Case Report

Atypical convexity meningioma presenting with photophobia and skull erosion✩✩✩

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ABSTRACT

A 41-year-old man presented with photophobia. The patient showed a choked disc and right-sided quadrantanopia with an intact sphincter reaction to light stimulation. Computed tomography revealed an isodense mass in the right frontal convexity, accompanied by extensive perifocal brain edema and smooth-contoured skull erosion. On cerebral magnetic resonance imaging, the tumor was dural-based, appeared inhomogeneous intensity on both T1- and T2-weighted sequences, and was intensely enhanced. Magnetic resonance angiography revealed unusually ectatic right-sided middle meningeal and superficial temporal arteries. The tumor, which was elastic hard, highly vascular, and severely adhered to the frontal cortices, was completely resected. The microscopic findings of the resected specimen were consistent with angiomatous meningioma. The patient’s photophobia resolved after surgery, with resolution of the optic chiasm compression. Meningiomas arising in the frontal convexity may cause photophobia. Angiomatous meningioma should be considered when a broad-based tumor is found in the cerebral convexity accompanied by skull erosion and extensive perifocal edema.

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Introduction

Meningiomas are the most common primary brain tumors occurring in adulthood. They commonly grow as subdural masses eventually compressing the cerebral parenchyma. Hyperostosis in the adjacent skull and dural tail, frequently involving tumor cells, is considered a characteristic finding of meningiomas [1–3]. Conversely, tumor-associated skull erosion and the absence of a dural tail have been documented as radiological findings of meningioma mimics [4–8]. Angiomatous meningioma is a rare subtype of World Health Organization grade I meningioma. It accounts for less than 1% of all intracranial tumors and 2.1% of all meningeal tumors.

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Fig. 1 – Perimetry of the patient shows right-sided inferonasal quadrantanopia.

Fig. 2 – (A) Axial computed tomography scan shows an isodense mass in the right frontal convexity (asterisk) accompanied by extensive perifocal edema. Axial (B) and coronal (C) bone target computed tomography scans at the same level reveal smooth-contoured skull erosion adjacent to the tumor, involving the inner table and diploe (arrows). The outer table is intact and hyperostotic changes are not observed.
Variable cyst components, extensive peritumoral edema, and multiple areas of vascular signal voids are considered the radiological hallmarks. Microscopically, tumor cells are consistent with low-grade meningiomas, characterized by prominent microvessels of varying sizes [9–12].

Photophobia is a well-known symptom; however, its signaling pathways and neurophysiological features are not well understood. It has been observed in patients with unruptured anterior communicating artery aneurysms, pituitary adenomas, craniopharyngiomas, clival chordomas, and demyelinating lesions affecting the optic pathway [13–16].

Here, we present a unique case of angiomatous meningioma that presented with photophobia and was accompanied by skull erosion and an inconspicuous dural tail on neuroimaging.

Case report

A previously healthy 41-year-old man presented with photophobia that had persisted for one month. At the time of presentation, the patient did not exhibit any focal neurological deficits. His pupils were isocoric with an intact sphincter reaction to light stimulation. Ophthalmological examination revealed a choked disc on both optic fundi and right-sided quadrantanopia (Fig. 1).

Computed tomography revealed an isodense mass in the right frontal convexity accompanied by extensive perifocal brain edema. On the bone target images, smooth-contoured skull erosion involving the inner table and diploic was identified adjacent to the tumor. The outer table was intact, and no
Fig. 4 – Magnetic resonance angiography, oblique view, shows an unusually ectatic middle meningeal (arrows) and superficial temporal (dashed arrows) arteries on the right side.

Fig. 5 – Photomicrographs of the resected specimens show tumor tissue comprising cells with oval-shaped nuclei and intervening vasculature of varying sizes (A). Few mitotic figures are observed. Immunohistochemical examination shows positive staining for epithelial membrane antigen (B) and progesterone receptor (C) and negative staining for STAT6 (D). A: hematoxylin and eosin stain, ×400; B: epithelial membrane antigen, ×400; C: progesterone receptor, ×400; D: STAT6, ×400.

hyperostotic changes were found (Fig. 2). On cerebral magnetic resonance imaging (MRI), the tumor was dural-based, measured $33 \times 35 \times 37$ mm, appeared inhomogeneous intensity on both T1- and T2-weighted sequences, and was intensely enhanced (Fig. 3). Moreover, intralesional cysts were found peripherally (Fig. 3D). Magnetic resonance angiography revealed unusually ectatic middle meningeal and superficial temporal arteries on the ipsilateral side of the tumor (Fig. 4). The patient underwent tumor resection. The tumor was elastic hard, highly vascular, and supplied by numerous branches from the
superficial temporal artery that passed through the skull, as well as branches from the middle meningeal artery. Furthermore, it was severely adhered to the frontal cortices. With circumscribed dissection followed by internal decompression with the aid of a cavitron ultrasound surgical aspirator, eventually a gross total resection was achieved with unusually thickened dura mater around the tumor attachment (Simpson grade I). Microscopic findings of the resected tumor showed anaplastic cells with oval-shaped nuclei and intervening rich vasculature. Few mitotic figures were observed. There were no necrotic foci. Immunohistochemical examination showed positive staining for epithelial membrane antigen and progesterone receptor and negative staining for STAT6 (Fig. 5). These findings were consistent angiomatous meningiomas. The patient’s postoperative course was uneventful. His photophobia and visual defects resolved 2 months after surgery. Cerebral MRI performed on postoperative day 5 showed remarkable resolution of compression exerted on the optic chiasm that had been caused by the swollen right frontal lobe (Fig. 6).

**Discussion**

In the present case, photophobia was the main clinical symptom associated with the tumor that resolved after tumor resection. Given that considerable optic nerve compression detected on presurgical MRI resolved promptly after surgery, extensive brain edema accompanying the meningioma was thought to contribute to the photophobia. Photophobia caused by peritumoral edema indirectly compressing the optic pathway has not yet been reported. On the other hand, previous study documented that the trigeminal system and pretectal nuclei may be associated with the pathophysiology of photophobia [13]. Further case accumulation is needed to elucidate the mechanisms and neural pathways of photophobia.

Angiomatous meningioma is a rare subtype of benign meningioma that is radiologically characterized by cyst components, extensive peritumoral edema, and multiple areas of vascular signal voids [11,12]. The present case was accompanied by extensive perifocal edema and skull erosion, while dural tail was inconspicuous. The most useful radiological features for differentiating meningiomas from their mimics were reported to be the presence of skull erosion and absence of a dural tail [5]. However, these findings were applicable to the present meningioma case. Therefore, it may be recommended that angiomatous meningioma be involved in the differential diagnosis when a broad-based tumor is found in the cerebral convexity with extensive perifocal edema and erosion in the adjacent skull. The dural tail is an abnormal MRI finding commonly observed in meningiomas but is not specific to them. Because of the frequent presence of tumor invasion in dural tail, resection of the dura mater, as widely as possible around the tumor attachment, is recommended [1,3]. Further investigations are needed to elucidate why some meningiomas present with skull erosion instead of hyperostosis.

In conclusion, meningiomas arising in the frontal convexity may cause photophobia. Angiomatous meningioma should be considered when a broad-based tumor is found in the cerebral convexity accompanied by skull erosion and extensive perifocal edema.

**Author contributions**

All the authors contributed equally to this study.
Ethical standards

We declare that the present study has been approved by the institution’s guidelines for human research and was performed in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki and its later amendments.

Patient consent

The patients documented in the manuscript fully understood and agreed that the authors use the information materials of the patients in anonymized manner for possible publication in Radiology Case Reports.

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