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Convexity *En Plaque* Meningioma Manifesting as Subcutaneous Mass: Case Report

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Abstract

A 67-year-old woman sensed a slowly growing, painless hard mass in the left parietal region. Cranial computed tomography showed focal bony erosion and homogeneous sclerotic change at the affected site. Magnetic resonance (MR) imaging revealed an enhanced subcutaneous mass and irregularly thickened dura mater. Intraoperatively, the subcutaneous tumor was found to be strongly adhered to the temporalis muscle. The outer table was eroded adjacent to the subcutaneous tumor, whereas the bony structures of the inner table were intact. The dura mater underneath had irregular-shaped, yellowish convolutions both on the outer and inner surfaces. The patient underwent total tumor resection with sufficient normal margins. The histological diagnosis was World Health Organization (WHO) grade I meningioma, with finger-like outward extensions through the dura mater and overlying skull, and infiltration among into the temporalis muscle fibers. Meningiomas may form a subcutaneous mass without intracranial growth.

Key words: en plaque meningioma, convexity, subcutaneous mass, tumor extension

Introduction

En plaque meningioma is an uncommon but distinct entity which shows carpet-like proliferation along the dura mater, in contrast to ordinary, massive meningiomas occurring as an intracranial mass with broad attachment to the dura mater. En plaque meningiomas typically develop in the sphenoorbital regions and manifest as ocular motor paresis, visual impairment, and proptosis,[6] whereas those arising in the convexity are rare.[3,5] Hyperostotic changes of the affected skull are thought to be characteristic findings of en plaque meningioma but may be confused with other pathological conditions such as fibrous dysplasia, osteoma, primary meningeal fibrosarcoma, and meningeal sarcoidosis.[2,4,7] The molecular genetics of meningiomas have recently been investigated but still far from a systematic understanding.[1,8] Here we describe a case of en plaque meningioma of the convexity presented with peculiar extracranial extensions identified by histological examination.

Report

A 67-year-old woman sensed a painless hard mass in the left parietal region slowly growing for 2 months. Her medical history was unremarkable and did not include previous head trauma. Cranial computed tomography scans showed focal bony erosions and homogeneous sclerotic change at the affected site (Fig. 1A, B). Magnetic resonance (MR) imaging revealed an enhanced subcutaneous mass and irregularly thickened dura mater below. The diploic tissue of the affected skull was abnormally thinned (Fig. 1C, D).

The patient underwent tumor resection. Intraoperatively, the subcutaneous tumor, measuring 16 × 13 × 5 mm, was found to be firmly adhered to the temporalis muscle. Therefore, the subcutaneous mass was resected en bloc with the surrounding muscle. The affected skull was removed as a single bone flap with sufficient margins guided by neuronavigation. The outer table of the pathological bone was eroded adjacent to the subcutaneous tumor, whereas the inner table appeared yellowish in color and had macroscopically intact bony structures. The inner table was not adhered to the underlying dura mater and got separated from it with ease. The dura mater contained irregular-shaped, yellowish convolutions both on the outer and inner surfaces, and was resected circumferentially with adequate margins (Fig. 2). The arachnoid membrane below the affected dura was intact. The histological diagnosis was consistent with benign meningothelial meningioma [World Health Organization (WHO) grade I], with finger-like outward extensions

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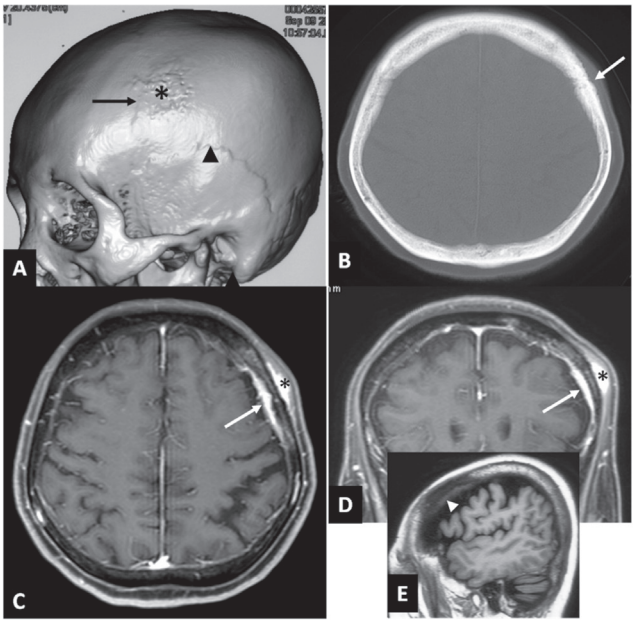

Fig. 1 Presurgical three-dimensional (A) and axial (B) computed tomography scans showing bony erosions (A, *asterisk*) and homogeneous sclerotic change (B, *arrow*) in the left parietal bone. Coronal suture (A, *arrow*); squamosal suture (A, *arrowhead*). Pre-contrast sagittal T_1-weighted (E), and post-contrast axial (C) and coronal (D) T_1-weighted magnetic resonance images demonstrating an intensely enhancing subcutaneous mass, 1.5 × 1.5 cm in diameter (*asterisk*), and irregular thickening of the dura mater underneath (E, *arrowhead* and C, D, *arrow*).

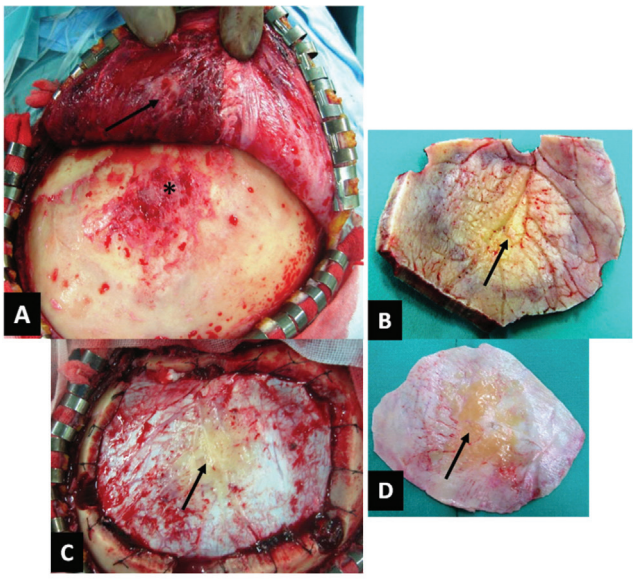

Fig. 2 Intraoperative photographs demonstrating the subcutaneous mass firmly adhered to the temporalis muscle (A, *arrow*), eroded outer table (A, *asterisk*), and the affected dura mater with irregular-shaped, yellowish convolutions both on the outer (C, *arrow*) and inner (D, *arrow*) sides, whereas macroscopically intact surface of the inner table (B, *arrow*).

passing through the dura mater and overlying skull, and infiltration amongst into the temporalis muscle fibers (Fig. 3). The histological appearance was identical for the subcutaneous, diploic, and dural lesions. The Ki-67 labeling index was less than 1%.

Discussion

In the present case, the abnormally thickened focal dura mater observed on MR imaging was identified as sheet-like proliferations of benign meningioma both on the inner and outer surfaces of the dura mater (en plaque meningioma), with histological appearance revealing extracranial tumor extension through the diploe of the overlying skull and tumor-associated bony erosions in the outer table. The extracranial tumor extension had formed a subcutaneous mass with infiltration among the temporalis muscle fibers. On the basis of these findings, we assumed that the present tumor initially occurred as en plaque proliferation, then infiltrated into the overlying skull with finger-like extensions, and finally formed a subcutaneous mass (Fig. 4). Primary ectopic subcutaneous meningioma with intracranial extension may be an alternative explanation for the present case, but it seems less likely considering the mass size which is disproportionally small for causing bony erosions.[9] Alternatively, present tumor initially might have occurred as intraosseous meningioma and thereafter extended bidirectionally both to the intra and extracranially. However,

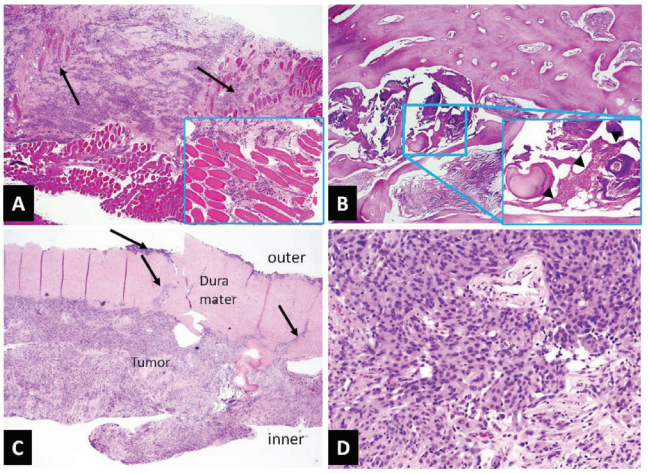

Fig. 3 Photomicrographs of the resected specimens showing A: tumor infiltration amongst into the temporalis muscle fibers (*arrows*) (H-E stain, ×10; inset, ×50), B: tumor extension passing through the diploe (*arrowheads*) (H-E stain, ×10; inset, ×50), C: sheet-like proliferation of the tumor on the inner surface of the dura mater with finger-like outward extensions (*arrows*) (H-E stain, ×10), and D: histological appearance of the tumor consistent with WHO grade I meningothelial meningioma (H-E stain, ×50). WHO: World Health Organization, H-E: hematoxylin-eosin.

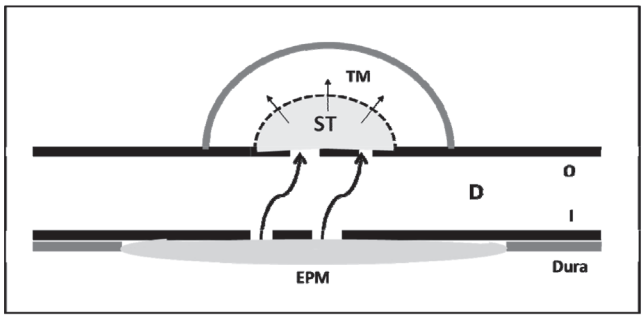

Fig. 4 Schematic illustration showing the assumed pathway of tumor extension (*arrows*) in the present case. D: diploe, EPM: en plaque meningioma, I: inner table, O: outer table, ST: subcutaneous tumor, TM: temporalis muscle.

considering that the lesion did not form a mass in the diploic tissue with abnormally thinned diploe in the affected site, this speculation seems to be less possible. To our knowledge, there has not been a report of subcutaneous meningioma occurring on the cranial convexity, not accompanying intracranial mass.

The molecular genetics of massive meningiomas have recently been investigated,[1,8] while the genetic peculiarities of en plaque meningiomas are little known. Differences in the genetic background between these two entities are also not known. In the present case, en plaque and subcutaneous, massive meningiomas occurred simultaneously in the same patient. Furthermore, in contrast with the macroscopically intact inner table adjacent to the en plaque lesion, the outer table was eroded by the massive tumor overlying it. To date, we do not explain the underlying mechanism of them. Further investigation is needed to understand the biological behavior of meningiomas.

Total resection is the rule for the treatment of low-grade meningiomas. In the present case, en bloc tumour resection involving the affected dura mater, skull, and temporalis muscle was achieved. Meningiomas should be assumed as differential diagnosis of subcutaneous mass lesion even in the absence of an accompaniment of intracranial tumor growth.

Conflicts of Interest Disclosure

The authors have no personal, financial or institutional interest in any of the drugs, materials, or devices in the article. All authors who are members of the Japan Neurosurgical Society (JNS) have registered online self-reported conflict of interest disclosure statement forms through the website for JNS members.

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