CASE REPORT

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Chordoid Meningioma

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A case of chordoid meningioma occurring in a 61-year-old woman who did not have a Castleman's disease is presented. The patient had suffered from headache and motor dysphasia. Laboratory findings are normal. The tumor, located in the left frontal region and associated with peritumoral edema, was totally resected. Surgical specimen revealed a solid mass with irregular surface that measured 3.5×4.5×4cm. Immunohistochemical staining revealed that the tumor cells expressed epithelial membrane antigen(EMA) focally, but not S-100 protein and glial fibrillary acid protein (GFAP), and the Ki-67 proliferative index of the tumor was 9%. The neoplasm was diagnosed chordoid meningioma of the World Health Organization(WHO) grade II. After total resection, her preoperative headache and dysphasia were disappeared.

KEY WORDS : Meningioma · Chordoid meningioma · Castleman's disease.

Introduction

Meningiomas are very common intracranial tumor. Most of these tumors are benign and surgical resection is not difficult. But 2~10% of meningiomas are tend to be malignant[14]. This tumor has high recurrent rate and is very difficult to resect totally. Chordoid meningioma is a relatively rare histological subtype and accounts for 0.5% of all meningiomas[5]. It is associated with high tendency to recur[5,10]. This tumor cells resemble those of a chordoma[6], but there are large areas of lymphocytic cells and plasma infiltration. Many cases of chordoid meningiomas have presented with microcytic normo-or hypochromic anemia, or Castleman's disease in young individuals[5-9]. We report a case of chordoid meningioma in a 61-year-old woman who had not systemic disease.

Case Report

History and Neurological examination

A 61-year-old female complained of moderate headache and motor dysphasia. There was no history of trauma or significant medical problems.

Radiologic and laboratory findings

Skull X-ray showed no abnormal findings as osteolytic or -blastic lesions. Brain magnetic resonance imaging(MRI) revealed a round solid mass in left frontal region(Fig. 1). This mass measured 3.5×4.5×3.5cm. The lesion was highly homogenous enhanced extra-axial mass in the gadolinium-enhanced T1-weighted image. Mild peritumoral edema and dural tail sign were showed. Margin of tumor was obscured because of loss of cerebrospinal fluid space. Laboratory findings are normal.

Operative findings

The bone flap was intact, but the dura was infiltrated by

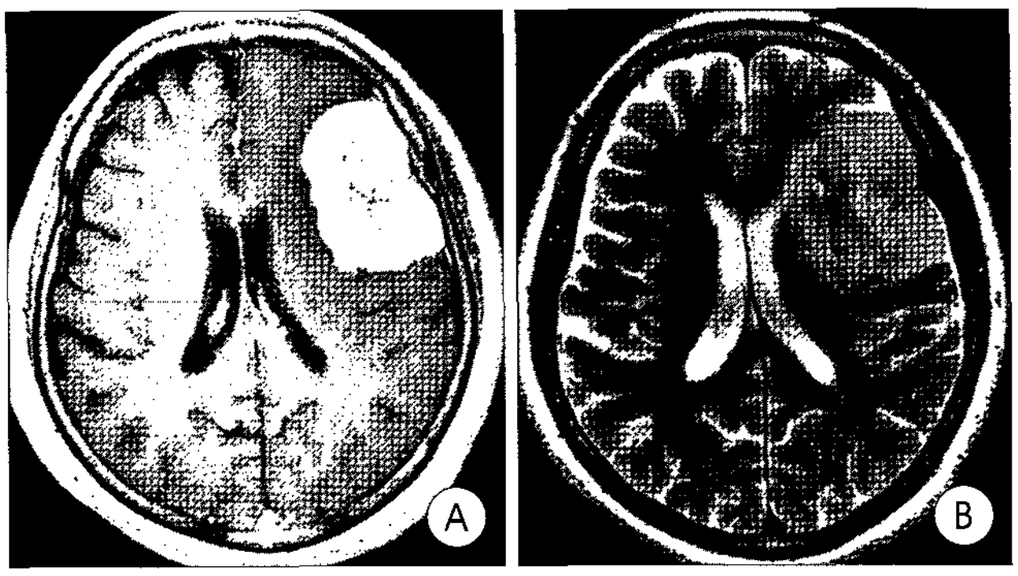

Fig. 1. A : Axial T2-weighted magnetic resonance (MR) image demonstrating a homogeneously iso- intensity mass approximately 4.5cm in diameter with peritumoral edema. B : Axial T1-weighted gadolium-enhanced MR image demonstrating a homogeneously well-enhancing mass with broad base in dura of left frontal convexity.

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the tumor. The dura was opened, the tumor was debulked internally and completely removed with the affected dura. Duroplasty was performed with customized synthetic dura. Immediately post operative clinical course was uneventful.

Histopathologic findings

Gross examination of the surgical specimen revealed a solid mass with irregular surface. The cut surface were pale gray and somewhat myxoid. Microscopically, large portion of the tumor consisted of characterized spindle shaped tumor cells with eosionophilic cytoplasm and trabeculated tumor cell with vacuolation in myxoid matrix, others were solid enough to suggest a meningothelial pattern of meningioma (Fig. 2). The cellularity of tumor was increased and mitosis were noted 8-9 per 10 high power fields in the most active areas. Immunohistochemical staining revealed that the tumor cells expressed epithelial membrane antigen(EMA) focally, but not S-100 protein and glial fibrillary acid protein(GFAP), and the Ki-67 proliferative index of the tumor was 9%. The neoplasm was identified chordoid meningioma as WHO grade II.

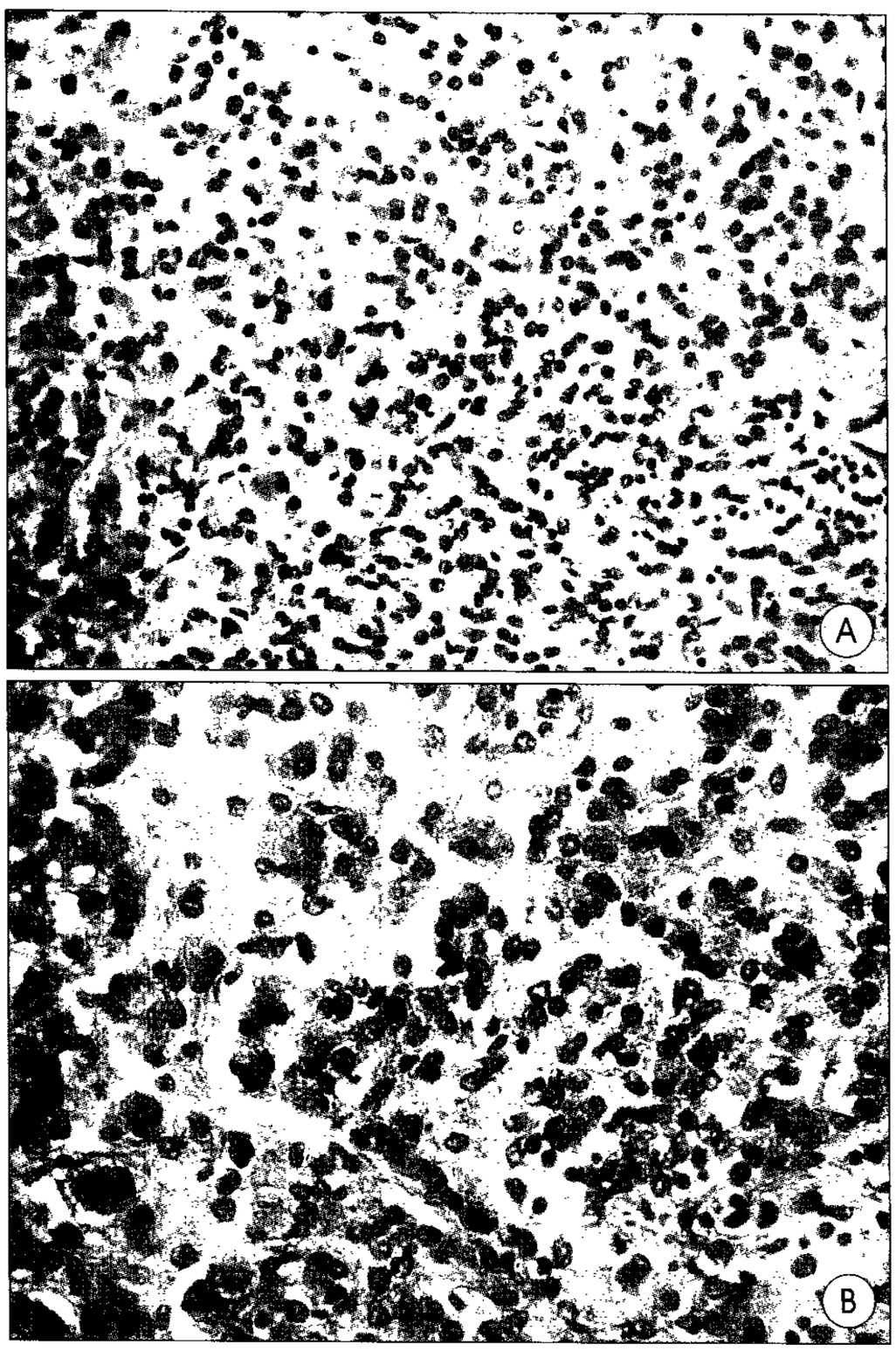

Fig. 2. Photomicrorgraph of the tumor specimen. A : Similar to chordoma, spindle shaped tumor cell with eosionophilic cytoplasm (H&E, x100). B : Trabeculated tumor cell with vacuolation in myxoid matrix(H&E, x400).

Postoperative course

The patient's postoperative course was uneventful. Her preoperative headache and dysphasia were disappeared. The brain computed tomography(CT) at 3months after operation showed no tumor recurrence.

Discussion

Meningioma is common intracranial tumor that occurs usually in the fifth and sixth decades of the life and accounts for approximately 15% of primary CNS tumors[10]. The most recent WHO classification of brain tumors divides meningeal tumors into three groups : 1) benign meningiomas of WHO Grade I, 2) atypical meningiomas of WHO Grade II, 3) anaplastic meningiomas of WHO Grade III. In addition, there are rare variant that are associated with less favorable clinical outcome, and that correspond to WHO Grades II and III[10]. Chordoid meningioma is rare type and its incidence is 0.5% of all meningiomas[5]. It is classified as WHO grade II and has high rate of recurrence(85.7%) after subtotal resection[5,10].

The term "chordoid meningiomas" was used by Kepes et al. to describe a meningeal tumor of young patients associated with iron-resistant microcytic anemia and/or dysgammaglobulinemia in 1988[6,7]. And Nho et al. reported chordoid meningioma with monoclonal gammopathy[11]. These authors also described a meningeal tumor with a chordoma-like histological appearance in patients, and noted an association between this lesion and peritumoral lymphoplasmacellular infiltration causing Castleman's disease[2,3,6,7,8,9].

The published reports on chordoid meningioma consist mainly of single case reports[6,8,9]. Although these cases were associated with systemic manifestation of Castleman's disease, Couce et al.[5] found no significant link between chordoid meningioma and systemic or hematological abnormalities in adults. He concluded that chronic inflammatory infiltration not a mandatory histopathological criterion for diagnosing chordoid meningioma and suggested that this relationship pertains only to chordoid meningioma of childhood[5]. In our case, the patient had not sytemic or hematological abnormalities and not revealed chronic inflammatory infiltration. Most of cases of chordoid meningiomas have occurred in the first decade or first two decades. But Couce et al. reported in his 40 reviews of 42 cases that the patient's mean age was 44years (range from 12 to 77years)[5]. We thought that the patient's age is not specific finding in chordoid meningiomas as our patient being 61years old.

Diagnosis of chordoid menigioma should be based on the

chordoma-like histological appearance of the tumor as well as on the meningothelial cell-like pattern that should be recognizable in at least some areas. Differential diagnosis should be established from chordoma, myxoid chondrosarcoma, recently described chordoid glioma, and certain tumor-like conditions associated with mass and lymphoplasmacellular infiltration. The differential diagnosis for chordoid meningiomas includes chordoid neoplasm, and chordoma is the most important tumor to be excluded. Chordoid meningioma can be distinguished from chordomas based on tumor location and immunohistochemical findings. Chordoma usually located extradural midline structures, such as clivus and sellar area. Immunohistochemisty for S-100 protein and EMA also help to distinguish from theses. Unlike chodoid meningioma, chordoma cells are positive for S-100 protein and negative for EMA[1]. Extra skeletal myxoid chondrosarcomas have not been described in the central nervous system. They are usually positive for vimentin, synaptophysin, S-100 protein and EMA[12]. Lastly, recently described third ventricular "chordoid glioma", unlike the chordoid meningioma, tumor cells show strong positive staining for GFAP[13].

Conclusion

Chordoid meningiomas are very rare subtypes of meningiomas. They are classified as WHO grade II. Although, many single chordoid meningioma cases reported associate with systemic or hematologic manifestations, our case had not associated with this manifestations. We described this case with particular interest because of its rarity and the challenges of differentiating chordoid meningioma from other chordoid neoplasms.

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