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증 례

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-증례보고-

연세대학교 의과대학 신경외과학교실,[1] 국민건강보험공단 일산병원 신경외과,[2] 포천중문 의과대학 신경외과학교실[3]
이창규[1] · 이윤호[2] · 심규원[1] · 김동석[1] · 최중언[3]

Disseminated Anaplastic Meningioma without Solid Mass
- Case Report -

Chang-Kyu Lee M.D.,[1] Yun-Ho Lee M.D.,[2] Kyu-Won Shim M.D.,[1]
Dong-Seok Kim M.D., Ph.D.,[1] Joong-Uhn Choi M.D., Ph.D.[3]
Department of Neurosurgery,[1] Yonsei University College of Medicine, Seoul, Korea
Department of Neurosurgery,[2] National Health Insurance Corporation, Ilsan Hospital, Goyang, Korea
Department of Neurosurgery,[3] Pochon CHA University College of Medicine, Sungnam, Korea

ABSTRACT

Meningiomas arising from the dural coverings of the brain are most common benign intracranial tumor. Mostly, meningiomas are dural based mass lesions and anaplastic meningioma is an uncommon variant of meningioma. We present a case of primary disseminated anaplastic meningioma without solid mass.

A 19-year-old man admitted to the department of neurology because of a 6-month history of headache with nausea, vomiting, and back pain. Magnetic resonance images (MRI) of the brain showed lesions at the right mesial temporal lobe and ipsilateral trigeminal nerve, which showed irregular Gadolinium-enhancement. However, the lesions had no definitive mass formation. Fluid-attenuated inversion recovery (FLAIR) MRI demonstrated high signal intensity on bilateral cingulate gyrus. Cerebrospinal fluid examination revealed a high opening pressure of 400 mmH_2O. Protein was elevated with 338 mg/dL. White blood cell (WBC) level was high with 29 per high power field (HPF), and glucose decreased with 3 mg/dL.

The patient underwent surgical resection for pathologic diagnosis. Pathological diagnosis was anaplastic meningioma showing rhabdoid features with multiple parenchymal and perivascular invasion He underwent radiation therapy including whole neuraxis.

Even though primary leptomeningeal enhancement without solid mass in MRI and more suggestive to inflammatory disease on CSF examination are showed, it should not be rule out the possibility of malignancy. The pathological diagnosis of suspected lesion with unusual situation and uncertain behavior is extremely important.

KEY WORDS : Anaplastic meningioma · Carcinomatous meningitis · Meningoencephalitis · Rhabdoid meningioma.

Introduction

The meningiomas are slow-growing tumor that forms in one of the inner layers of the meninges. They are mostly benign nature of the brain tumors and carry the possibility of complete cure. Thus the long-term outcome of a patient with this tumor is generally good. Meningiomas are rarely malignant in their behavior but when malignant, meningiomas grow rapidly and are destructive.[5,13,14] Malignant meningiomas are difficult to treat and often recur even though surgical tumor removal. The World Health Organization classification sys-

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교신저자 : 심규원, 120-752 서울 서대문구 신촌동 134번지
연세대학교 의과대학 신경외과학교실
전화 : 02) 2228-2152, 전송 : 02) 393-9979
E-mail : shimkyuwon@yuhs.ac

tem defines both grade II and grade III meningioma as malignant, for example, clear cell, chordoid, rhabdoid and papillary type.[5,15] They are attached to the dura and compress the underlying brain without invading it.[4] Dural invasion found macroscopically in 49−72% of meningiomas, microscopically in 57−63%.[4] Even though invasion of the dura and dural sinus is not uncommon, meningiomas are usually easily separated from the pia mater. And usually metastatic brain tumor or spinal cord tumor was noted with leptomeningeal spread. But meningioma with leptomeningeal carcinomatosis is rare and aggressive.[13,14] Rhabdoid meningiomas are highly aggressive tumors known for their recurrence and often carry a poor prognosis.[5,13,14] Also, they recurred with dissemination along the cerebral spinal fluid spaces rarely. We report a case of disseminated anaplastic meningioma with rhabdoid features without solid mass.

Case

A 19-year-old man presented with headache, nausea, vomiting, and back pain for 6 months. He did not have a family history like neurofibromatosis. The symptoms developed after he went swimming in the river. He was checked with magnetic resonance images (MRI) at another hospital at 6 months ago, it did not show any abnormal finding. While his headache was not subsided, he admitted to that hospital. White blood cells (WBS) [10/10 high power field (HPF)], protein (83 mg/dL), and glucose (51 mg/dL) were reported in the cerebrospinal fluid (CSF) examination and culture study for bacterial and fungal infection was negative. Because the result of culture study for mycobacterium is needed for 8 weeks, neurologists diagnosed as tuberculous meningoencephalitis, clinically. Therefore, the medication for tuberculous meningoencephalitis was taken in that hospital. However, his headache was intractable to medication. Three months later, the patient visited emergency room in our hospital with aggregated headache and back pain. Because of his history of presumed diagnosis of tuberculous meningoencephalitis, the department of neurology performed a spinal tap for CSF examination. The CSF examination revealed a high opening pressure of 400 mmH_2O cerebrospinal fluid. Protein was elevated with 338 mg/dL. WBC level was high with 29 per 10 HPF, and glucose decreased with 3 mg/dL. The CSF cytology was not seen any malignancy and inflammatory cells. Culture study for bacterial and fungal in CSF was sterile. On the MRI of the brain, T2 weighted images showed increased signal intensity at right mesial temporal lobe and T1 weighted images with Gadolinium revealed disseminated enhancement at right trigeminal nerve and leptomeninges of the right mesial temporal lobe without solid mass. Fluid-attenuated inversion recovery (FLAIR) MRI demonstrated high signal intensity on both cingulate gyri (Fig. 1). Neurologists diagnosed intracranial hypertension with meningoencephalitis, and consulted to our department for surgical management of intracranial hypertension. We performed lumbo-peritoneal shunt. Despite of the lumbo-peritoneal shunt, headache and back pain was still remained. One month later, follow-up MRI of the brain revealed more increased enhancement at right mesial temporal lobe and newly developed leptomeningeal enhancing lesion at right parietal lobe compared with prior images (Fig. 2). In the lumbar spine MRI showed multiple enhancements at nerve root (Fig. 3). We performed a surgical resection of anterior temporal lobe including anterior mesial temporal area, measuring 5.5×4 cm to confirm its histopathology (Fig. 4). Histopathological examination showed a malignant tumor and multiple parenchymal invasions of the tumor. The tumor cell was seen in meninges, mostly. The Ki-67 labeling index was more than 10%. It was positive for vimentin, but not epithelial membrane antigen (EMA) (Fig.

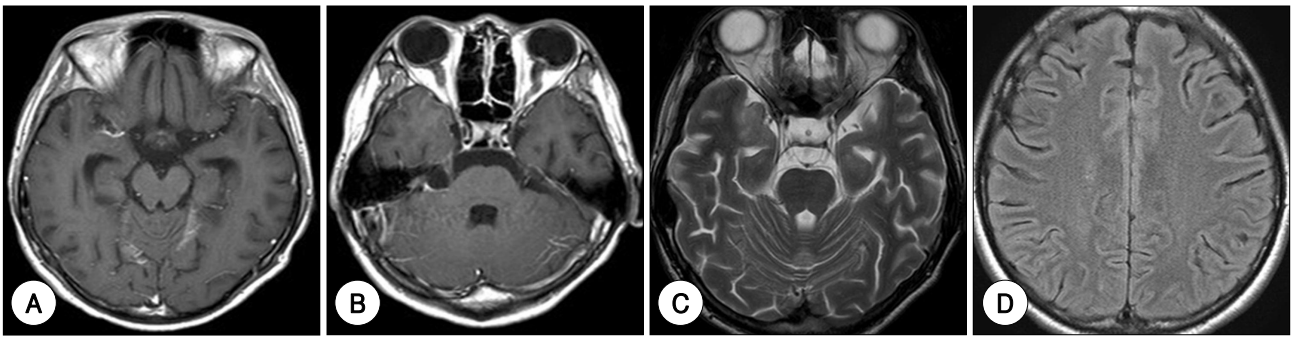

Fig. 1. T1-weighted gadolinium-enhanced axial magnetic resonance images (MRI) scans showing enhancement of the mesial temporal lobe through the leptomeninges (A) and right trigeminal nerve (B). T2-weighted image showing high signal intensity on right mesial temporal lobe (C). FLAIR image showing high-signal intensity of bilateral cingulated gyrus (D).

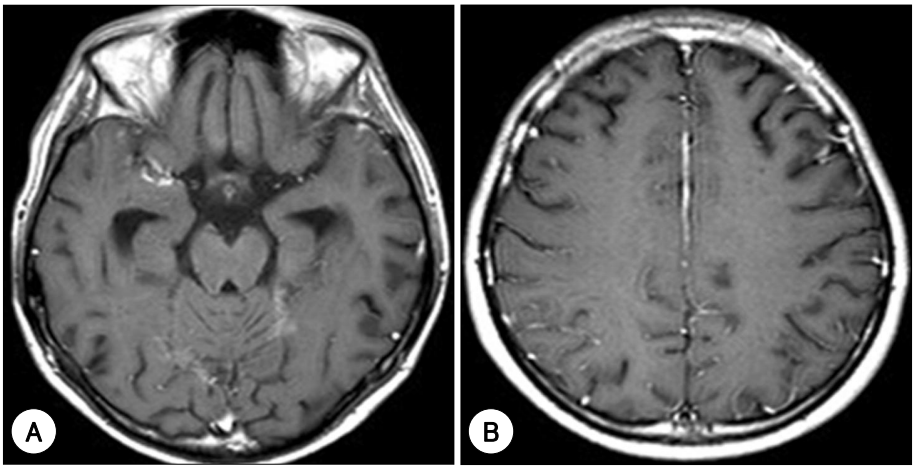

Fig. 2. One month later, T1-weighted gadolinium-enhanced axial MRI scans showing increased enhancement at right mesial temporal lobe(A) and a newly developed leptomeningeal enhancing lesion at the right parietal lobe(B).

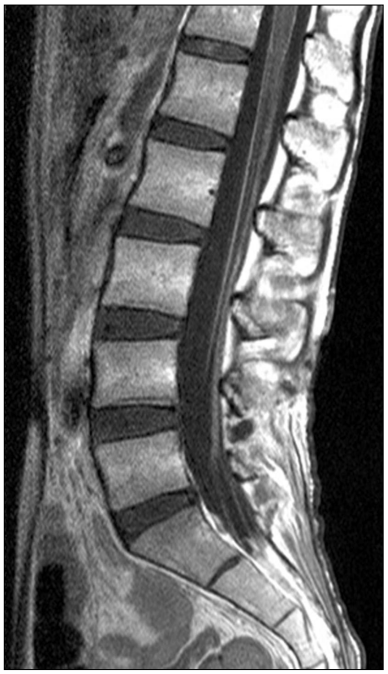

Fig. 3. T1-weighted gadolinium-enhanced sagittal spinal MRI showing multiple enhancements of the nerve root.

5). Because the tumor represented a leptomeningeal spread pattern without solid mass, we checked immunohistochemical stain to distinguish hematologic malignancy and anaplastic lymphoma. Immunohistochemical stain resulted all negative for leukocyte common antigen(LCA), CD3, CD20, T-cell-restricted intracellular antigen(TIA), CD43, anaplastic lymphoma kinase(ALK), and Ki-1. Also, HMB45 immunohistochemical stain for distinguish to melanoma was negative. Finally, histological diagnosis showed most likely anaplastic meningioma showing rhabdoid features with multiple brain parenchymal invasion and perivascular spaces. He underwent whole neuraxis external fractionated radiotherapy of 45 Gy.

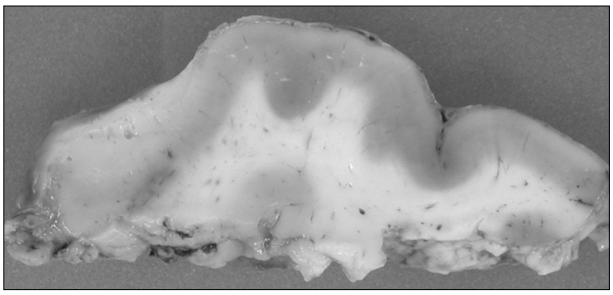

Fig. 4. Photography of the specimen is anterior temporal lobe, measuring 5.5×4 cm sized. The external surface showing a focal fibrosis.

However, after the radiation therapy, the patient declined in general condition. Furthermore he had neutropenic fever and sepsis. Despite the radiation therapy, he was dead 3 months later. An autopsy or post-radiative imaging study could not perform because of parent's disagreement and patient's septic condition

Discussion

Histological grading of meningiomas is based on the current World Health Organization(WHO) classification. Most are WHO grade I, reflecting their benign nature. However, atypical meningiomas(WHO grade II), which make up 5−7%, and anaplastic variants(WHO grade III), 1−3%, are recognized by several histological characteristics.[15] Anaplastic meningioma is defined by histological features, which include either obviously malignant cytology or a high mitotic index-20 mitoses>10 High power field(HPF).[10] Expression of proliferation markers such as Ki-67 has shown increases in the labeling index with WHO classification. The Ki-67 of grade III or anaplastic meningioma was elevated from 5.6 to 19.5%.[6] Invasion of the brain alone is not sufficient for a diagnosis of ana-

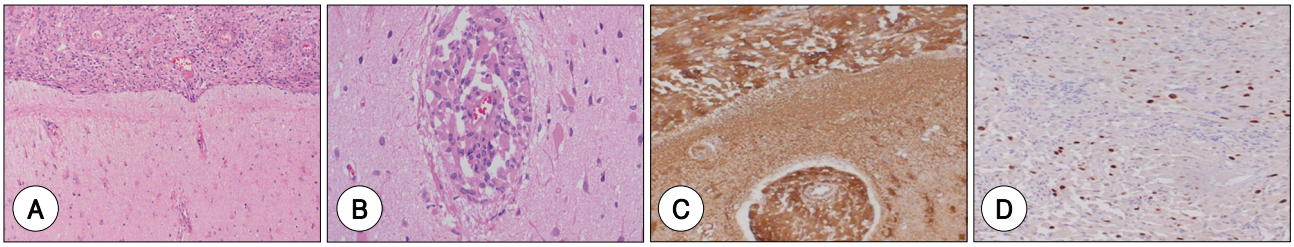

Fig. 5. Histopathological images of the resected right mesial temporal lobe. The tumor cells consisted of rhabdoid features and infiltrated the parenchymal (A) (×50) and perivascular spaces with high mitotic features (B) (×200). Positive for vimentin (C) and high Ki-67 index (D).

plastic meningioma. But, brain invasive lesions related to the high recurrence rate, which is likely the result of microscopic residual tumor tissue within the parenchyma that used not to be resected because of potential morbidity. Rhabdoid meningioma was adopted as a new subtype of meningioma in the WHO Classification of tumors, 2000.[12] This tumor is defined by rhabdoid cells, which have a specific microscopic appearance with eccentric cytoplasm and paranuclear inclusions.[12] Rhabdoid meningioma more tends to have cystic component, prominent peritumoral edema, and bony involvement on MRI.[5] The optimum treatment of anaplastic meningiomas has not been well defined and prognosis is poor, survival following gross total resection of tumor without adjuvant therapy is less than 2 years. Chemotherapy for malignant meningiomas has not shown any convincing effect in several literatures.[3,8,9] Radiotherapy in the treatment of meningiomas is still debatable. However, Pourel et al. reported radiotherapy is a standard treatment for unresectable lesions and histologically proven atypical or anaplastic meningiomas, which still have a poor prognosis.[11] Although some cases were reported for leptomeningeal spread in WHO grade II or III meningiomas,[1,7,13,14] most of cases showed dural attached mass lesion as well as leptomeningeal spread. In 3 cases of leptomeningeal infiltrating meningiomas, histological examination revealed rhabdoid and papillary meningioma with high proliferation rate.[1,14] Another case diagnosed clear cell meningioma.[7] Diffuse leptomeningeal enhancement is usually associated with meningitis or meningoencephalitis, which may be bacterial, viral, or fungal. Also, neoplasms may spread into the subarachnoid space and may produce leptomeningeal enhancement that is often called "carcinomatous meningitis". Leptomeningeal enhancing pattern in neoplasms is generally distinguish to viral or bacterial meningitis, which can produce thick, lumpy, or nodular enhancement, similar to that of fungal disease. In our case, MRI of the brain demonstrated diffuse leptomeningeal, mainly right mesial temporal lobe, and right trigeminal nerve enhancement without dural attached mass lesions. Moreover, laboratory data initially was more suggestive of inflammatory diseases, not to tumor. Therefore, initial diagnosis was confused. No one could think the enhancing lesion to be malignant tumor before its pathological report.

Conclusion

We report an extremely rare case of disseminated primary intracranial anaplastic meningioma without solid mass. In this case, the pathological diagnosis of suspected lesion with unusual situation and uncertain behavior is extremely important.

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