

A rare case report: giant meningioma with extracranial extension



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ABSTRACT

Background: Meningioma is the most frequently occurring primary central nervous system (CNS) tumor, and it is typically found during brain imaging. It has a prevalence of 53 per 100,000 people. Extracranial extension in skull-base meningioma is rare and presents a female preponderance

Case Description: We presented an Indonesian female who underwent a brain MRI depicting atypical giant meningioma with extracranial extension. The surgery was conducted, and pathological examination confirmed as a typical meningioma WHO grade I. Recognizing and identifying the entity's unique imaging characteristics are paramount in successfully diagnosing and initiating the most effective treatment.

Conclusion: This report is expected to help diagnose giant meningioma, which is a rare case, thus reducing the occurrence of misdiagnosis.

Keywords: Extra-cranial extension, Meningioma, MRI.

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INTRODUCTION

Meningioma is the most frequently occurring primary central nervous system (CNS) tumor, and it is typically found during brain imaging. It has a prevalence of 53 per 100,000 people.^{1,2} Meningioma arises from the meningotheial or the arachnoid cap cells in the dura tissue. These tumors are generally located at the skull base, the skull vault, and other areas with dural reflections, such as the tentorium cerebelli, falx cerebri, and near the dural venous sinuses.³ World Health Organization (WHO) categorizes meningioma into three groups. Eighty percent of cases of meningiomas are benign tumors or grade I. Subsequently, meningiomas classified as grade II or atypical, which account for approximately 20% of all meningiomas, have a more aggressive behavior along with a higher risk of morbidity and recurrence. Furthermore, grade III meningiomas have a high mortality rate and comprise 1% to 2% of all meningiomas.⁴

Extracranial extension in skull-base meningioma is rare and presents a female preponderance. Extracranial growth may be the main symptom, or it may be hidden as the tumor silently grows through the

base of the skull and into the sinuses. Extracranial growth can result in facial deformity and distortion of cranial shape, causing aesthetic concerns in patients long before neurological symptoms develop. Surgical treatment of such tumors can be challenging as it requires creativity and improvisation on the part of the neurosurgeon during surgery.⁵ Studies on meningiomas with extracranial extension are rare and are mostly presented as case reports.⁶ In this case, we described a giant extracranial extension of intracranial meningioma with histopathology findings of meningotheial meningioma type.

CASE DESCRIPTION

A female patient, 39 years old, visited our hospital with a lump on her right forehead that was first noticed seven years ago. Over the past year, the lump has grown to approximately 15 cm, but the patient did not seek medical attention as she did not experience any pain. Recently, she has been complaining of headaches for the past three months. No account of seizures, loss of consciousness, limb weakness, or vision changes in either eye or hearing loss was reported. The patient has no history of head trauma or oncological issues but

has used hormonal contraception for two years.

A large, well-demarcated solid lesion measuring 14.8 x 16.0 x 13.8 cm at the extra-axial of the right frontotemporal base skull was shown during an MRI scan of the head. The right frontotemporal bones have been damaged and have expanded into the soft tissue of the right frontotemporal region. The mass with surrounding perifocal edema has pressed on the midbrain, pons, and right ventricle, resulting in a midline shift to the left side. The mass has also extended into the right sphenoid sinus, filling the right orbital cavity, and pressed on the right eyeball, resulting in proptosis. In contrast, the right opticus nervus is still preserved (**Figure 1**). Gadolinium intravenous administration showed heterogeneous contrast enhancement mass with prominent vascular, in which the feeding arteries from the branch of the right external carotid artery (ECA) and middle cerebral artery (MCA) were seen.

DISCUSSION

The incidence of meningioma in women relative to men is approximately 2:1, with the most dominant age group being individuals aged between 35-44

years.⁷ Meningioma can be caused by a variety of variables, including radiation, work-related exposure to pesticides and herbicides, obesity, family history, molecular characteristics, and hormones most of the time.^{1,7} Patient's hormonal contraceptive use history may increase meningioma risk.

The skull base (43.51%) of meningioma is the most frequent location, with the sphenoid and middle cranial fossa predisposition.¹ Meningiomas usually manifest with broad types of presentation; however, the location and compression of adjacent brain and vascular structures can lead to focal neurologic deficits (including cranial nerve deficits).⁸ Skull-base meningiomas present more often with neurological deficits, and non-skull base meningiomas are more likely to present with seizures.¹ Symptoms frequently seen include headache, focal cranial nerve deficit, seizure, cognitive change, weakness, dizziness, gait change, pain/sensory change, proptosis, syncope, and asymptomatic.¹ Our patient had not felt any symptoms for the last seven years except for a lump on her forehead. The lump has progressively grown in the previous year, causing proptosis and headaches.

CT can identify calcification and hyperostosis in meningiomas, but MRI is better for tissue and edema analysis. Meningiomas appear hypointense to isointense on T1 and isointense to hyperintense on T2. Moreover, their contents show no restriction in the diffusion-weighted sequence, comparable to CSF, and no surrounding edema. A well-defined border, homogenous enhancement, calcification, and a dural tail are commonly associated with benign meningiomas. Although dural tails are not pathognomonic for meningiomas, they are a common characteristic in 72% of cases. Furthermore, tumor extension over skull base foramina, intra-tumoral necrosis, cortical invasion, intertumoral cystic alterations, edema volume, and ADC reduction (apparent diffusion coefficient) are associated with more aggressive and higher-grade meningiomas.³ Compared to normal brain tissue, magnetic resonance spectroscopy usually reveals elevated peaks for choline and alanine and lower peaks

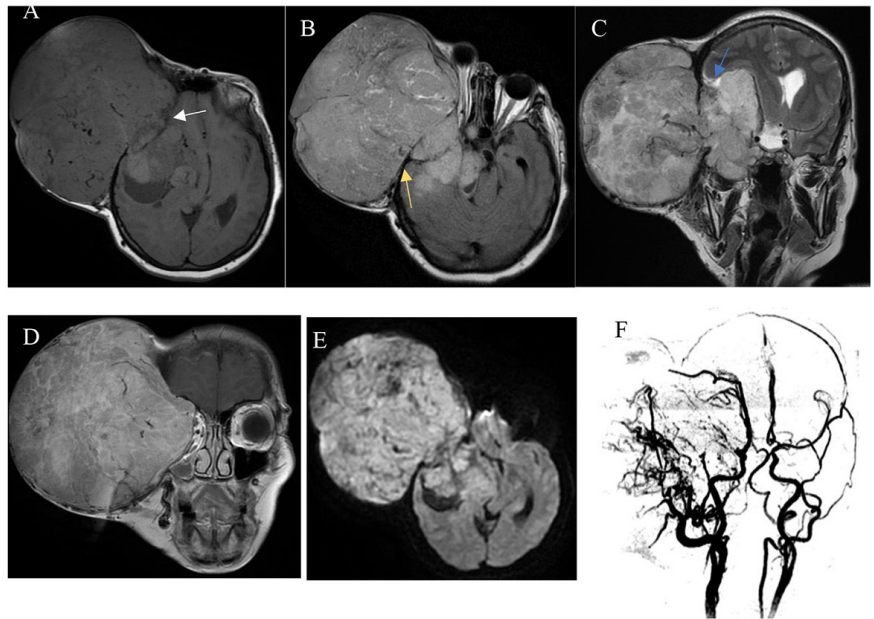


Figure 1. (a) T1-weighted axial plane image showed the extra-axial solid mass at right frontotemporal convexity with a well-defined border low signal destruct bone and extends to extracranial (white arrows). (b) T2Flair axial plane and (c) T2-weighted coronal plane showed slightly high signal intensity (yellow arrows) and also extended into the right sphenoid sinus, filling the right orbital cavity and pressing on the right eyeball causing proptosis, with right opticus nervus still preserved with cerebrospinal fluid (CSF) cleft sign (blue arrow). (d) T1-weighted coronal plane with contrast injection showed substantial heterogeneous contrast enhancement (red arrows). (e) Diffusion-weighted imaging (DWI) showed restricted lesions. (f) Magnetic resonance angiography (MRA) showed feeding mass from the right MCA and ECA branches.

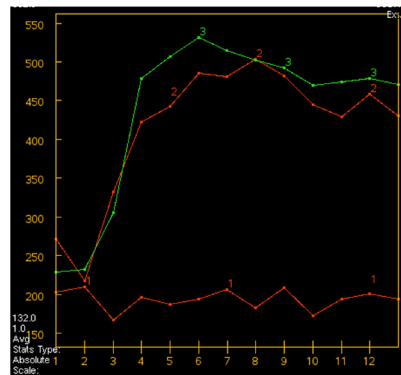
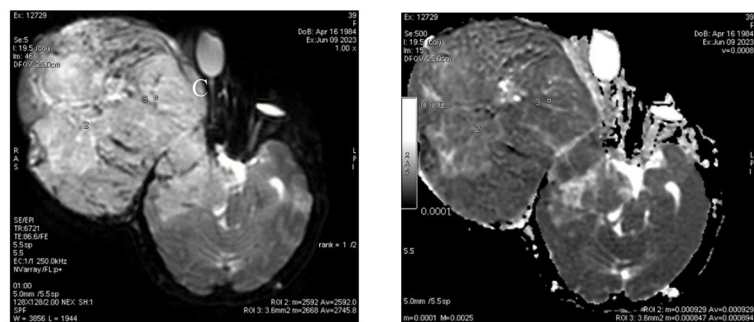


Figure 2. DWI shows slightly restricted diffusion with apparent diffusion coefficient (ADC) value $0.894 - 0.929 \times 10^{-3}$ (a, b), Dynamic contrast-enhanced (DCE) MR perfusion of time signal intensity curve (TIC) type 4 (washout phase)

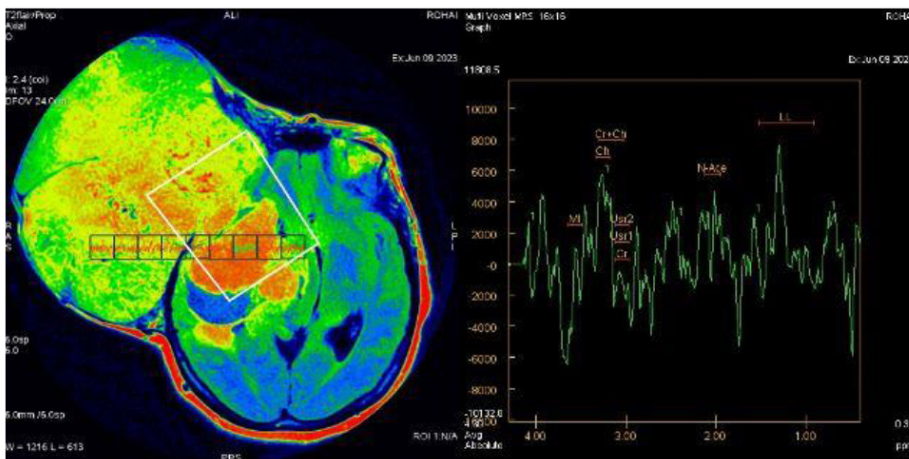


Figure 3. Spectroscopy on the lesion reveals elevated peaks for choline and lactate and lower peaks for creatinine.

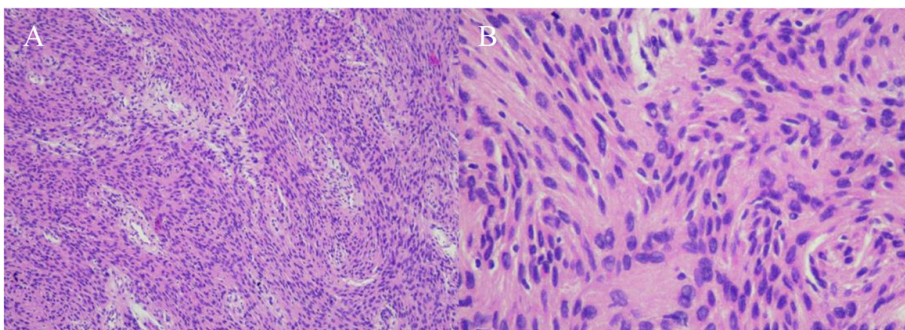


Figure 4. Histology of a tissue section partially covered by the skin's epidermis with a tumor growth arranged in lobules and small syncytia within the connective tissue. Consists of meningeothelial cell proliferation with round nuclei, smooth chromatin, sufficient cytoplasm, and some with intranuclear pseudo-inclusion. Mitosis is difficult to find. No signs of malignancy were evident. Magnification (a) x100, (b) x400.

for creatinine and N-acetyl aspartate. As in necrotic tissue, atypical meningiomas exhibit a lactate peak.⁹

Based on the literature, when a meningioma is symptomatic or growing larger, complete surgical excision of the tumor, surrounding dural attachment, and affected bone is advised. Endovascular embolization is not typically a recommended treatment option. However, it may be considered in specific cases as a preoperative measure.⁸ Our patient had undergone a complete surgical excision of the tumor with prior embolization, and then histopathological examination was conducted on the excised tissue. Based on histology or subtype, the three malignancy categories (CNS WHO grades 1-3) in WHO 2021 are equivalent to those in WHO 2016. The

malignancy grading of meningiomas has been modified to a within-tumor grading, regardless of subtype, and they have now considered a single tumor type with 15 subgroups. In grade I, there are <4 mitoses per 10 high power fields (HPFs), grade II 4-19 mitoses per 10 HPFs, and grade III >20 mitoses per 10 HPFs.^{1,10}

In this case, we found a presentation similar to atypical meningioma with right frontotemporal convexity with giant extracranial extension, heterogenous contrast enhancement, reduced ADC value, and elevated lactate intratumoral which means intra-tumoral necrosis. However, there is no established ADC threshold to differentiate between benign and high-grade meningiomas, and ADC cannot accurately predict proliferation potential and cellularity in meningiomas.¹¹

Meningiomas that expand outside the cranium are uncommon, accounting for only 1-2% of all meningiomas.⁶ Nevertheless, histopathology shows that there is no mitoses per 10 HPFs and brain cell invasion, which consists of meningeothelial cell proliferation.

CONCLUSION

Meningiomas with giant extracranial extend are very rare. The appearance of an atypical meningioma could be a typical meningioma based on histopathological findings that correspond to WHO grade 1, typical meningioma with meningiothelioma type.

CONFLICT OF INTEREST

In this review article, there is no potential conflict of interest.

FUNDING SOURCE

Not applicable

AUTHOR CONTRIBUTION

All authors contributed to this article.

CONSENT FOR PUBLICATION

Written informed consent was obtained from the patient to publish this case report and accompanying images.

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