

# A rare case of paediatric meningioma masquerading as intra-axial lesion

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## SUMMARY

**Meningiomas are neoplasm arising from meningoepithelial cells, most commonly in the fifth to sixth decade of life. Meningiomas are rare in paediatric population, accounting for 0.4-4.1% of all paediatric tumours and less than 3% of paediatric brain tumours. However, meningiomas represent the most common dural based tumours in children. We describe a rare case of paediatric fibroblastic meningioma within the left middle cranial fossa masquerading as an intra-axial mass lesion. Our discussion will be centred on atypical features of paediatric meningiomas and differential diagnosis of extra-axial mass lesion in the paediatric population.**

## CASE HISTORY

A 5-year-old boy with no past medical issue presented with a 6-month history of seizures, associated with nausea and headache. There was no focal neurological deficit on presentation.

CT scan was performed and shows a well-defined hyperdense mass within the left middle cranial fossa in the left temporal lobe with minimal perilesional oedema (Fig. 1A). Due to the lack of dural attachment, the mass was initially thought to be an intra-axial lesion.

MRI was performed for further characterization. The lesion is predominantly T2w hypointense and shows no evidence of intralesional haemorrhage or calcification. The presence of CSF cleft between the lesion and adjacent left temporal lobe confirms an extra-axial location of the tumour (Fig. 1B). Homogeneous enhancement is noted with prominent vessels along its superior and posterior margins. Of note is also circumferential encasement of the M2 segment of the left middle cerebral artery (MCA) (Fig. 1C).

The patient eventually underwent surgical excision of this lesion. Intra-operatively, the tumour was noted to be of hard consistency with moderate vascularity. It did not have a definite dural attachment and was adherent to the left temporal lobe. The main bulk of the tumour was successfully removed. A post-operative MRI was performed and showed residual tumour encasing the left MCA.

On histological examination, bland spindle cells were seen with fibrous tissue, in keeping with fibroblastic meningioma. Glial fibrillary acidic protein (GFAP) immunostain was negative, excluding a tumour of glial origin.

Post-surgical recovery was uneventful until post-operative day-5 when the patient developed an acute left striatocapsular infarct, likely due to tumour encasement of the left proximal MCA. Patient was eventually discharged, and subsequent MRI three months later demonstrates an increase in size of the residual tumour (Fig. 2A and 2B). The patient's family was not keen for radiation therapy and opted for expectant management.

## DISCUSSION

Meningiomas are rare in the paediatric population, accounting for 0.4-4.1% of all paediatric tumour<sup>1</sup> and less than 3% of paediatric brain tumours.<sup>2</sup> Paediatric meningiomas are known to show atypical features in comparison to their adult counterpart. A case series reported by Arizhavagan et al.,<sup>3</sup> found intraventricular location to be the most common within the paediatric population. In our case, the tumour location within the left middle cranial fossa is relatively rare.

Paediatric meningiomas are also known to mimic intra-axial lesion because they often lack a definite dural attachment,<sup>2</sup> as in our case. However, the presence of a CSF cleft sign and cortical buckling on subsequent MRI study as well as circumferential encasement of the left M2 segment point towards an extra-axial lesion. There was only minimal parenchymal vasogenic oedema despite the large size of the lesion, again unusual for an intra-axial lesion.

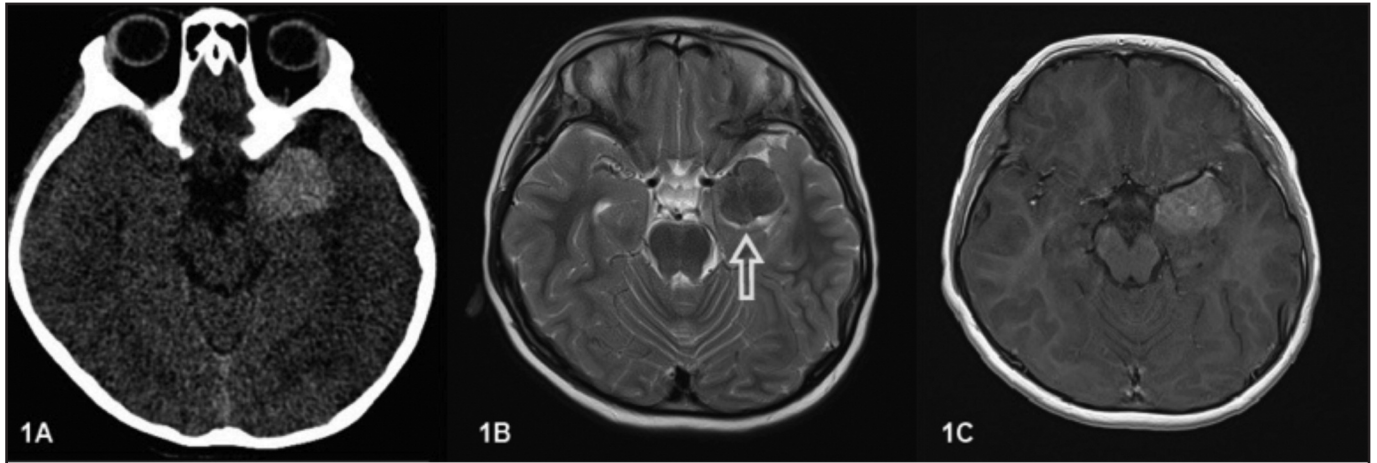
Differential diagnoses of an extra-axial lesion within the paediatric population include meningioma, haemangiopericytoma and primary dural lymphoma. Haemangiopericytomas are usually lobulated with heterogeneous enhancement, unlike the lesion described in our case which demonstrates a smooth margin and relatively homogenous enhancement. In addition, haemangiopericytomas are often associated with erosion of adjacent osseous structure,<sup>4,5</sup> which is absent in our case. Primary dural lymphoma, on the other hand, is usually associated with marked vasogenic oedema of adjacent brain parenchyma.<sup>4</sup>

In terms of management, meningiomas are reported to be more aggressive in the paediatric age group and clinical outcome is less predictable.<sup>2</sup> In our case, patient developed an acute left striatocapsular infarct during the post-surgery period, which may be related to the presence of vessel encasement. In addition, there is significant increase in size of residual tumour three months after the surgery, an

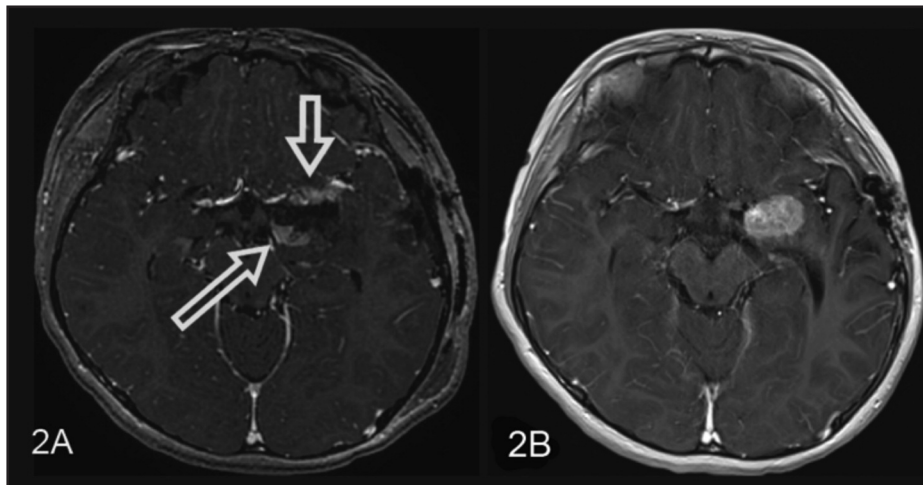
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**Fig. 1:** 1A: Non-contrast CT study shows a hyperdense mass within the left middle cranial fossa with no definite dural attachment. This was initially thought to be an intra-axial mass. 1B: Axial T2-weighted image shows a well circumscribed hypointense lesion within the left middle cranial fossa, separated from the left temporal lobe by a CSF cleft (arrow). 1C: Axial T1-weighted post contrast image shows encasement of the left middle cerebral artery by the homogeneously enhancing lesion.



**Fig. 2:** Contrast-enhanced T1-weighted image acquired 3 days after surgery shows residual enhancing tumour along the left MCA (short arrow), as well as along the posteromedial margin of the surgical cavity (long arrow) (Fig. 2A). The residual enhancing tumour shows increase in size in a follow up study performed three months after surgery (Fig. 2B).

unusual course for a WHO grade 1 meningioma. As meningiomas are uncommon in the paediatric population, there are no large multi-institutional studies to ascertain the standard treatment regime. Radiation therapy is generally avoided in the paediatric population unless deemed clinically necessary. It is also unclear if adjuvant radiation therapy will affect the overall risk of eventual malignant transformation.<sup>2</sup>

**CONCLUSION**

Paediatric meningiomas are rare and often demonstrate atypical features, rendering the radiological diagnosis challenging. One needs to be familiar with these features to clinch the correct radiological diagnosis. Knowledge of the differential diagnoses of an extra-axial mass lesion within the paediatric population as well as their radiological appearances are also equally important.

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