

## Chordoid Meningioma : A Case Report

The term "chordoid meningioma" means meningioma, which is pathologically similar to chordoma, and previously reported that rarely associated with microcytic anemia and/or dysgammaglobulinemia especially in pediatric population. We present a case of this rare variant, which comprises less than 0.5% of all meningiomas. A 33-yr-old man visited our hospital, complaining visual field defect worsening over 7 yr. Neurological examination showed left homonymous hemianopsia. The brain magnetic resonance imaging revealed well enhancing right temporo-occipital mass with cystic portion. Histopathologic findings of resected tumor were compatible with chordoid meningioma which included trabeculae of eosinophilic, vacuolated cells in a myxoid matrix with prominent lymphoplasmacellular infiltration. The neoplastic cells were positive for vimentin and epithelial membrane antigen and negative for glial fibrillary acidic protein and cytokeratin. This is an adult case of chordoid meningioma without anemia or dysgammaglobulinemia.

Key Words : *Meningioma; Chordoma*

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## INTRODUCTION

The term "chordoid meningiomas" was first used by Kepes et al. in 1988 to describe a meningeal tumor among young patients. They reported 7 collected cases, and all of them exhibited hypochromic/microcytic anemia, and one patient showed dysgammaglobulinemia with bone marrow plasmacytosis (Castleman syndrome) which disappeared after tumor removal (1). Such tumors were composed of eosinophilic vacuolated cells disposed in chordoma-like clusters and cords in a myxoid matrix, and often featured a prominent lymphoplasmacellular infiltrate. The 1993 World Health Organization (WHO) classification of Tumors of the Central Nervous System accepted lymphoplasmacyte-rich meningioma and chordoid meningioma as variants of meningioma (2). These tumors contain abundant lymphoplasmacellular infiltration and were previously reported to be frequently associated with hematological abnormalities (3-5). More recently, however, many cases of chordoid meningioma with no hematological abnormalities (6-8) are being reported (Table 1). The chordoid meningioma comprises approximately 0.5% or less of all meningiomas (6). In Korea, the first reported case was a 55-yr-old woman who exhibited polyclonal gammopathy, which is one of the signs of Castleman syndrome (9). We report the second case of chordoid meningioma, not associated with any hematologic abnormality or Castleman syndrome.

## CASE REPORT

A 33-yr-old man visited our hospital and his chief complaint was visual field defect worsening over 7 yr, especially in left infero-temporal quadrant zone. Otherwise his past medical history was unremarkable.

Neurological examination showed no focal deficit except left homonymous hemianopsia. In a routine laboratory test, the hemoglobin, mean corpuscular volume and mean corpuscular hemoglobin were 13.7 g/dL, 90.2 fL and 30.6 pg, respectively. The total protein, albumin and globulin were 7.7 g/dL, 4.5 g/dL and 3.2 g/dL, thus albumin-globulin ratio was 1.4 (reference range 1.3-2.2). There was no laboratory finding suggesting hematologic abnormality or dysgammaglobulinemia. Magnetic resonance imaging scan revealed right temporo-occipital mass, which showed iso-signal intensity on T1&T2 weighted image and intense uniform enhancement of solid portion following intravenous gadolinium administration (Fig. 1). Thus our preoperative diagnosis was meningioma.

Right temporo-occipital craniotomy was performed, and yellowish hard mass was seen with scant vascularity. After puncturing peripheral cystic portion, yellowish fluid was drained. There was no gross invasion to adjacent brain tissue, and the mass was removed totally with adhering dura.

The visual field defect was improved immediately after operation, and there is no evidence of tumor recurrence during the 7-month follow-up period.

Table 1. Literature review of chordoid meningioma cases

Authors (Reference No.)	Age (yr)	No. of cases	Systemic effect (No. of patient)
Kepes et al., 1988 (1)	8-19 (mean:14.8)	7	Anemia (7), dysgammaglobulinemia (1)
Glasier et al., 1993	15	1	Not available
Zuppan et al., 1994	10	1	No systemic effect
Kumar et al., 1996	5	1	Not available
Civit et al., 1997	21	1	Anemia
Kobata et al., 1998 (14)	15	1	Anemia
Kajiwara et al., 1999	52	1	Not available
Couce et al., 2000 (6)	12-77 (mean:47.4)*	42	No systemic effect
Yano et al., 2000 (8)	55	1	No systemic effect
Lee et al., 2001 (9)	44	1	Dysgammaglobulinemia
Mori et al., 2001 (7)	62	1	No systemic effect

\*Only 2 patients were young.

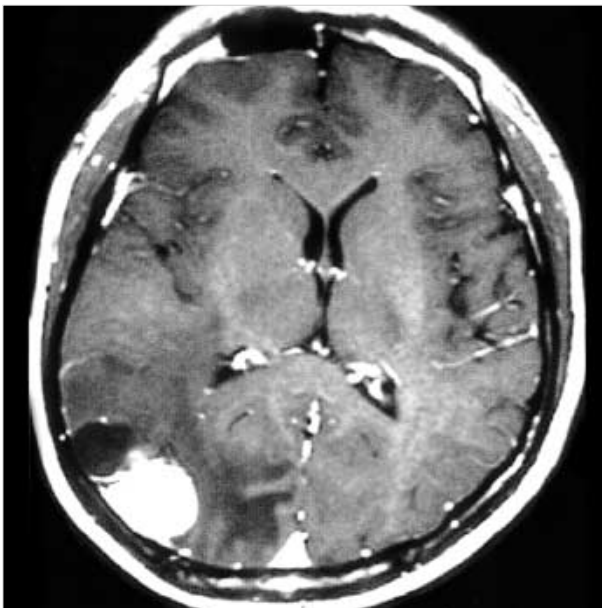


Fig. 1. Axial T1 post-contrast magnetic resonance image shows uniformly enhanced right temporo-occipital mass with peripheral cystic formation.

### Pathological finding

Histologically, the tumor mainly consisted of trabeculae or cords of eosinophilic vacuolated cells in the abundant mucoid matrix (Fig. 2). This chordoid area comprised the majority (about 95%) of the tumor, which intermixed with small areas of conventional meningioma. There were prominent inflammatory cell infiltrates within the mass as well as around the tumor margin. Neither nuclear pleomorphism including mitosis nor necrosis was found. Immunohistochemically, tumor cells showed a typical membranous staining of meningioma for epithelial membrane antigen in focal areas (Fig. 3) and diffuse cytoplasmic staining for vimentin (Fig. 4). None of tumor cells expressed glial fibrillary acidic protein, cytokeratin (CAM5.2), S-100 protein or p53. Ki-67

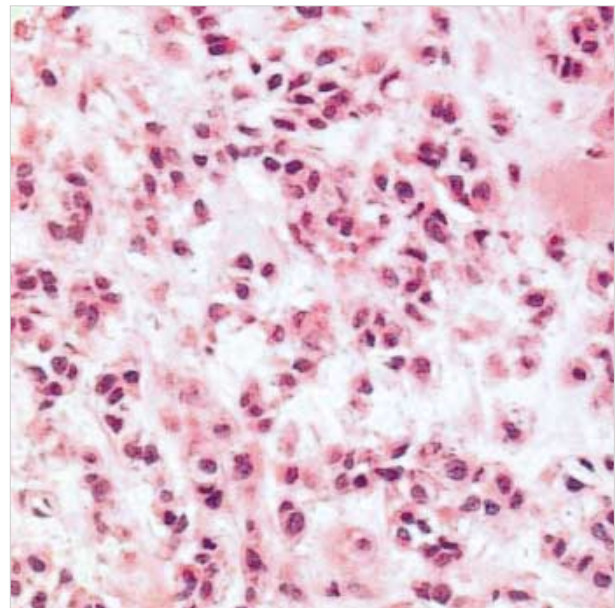


Fig. 2. The histopathology of the tumor shows microscopic finding very similar to that of chordoma. Note cords or trabeculae of epithelioid tumor cells in a myxoid stroma (H&E stain,  $\times 200$ ).

was positive in a few tumor cells. The inflammatory cells consisted of mixed B- and T-lymphocytes but T-lymphocytes predominated within the mass (Fig. 5, 6). All these findings were consistent with those of chordoid meningioma (6, 10, 11).

### DISCUSSION

Chordoid meningiomas show trabeculae or cords of eosinophilic vacuolated cells in a myxoid matrix, and thus may mimic the histological appearance of chordoma. Typical physaliphorous tumor cells, however, are absent in chordoid meningiomas. In addition, the presence of histologic and immunohistochemical features of a classical meningioma, i.e., whorl formation and immunohistochemical staining for vimentin and epithelial

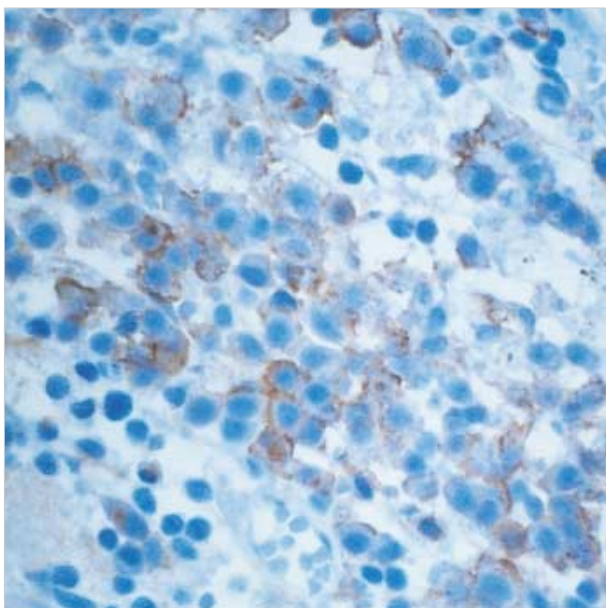


Fig. 3. Tumor cells show membranous expression of epithelial membrane antigen (immunohistochemical stain,  $\times 400$ ).

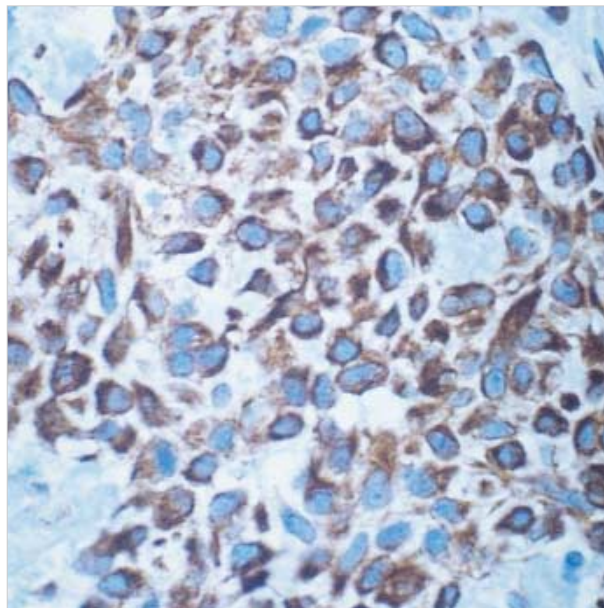


Fig. 4. Tumor cells are strongly reactive for vimentin (immunohistochemical stain,  $\times 400$ ).

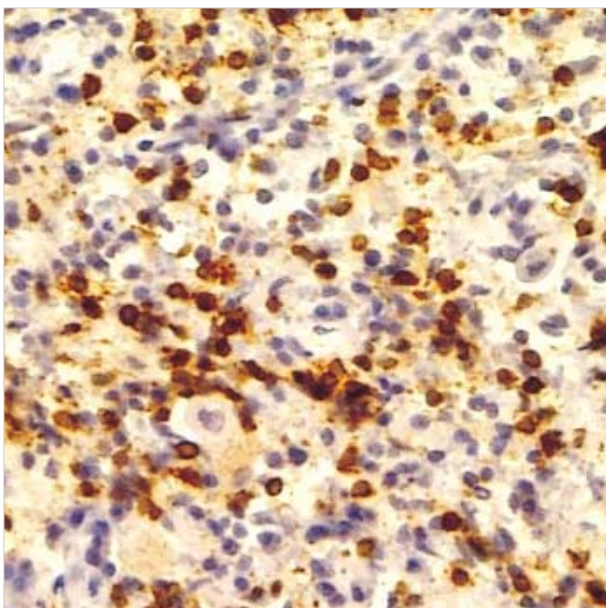


Fig. 5. T-lymphocytes predominate within the mass (immunohistochemical stain-CD3,  $\times 200$ )

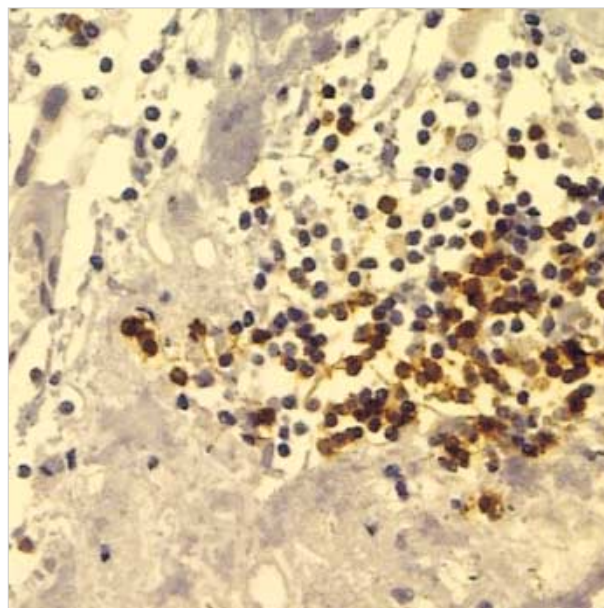


Fig. 6. B-lymphocytes also present within the mass (immunohistochemical stain-CD20,  $\times 200$ )

membrane antigen in the absence of prominent cytokeratin expression, facilitates the differential diagnosis (6, 11).

The differential diagnosis of chordoid meningioma includes chordoid chordoma, myxoid chondrosarcoma, chordoid glioma and other variants of meningiomas (4, 6, 11-14). Chordomas usually locate in the midline and show strong widespread staining for cytokeratin and epithelial membrane antigen (4). Myxoid chondrosarcoma has not been described in the

central nervous system, and usually positive for S-100 protein (13). Chordoid glioma is strongly glial fibrillary acidic protein-positive (12). Microcystic variant of meningioma shows considerable overlap with some features of chordoid meningioma, because it may display certain degrees of myxoid stromal changes and cytoplasmic vacuolation (11, 14). However, this variant lacks inflammatory cell infiltrates. Lymphoplasmacyte-rich meningiomas do not exhibit chordoid features (15).

In clinical aspect, the chordoid meningioma corresponds to WHO grade II (atypical meningioma) because of its more aggressive clinical behavior, and is recommended to be followed up at regular intervals after surgery (10, 11).

About association of chordoid meningioma and lymphoplasmacyte-rich meningioma with Castleman syndrome (1, 3, 5, 14), as may dysgammaglobulinemia and microcytic hypochromic anemia especially in pediatric population, it is postulated that the cause of Castleman syndrome is immunological host reactions to the tumor (1). Castleman's disease presents with localized or disseminated lymphadenopathy (giant lymph node hyperplasia) with systemic symptoms among some patients. The disseminated form is often accompanied by anemia and polyclonal hypergammaglobulinemia, and the condition seems to be related to an overproduction of interleukin 6, possibly produced by human herpesvirus 8 (16). Patients with localized disease can be treated effectively with local therapy, while the initial treatment for patients with disseminated disease is usually done with systemic glucocorticoids (16). In the neurosurgical era, many cases with chordoid meningioma revealed that such systemic manifestations disappeared after the removal of the tumor (1, 3, 9, 14, 15).

It is noteworthy that chordoid and lymphoplasmacyte-rich meningiomas were strongly associated with predominantly B-cells and plasma cells (1, 5, 14, 15) especially in young patients and many of the cases were also associated with Castleman syndrome. However, Couce et al. reviewed 42 cases of chordoid meningioma, mostly among adults (mean age of 47.4 yrs), and reported that none of them showed anemia and/or dysgammaglobulinemia (6). Immunohistochemical staining results of available 5 cases confirmed that the infiltrating lymphocytes were predominantly T-cells (6). They concluded the chordoid meningiomas occur primarily in adults, and there was no significant association with systemic manifestations at least in adults. It seems that B-cells and plasma cells cause these systemic manifestations. However, chronic inflammatory infiltrates are not necessarily composed of B-cell lineage, the chordoid meningioma may not be associated with Castleman syndrome. There have been a few case reports that support this (7, 8) and so does ours. Our case was a 33-yr-old man, and he did not show any sign of Castleman syndrome. A more comprehensive study will be required to know whether B-cells are more prone to infiltrate the chordoid meningioma of childhood, and if so, why it is.

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