Extracranial Extension of Intracranial Atypical Meningioma En Plaque with Osteoblastic Change of the Skull

Se Youn Jang, M.D.,1 Choong Hyun Kim, M.D., Ph.D.,2 Jin Hwan Cheong, M.D., Ph.D.,3 Jae Min Kim, M.D., Ph.D.2

Department of Neurosurgery,1 Seoul Medical Center, Seoul, Korea
Department of Neurosurgery,2 Hanyang University Guri Hospital, Guri, Korea

Meningioma is a common primary tumor of central nervous system. However, extracranial extension of the intracranial meningioma is unusual, and mostly accompanied the osteolytic change of the skull. We herein describe an atypical meningioma having extracranial extension with hyperostotic change of the skull. The patient was a 72-year-old woman who presented a large mass in the right frontal scalp and left hemiparesis. Brain magnetic resonance imaging and computed tomography scans revealed an intracranial mass, diffuse meningeal thickening, hyperostotic change of the skull with focal extension into the right frontal scalp. She underwent total removal of extracranial tumor, bifrontal craniectomy, and partial removal of intracranial tumor followed by cranioplasty. Tumor pathology was confirmed as atypical meningioma, and she received adjuvant radiotherapy. In this report, we present and discuss a meningioma en plaque of atypical histopathology having an extracranial extension with diffuse intracranial growth and hyperostotic change of the skull.

Key Words: Atypical meningioma · Extracranial extension · Hyperostotic change · Meningioma en plaque.

INTRODUCTION

Meningiomas are the second common central nervous system (CNS) neoplasm in adults and account for 15–20% of all primary brain tumors. Although most meningiomas are benign, approximately 10% demonstrates a more aggressive clinical behavior and are classified as non-benign meningiomas. Extracranial meningiomas are 1–2% of all meningioma, and the majority has a secondary extension of the primary intracranial tumors and accompanies the osteolytic change of the skull. Some intracranial meningiomas may extend to skull leading to cranial hyperostosis. Osteoblastic intraosseous meningiomas may induce hyperostosis. Meningioma en plaque (MEP) represents an infiltration to the dura and sometimes invades the bone with the intraosseous tumor growth leading to significant hyperostosis. But, the concomitant appearance of scalp meningioma associated with intracranial atypical MEP accompanying hyperostosis has been rarely described. We herein describe an uncommon atypical MEP, which accompanied intracranial growth and extracranial extension with hyperostotic change of the skull.

CASE REPORT

A 72-year-old woman presented with a large mass in the right frontal scalp and left sided-motor weakness. The mass was progressively enlarged over 4 years, and motor weakness has been developed since 6 months before admission. Scalp tumor was a well-defined, subcutaneous firm mass, and located in the right frontal area adjacent to the midline. The enhanced computed tomography (CT) scans showed the well-enhanced mass, sized 6.2×2.6×5.1 cm in the right frontal scalp, and diffuse growth of intracranial tumor accompanying the hyperostotic change of the skull bone (Fig. 1A, B). Brain magnetic resonance images revealed the well-enhanced tumors with bony infiltration in the right frontal region, diffuse meningeal thickening with multiple cystic changes in the right hemisphere (Fig. 1C, D). Main feeder of tumor was the right external carotid artery, and anterior part of the superior sagittal sinus was compressed by tumor.
primary tumors. MEPs are presumed to account for 2-4% of intracranial meningiomas. These tumors are defined by an intracranial tumor growth leading to significant hyperostosis and a widespread, carpet-like, soft-tissue growth at the dura. MEP is predominantly found in the middle age, with the peak incidence between fourth and fifth decade years. The female-to-male ratio of incidence is about 3-5:1. MEP is a specific clinicopathological entity, which although locally invasive, usually bears the histology of World Health Organization (WHO) grade I meningioma. Atypical (WHO grade II) meningiomas constitute approximately 5-7% of meningiomas. Especially, atypical meningioma is rare but existed among MEPs. Li et al. reported 2 cases of MEP with atypical pathology out of 37 MEPs in the sphenoid wing. Hyperostosis is a well-known sign of meningiomas, which is observed in 4.5% of all types, but is more frequently observed in meningioma en plaque with an incidence of 13 to 49%. The histological type of meningiomas appears to have no relationship with hyperostosis, but the various histological types are found as MEP, which are frequently associated with hyperostosis. In addition, there is no relationship between the hyperostotic pattern and the histological type of meningioma.

The differential diagnosis includes a number of mesenchymal and epithelial tumors in the head, such as, paraganglioma, carcinoma, melanoma, schwannoma, olfactory neuroblasticoma, fibrous dysplasia, osteoma, osteoblastic metastasis, Paget’s disease, hyperostosis frontalis interna, erythroid hyperplasia, and sarcoidosis or others depending on the anatomic site of involve-

DISCUSSION

Meningiomas are the most common non-glial intracranial primary tumors. MEPs are presumed to account for 2-4% of intracranial meningiomas. These tumors are defined by an intracranial tumor growth leading to significant hyperostosis and a widespread, carpet-like, soft-tissue growth at the dura. MEP is predominantly found in the middle age, with the peak incidence between fourth and fifth decade years. The female-to-male ratio of incidence is about 3-5:1.

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Inward bulging of the inner aspect of the hyperostotic bone, irregularity of the bone surface, and intracranial changes are important imaging features that can be used in differential diagnosis of MEP. The most successful outcome of treatment for atypical meningioma can be obtained by the early and extensive surgical resection. Surgical intervention consists of the removal of all involved lesions including bones, dura, muscles, intracranial and intraorbital components. Radiation therapy has been suggested to yield an improvement in patient survival with some meningiomas of the CNS. Goyal et al. reported that radiation therapy was associated with less recurrence: 22 (78.5%) of the 28 patients who did not develop recurrence were the group that received radiation therapy. Park et al. reported that progression-free survival was significantly higher in patients undergoing radiation therapy after surgical resection than those not undergoing radiation therapy (58.7% vs. 44.3%, at 5 years, p=0.029). Maroon et al. have recommended postoperative radiation therapy when tumors were subtotally removed. It has been also indicated if there is the dural or cavernous sinus invasion of tumor, or tumor recurrence on neuroimaging. Radiation therapy for atypical meningioma should be considered if there are residual tumors or the recurrent tumor after initial radical removal of bone and tumor. In our case, radiation therapy has been also performed, however, patient outcome was not reached to satisfaction. The prognosis of meningioma is generally favorable if the complete excision is attempted. However, rare cases of meningioma like atypical MEP are aggressive and total resection is unfeasible. A few of meningiomas can develop as a malignant pathological type.

CONCLUSION

Meningiomas are mostly benign intracranial lesions and their coexistence with extracranial extension of atypical MEP is rarely reported. We hereby describe and discuss an atypical MEP having an extracranial extension with diffuse intracranial growth. Total resection of tumor is a choice of treatment for meningioma, however, management should be tailored depending on the status of patient.

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