordoid or clear cell components in our cases. One of the two recurrent tumors was designated as grade I, and the other was grade II. The mucoid matrix stained red with mucicarmine, pink with PAS, which was resistant to diastase, and bright with alcian blue at pH 2.5, consistent with acid mucin. Interestingly for the clear cell elements in three of the specimens, the cytoplasmic clearing was PAS-positive yet diastase-sensitive, consistent with glycogen accumulation. Lymphoplasmacytic infiltrate was moderate in one tumor (7%), mild in eight tumors (62%), and absent in four tumors (31%). The inflammatory cells were predominantly T cells (CD3+), with only scarce B cells (CD20+). There was a wide range of MIB-1 labeling indices (0.3–25.8%, mean 7.5%), which increased following tumor recurrence. All tumors showed diffuse positive

Table 2 Summary of the histopathologic findings

Case no.	Histologic grade	Increased cellularity	Small cell with a high n/c ratio	Prominent nucleoli	Sheet-like growth	Geographic necrosis
1	II	Yes	Yes	No	Yes	Yes
2	I	No	No	No	Yes	No
2[a]	I	No	No	No	Yes	No
3	II	Yes	Yes	No	No	Yes
3[a]	II	Yes	Yes	Yes	Yes	Yes
4	II	Yes	Yes	Yes	No	Yes
5	I	No	No	No	No	No
6	II	Yes	Yes	Yes	No	Yes
7	I	No	No	No	No	No
8	II	Yes	Yes	No	No	Yes
9	I	No	No	No	No	No
10	II	Yes	Yes	Yes	No	Yes
11	I	No	No	No	No	No

Case no.	Mitotic index (10 HPF)	Malignant cytology	MIB-1 labeling index (%)	Percentage of chordoid morphology	Additional morphology	Lymphocyte infiltration
1	8	Absent	11.5	96	Meningothelial, BI	Mild
2	0	Absent	0.6	97	Fibrous	Minimal
2[a]	1	Absent	2.1	97	Fibrous, SI	Mild
3	7	Absent	5.6	60	Transitional	Absent
3[a]	15	Absent	25.8	98	Transitional, BI	Absent
4	7	Absent	12.5	55	Clear cell	Mild
5	1	Absent	1.4	40	Meningothelial, SI	Mild
6	12	Absent	19.7	30	Clear cell	Mild
7	0	Absent	0.3	35	Fibrous	Absent
8	5	Absent	5.7	30	Metaplastic, BI	Moderate
9	2	Absent	2.7	60	Metaplastic	Minimal
10	6	Absent	6.8	55	Clear cell, BI	Minimal
11	1	Absent	2.8	60	Transitional	Mild

BI Brain invasion; *SI* skull invasion

[a] First recurrence

immunoreactivity with vimentin (100%) and EMA (100%), 3 tumors (23%) were positive for HMW, 3 tumors (23%) were positive for LMW, 11 tumors (85%) were positive for podoplanin (both membranous and cytoplasmic), and 11 tumors (85%) were positive for D2-40 (both membranous and cytoplasmic). Two tumors (15%) were positive for CD10, two tumors (15%) were positive for GFAP, and four tumors (31%) were positive for S100 (both nuclear and cytoplasmic).

Discussion

Meningiomas are common tumors, which constitute 13–20% of all primary intracranial tumors [25, 26]. The majority are slow growing and follow a benign clinical course. There are, however, rare meningiomas that meet atypical or anaplastic criteria, or belong to chordoid, clear cell, papillary or rhabdoid subtypes, and show an aggressive clinical behavior. Chordoid meningioma comprises 0.5–1.0% of meningioma [3–5]. To the best of our knowledge, 108 cases of CM have been published in the English literature [2–23], largely in the form of isolated case reports, with four series [2–5]. Although it was originally deemed a tumor with a preference for young individuals, the total number of reported patients under the age of 18 years is only 18 [5, 19]. Along with clear cell meningioma and atypical meningioma, CM is currently considered a grade II meningioma [24, 27], due to a high rate of recurrence, particularly following subtotal resection.

Kepes et al. [2] published the first series of CM in 1988 with emphasis on chordoma-like histology, young age,

Table 3 Immunohistochemical results—the extent of immunohistochemical staining is expressed semiquantitatively with the first number representing the percentage of tumor cells with observable staining and the second number representing the intensity of staining (+, ++, or +++)

Case no.	Antibody								
	EMA	HMW	LMW	Podoplanin	D2-40	Vimentin	CD10	GFAP	S100
1	100%, +++	0	0	10%,+	10%,+	100%, +++	0	0	0
2	100%, +++	0	0	0	0	100%, +++	0	0	0
2[a]	100%, +++	0	0	0	0	100%, +++	0	0	0
3	100%, ++	30%,+	40%,+	100%, ++	100, ++	100%, ++	0	0	40%,+
3[a]	100%, +++	70%,+	70%, ++	100%, +++	100, ++	100%, ++	0	0	60%, ++
4	100%, +++	5%, +++	10%, ++	70%, ++	60%, ++	100%, +++	0	0	0
5	70%, ++	0	0	80%, ++	70%, ++	90%, ++	0	10%, ++	10%, ++
6	100%, ++	0	0	80%, +++	60%, ++	100%, +++	0	5%,+	0
7	100%, ++	0	0	5%,+	5%,+	100%, ++	0	0	0
8	70%, ++	0	0	60%, ++	40%,+	100%, ++	5%,+	0	0
9	80%, ++	0	0	70%, ++	60%, ++	100%, ++	0	0	10%,+
10	80%, +++	0	0	70%, ++	60%, ++	70%, ++	10%, ++	0	0
11	100%, ++	0	0	80%,+	80%,+	100%, ++	0	0	0

[a] First recurrence

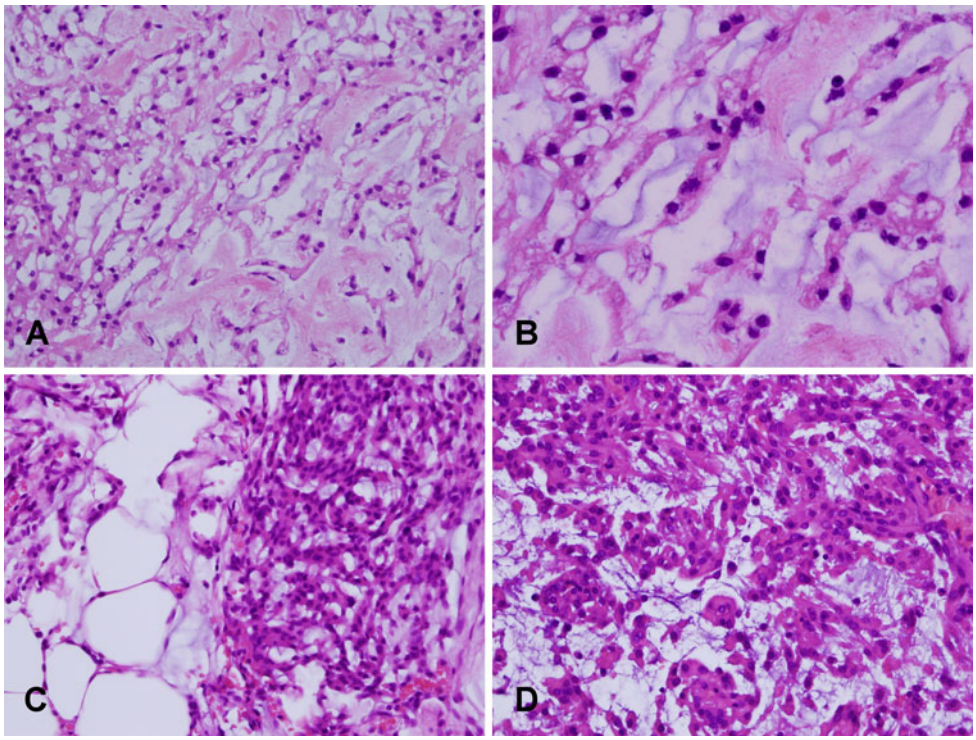

Fig. 1 **a** Epithelioid cells arranged in relatively irregular cords against mucoid matrix (case 1, H&E, ×200), **b** physaliferouslike cells with vacuolated cytoplasm (case 1, H&E, ×400), **c** epithelioid cells arranged in cribriforms against mucoid matrix (*right*), along with lipomatous element (*left*) (case 9, H&E, ×200), **d** epithelioid cells arranged in nests against mucoid matrix (case 5, H&E, ×200)

prominent peritumoral lymphoplasmacellular infiltrates, and association with systemic manifestations of Castleman syndrome. Subsequent articles demonstrated that CM was more often seen in adults than children or adolescents and not always associated with Castleman's syndrome. In fact, after the series of Kepes et al. [2], there were only a handful of CM case reports connected to systemic inflammatory syndrome [8, 14, 15]. And, as more and more people presented their studies, there was debate about whether it was B cell [4, 6, 14] or T cell [3, 15] committed to the peritumoral lymphoplasmacytic infiltrates [3, 4, 14, 15]. The majority of lymphocyte infiltrates in all meningiomas consisted of T cells [28], except for CMs of childhood [3, 29] and lymphoplasmacytoid meningiomas

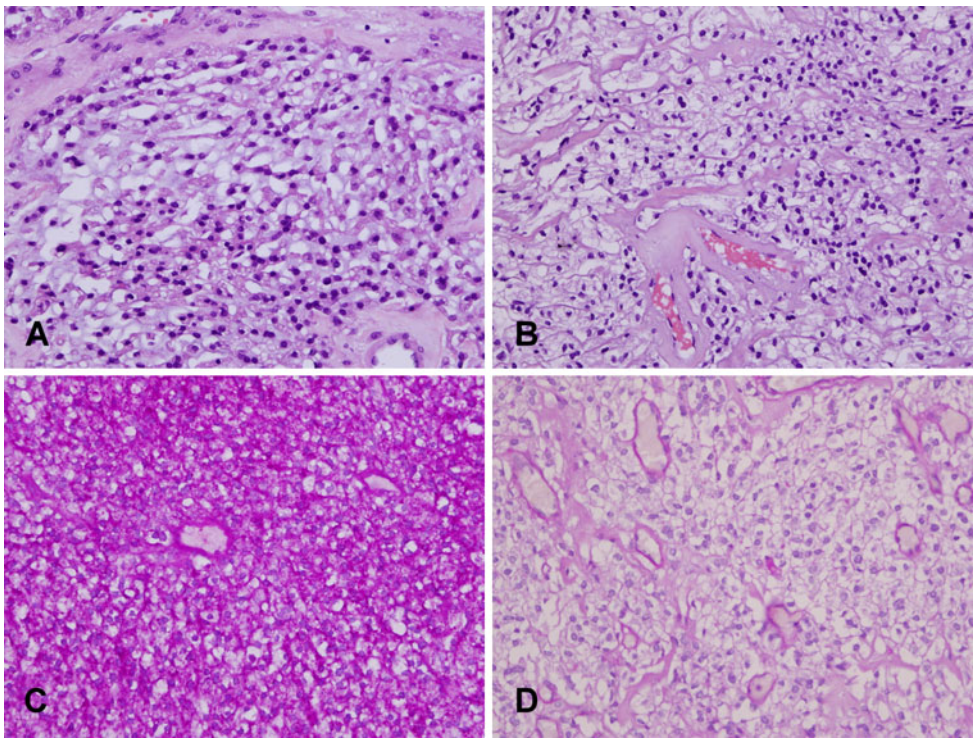

Fig. 2 (Case 10) **a** Epithelioid cells arranged in relatively regular cords against mucoid matrix (H&E, ×200), **b** sheets of clear cells divided by fibrous septa (H&E, ×200), **c** glycogen-rich cytoplasm (PAS, ×200), **d** absence of glycogen after diastase digestion (PAS-D, ×200)

that were strongly associated with B lymphocytes and plasma cells [6]. CM in young patients [3, 29] was mainly formed by B cells, due to association with Castleman syndrome [2], whereas a predominance of T cells was observed in adult cases [15]. In our cases, the inflammatory cells were predominantly T cells (CD3+), with only scarce B cells (CD20+), a result not very different from other subtypes of meningioma. Since all our cases are adults, why B cell infiltrates typify CMs of childhood and lymphoplasmacytoid meningiomas, but are lacking in CMs of adulthood, is open to speculation [15]. It remains a mystery surrounding pediatric CM cases with B cell infiltrates and Castleman syndrome noted in original descriptions of this entity.

Kepes et al. [2] reported a recurrence rate of about 28.6% (2/7). Counce et al. [3] observed a wide range of MIB-1 LI (0.4–11.4%; mean 5.2%) in their series and also reported a high rate of recurrence (39%). In contrast, Epari et al. [4] reported a low MIB-1 labeling index (<2% in all cases except two which showed 6 and 8%, respectively) and none of their cases had recurrence. Our cases also showed a wide range of MIB-1 LI (0.3–25.8%, mean 7.5%), which increased following tumor recurrence. Two patients respectively experienced recurrences at 6.8 and 14 years postoperatively (mean 10.4 years) even though they underwent a gross total resection of the primary tumors. The recurrence rate was 20% (excluding the patient who died immediately after first operation). Since mean time to recurrence was 10.4 years, and average follow-up was 41.4 months (range from 1 to 207 months), the vast majority of patients were followed up for a relatively short period of time. The follow-up on the recurrent cases was much higher (8.7 and 14.9 years) than the non-recurrent cases (range from 1 month to 3.2 years). If long-term follow-up could eventually be achieved for the non-recurrent cases, the recurrence rate might be higher. We also quantitated the percentage of chordoid architecture, which was previously addressed by Counce et al. [3]. They found that all recurrent tumors in their series were associated with a predominance of the mucin-rich, chordoid pattern [3]. In our study, there were nine specimens where the percentage of chordoid components was more than 50%, including two primary tumors of the patients with recurrences.

The presence of a WHO grade II meningioma should guide clinicians down a different management path, possibly resulting in closer follow-up and a lower threshold for re-operation in the event of recurrence. Surgery is the treatment of choice for meningiomas, and gross total resection should always be the goal of surgery. However, some meningiomas, particularly those involving the cavernous sinus, petroclival region, posterior aspect of the superior sagittal sinus, or optic nerve sheath, cannot be completely removed due to their relationship to vital neural or vascular structures [30]. The recommendation for such circumstances is surgery plus postoperative radiotherapy, especially when dealing with WHO grade II or grade III meningiomas. In the event of subtotally resected CMs, postoperative radiotherapy is the standard treatment.

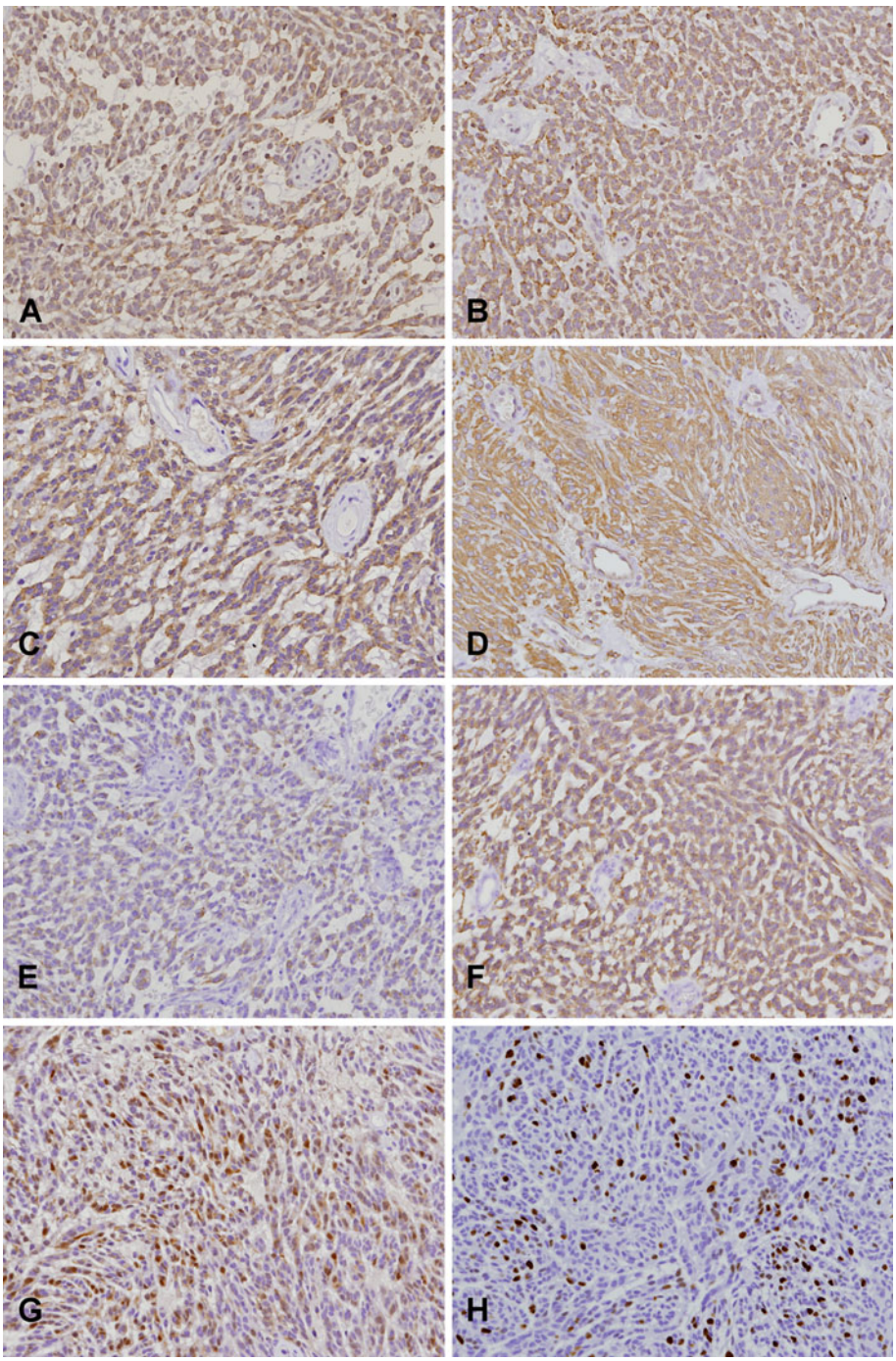

Fig. 3 (Case 3) Immunohistochemical findings. The tumor cells show membranous and cytoplasmic positivity for **a** D2-40 (×200), **b** podoplanin (×200), **c** membranous positivity for EMA (×200), **d** cytoplasmic positivity for vimentin (×200), **e** membranous positivity for HMW (×200), and **f** LMW (×200), **g** patchy positivity for S-100 (×200), **h** high expression of high MIB-1 labeling index (×200)

However, patients with residual or recurrent tumors following surgery may be candidates for stereotactic radiosurgery, and other systemic therapies may be considered for unresectable or recurrent tumors on a clinical trial basis.

With respect to the differential diagnosis of CM of which three cases also bear clear cell components, some tumors with vacuolated cytoplasm, abundant chondroid/myxoid matrix, and arising within or near the CNS, such as chondroid chordoma, extraskeletal and skeletal myxoid chondrosarcoma, low-grade chondrosarcoma, enchondroma, chordoid glioma, metastatic mucinous carcinoma, or metastatic renal cell carcinoma warrant special attention. The differential diagnosis with chordoma is of particular importance given its often midline skull base location. The diagnosis of CM is secured if typical meningothelial differentiation evidenced by whorl formation or intranuclear

inclusions is identified. In the event of overwhelming chordoid presence, the immunohistochemical study becomes of ultimate importance.

D2-40 is a monoclonal antibody that was initially developed against M2A, a 38-kD, mucin-type transmembrane glycoprotein, and fetal testis-related antigen, now known as podoplanin [31–33]. D2-40 has been used in the clinical setting as a selective marker of lymphatic endothelium [33, 34] and in the identification of various normal and neoplastic tissues [35]. Within the CNS, D2-40 stains both nonneoplastic (choroid plexus epithelium, ependyma, subependymal area, leptomeninges, and cerebellar Purkinje cells) and neoplastic (ependymomas, choroids plexus papillomas, meningiomas, gliomas, and medulloblastomas) tissues [36, 37]. D2-40 was also reported to stain normal cartilage, true chondroid tumors, CMs, and chordoid gliomas, but not chordomas [5, 38]. In a previous report concerning immunohistochemical marker expressions in distinguishing CMs with their histological mimickers [5], D2-40 showed positivity in a majority of skeletal myxoid chondrosarcomas, enchondromas, low-grade chondrosarcomas, CMs, and chordoid gliomas, but not extraskeletal myxoid chondrosarcomas or chordomas, making D2-40 very helpful in excluding the latter two entities. Chordomas have been shown to robustly express epithelial markers such as cytokeratins and EMA [39]. While S100 has been well established as a chondroid marker, both chordomas and chondrosarcomas typically express this protein [40, 41]. As in our cases, a minority of CM is focally positive for S100 and cytokeratins, markers typically associated with chordomas [3], sometimes rendering the markers nearly useless. Similar to S100, vimentin is very limited in utility due to poor specificity [42], and is shared by many entities. Because cytokeratins and EMA also highlight chordoid gliomas [5], the most telling marker turns out to be GFAP, with strong, diffuse staining supportive of chordoid gliomas. EMA is the most effective antibody for differentiating CMs from skeletal myxoid chondrosarcomas, low-grade chondrosarcomas, and enchondromas, whereas D2-40 is the most effective antibody for differentiating CM from extraskeletal myxoid chondrosarcomas and chordomas. GFAP proves to be sensitive and specific marker for chordoid glioma. As to metastatic mucinous carcinoma and metastatic renal cell carcinoma occurring within and near CNS, clinical data and imaging information are of great help, and a strong, diffuse expression of cytokeratins is indicative of carcinomas. Moreover, D2-40 and CD10 also effectively distinguish CM from renal cell carcinoma [36].

Our findings demonstrate that, in conjunction with clinical and radiographic findings, immunohistochemical evaluation with a panel of podoplanin, D2-40, EMA, cytokeratins, GFAP, and CD10 is most useful in distinguishing CM from chordoma, chordoid glioma, skeletal myxoid chondrosarcoma, extraskeletal myxoid chondrosarcoma, low-grade chondrosarcoma, enchondroma, metastatic mucinous carcinoma, and metastatic renal cell carcinoma. This series is different from previous ones in that it highlights an older age group, and the absence of the systemic complications that can accompany the presence of CM.

Conclusion

Since the original description of this histologic subtype, there have been a few studies in the literature that have demonstrated that (1) chordoid meningiomas perhaps have an increased propensity to recur, (2) this subtype is not always associated with Castleman's Syndrome, (3) this subtype can be associated with T-cell rather than B-cell infiltration, and (4) this histologic category can be seen in the elderly as opposed to only in younger age groups. Our study reinforces these points. And, in conjunction with clinical and radiographic findings, immunohistochemical evaluation with a panel of podoplanin, D2-40, EMA, cytokeratins, GFAP, and CD10 is very helpful in distinguishing CM from chordoma, chordoid glioma, skeletal myxoid chondrosarcoma, extraskeletal myxoid chondrosarcoma, low-grade chondrosarcoma, enchondroma, metastatic mucinous carcinoma, and metastatic renal cell carcinoma.

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