Polish Annals of Medicine

Journal homepage: https://www.paom.pl

Case Report

Neuroglial heterotopia in a neonate: The unusual suspect

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ARTICLE INFO

Article history
Received: May 5, 2024
Accepted: November 24, 2024
Available online: 11 December 2024

Keywords
Glioma
Neonate
Neuroglia
Neuroglial heterotopia

Doi
https://doi.org/10.29089/paom/196398

ABSTRACT

Introduction: Soft tissue masses in neonates often attract a wide variety of differential diagnosis, with hemangiomas and other types of vascular malformations being the most common benign lesions. However, in this mysterious case, neuroglial heterotopia, a rare congenital anomaly characterized by the presence of mature neural tissue outside the cranial cavity, was discovered instead. Extracranial presentations of these lesions are rarely encountered.

Aim: This case report aims to create awareness regarding the possibility of neuroglial heterotopia masquerading as a benign tumor in neonates and to highlight the importance of examination and confirmation with radiological investigations, to guide further management.

Case study: We describe a 3-month-old infant who presented with a rapidly growing soft, non-tender mass on the right side of her head that she had since birth. Radio imaging showed multiple cystic lesions located extra-temporally, which may represent lymphatic malformation, and excluded the chance of branchial cleft cyst as no intracranial sinus tract was seen.

Results and discussion: Surgical excision of the mass was performed, and histopathological evaluation confirmed the presence of mature glial tissue consistent with neuroglial heterotopia.

Conclusions: Extranasal neuroglial heterotopias are a distinct clinical entity. Accurate diagnosis, primarily using imaging, is essential to differentiate from other soft tissue masses in neonates and to guide management. Surgical excision remains the treatment of choice, and histopathological examination is required to confirm the diagnosis.

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1. INTRODUCTION

Soft tissue masses in neonates often attract a wide variety of differential diagnosis, with haemangiomas, hamartoma, and other types of vascular malformations being the most common benign lesions. Neuroglial heterotopia (NGH), also known as nasal glioma, is a rare congenital anomaly characterized by the presence of mature neural tissue, mainly in the nasal region. However, the presence of this tissue is also seen extranasally, which is rare.[1]

2. AIM

This case report aims to create awareness regarding the possibility of NGH masquerading as a benign tumor in neonates and to highlight the importance of examination and confirmation with radiological investigations, to guide further management.

3. CASE REPORT

A 3-month-old infant, born to non-consanguineous parents, presented with a rapidly growing soft, non-tender mass on the right temporal region, extending into the right cheek, that was present since birth. Antenatal scans were unremarkable, the delivery was uneventful, and there was no history of trauma before the discovery. The mass grew progressively from 1 × 1 cm to 6 × 6 cm over 3 months.

The mass is soft in consistency and had a smooth surface overlayed by normal skin. It was non-pulsatile and was fixed at the base. The mass also did not expand during straining or crying. The initial differential diagnosis included branchial cleft cyst, lymphatic malformation, cephalohematoma, and caput succedaneum.

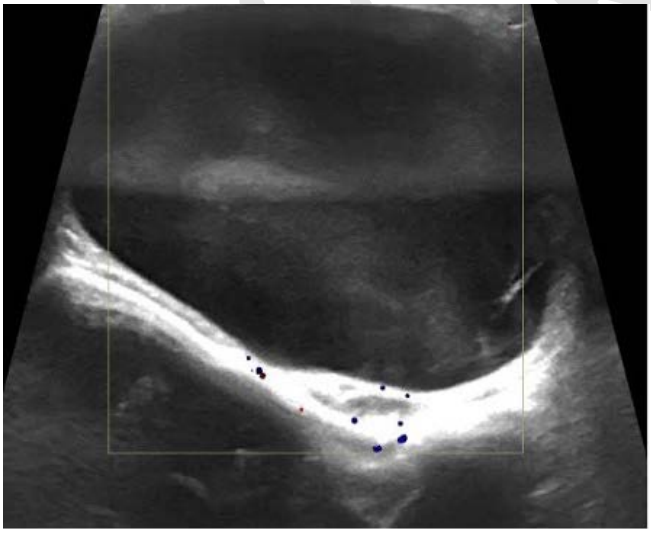

Figure 1. Ultrasonogram of the lesion: A well-defined anechoic lesion occupying the subcutaneous region. Heterogenous echogenic content and a few thin septations is seen within the lesion.

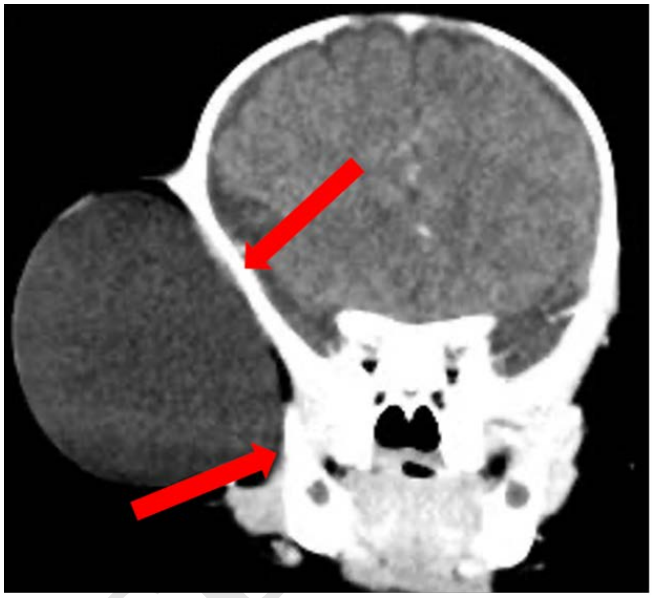

Figure 2. Coronal view of the CT. No intracranial communication is seen. A mass effect is seen, causing the remodelling of the right zygomatic and temporal bone (red arrows).

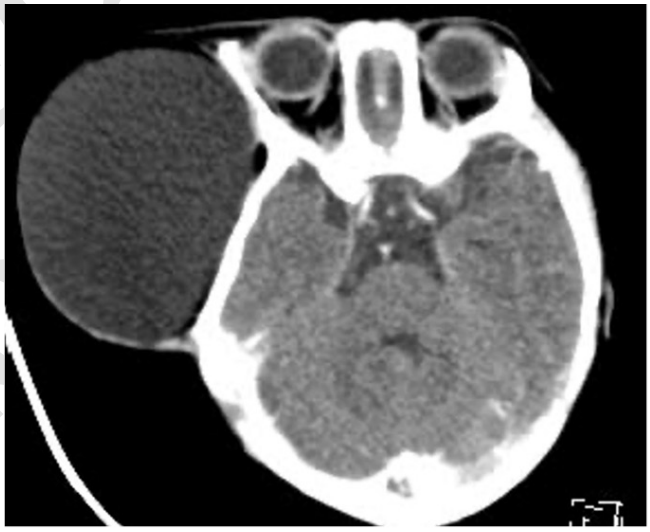

Figure 3. Axial view of the CT.

The child was subjected to an ultrasonogram and a computerized tomography, which revealed a cystic mass over the right temporal region, sized 6.6 × 5.4 × 6.1cm (AP × W × CC), causing a mass effect onto the right temporal and zygomatic bone, projecting a mass effect onto the adjacent calvarium causing remodelling of the right zygomatic and temporal bone. No intracranial communication was observed (Figure 1–3).

Surgical excision was done by successfully removing the mass with the capsules intact, and a no-tension skin closure of the defect was done (Figure 4–7). The mass was sent for a histopathological examination (HPE). HPE of the mass unexpectedly reported mature glial tissues, consistent with NGH. Post-operatively, the child had an indentation over the right temporal region, which resolved over time (Figure 8).

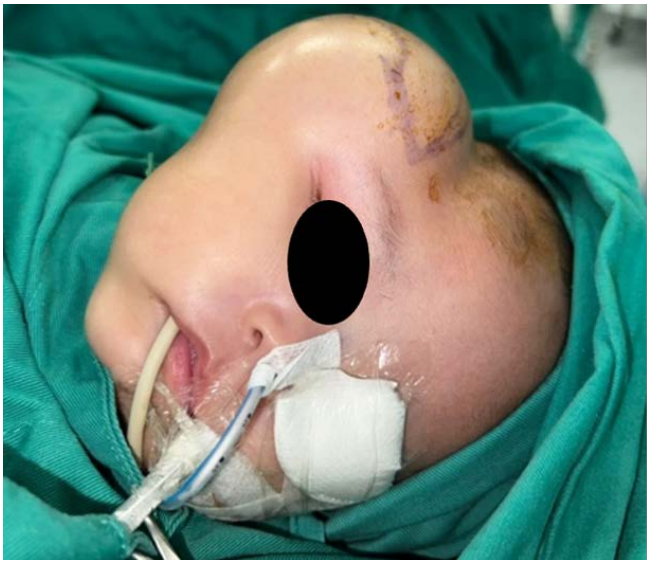

Figure 4. Pre-excision: Front view.

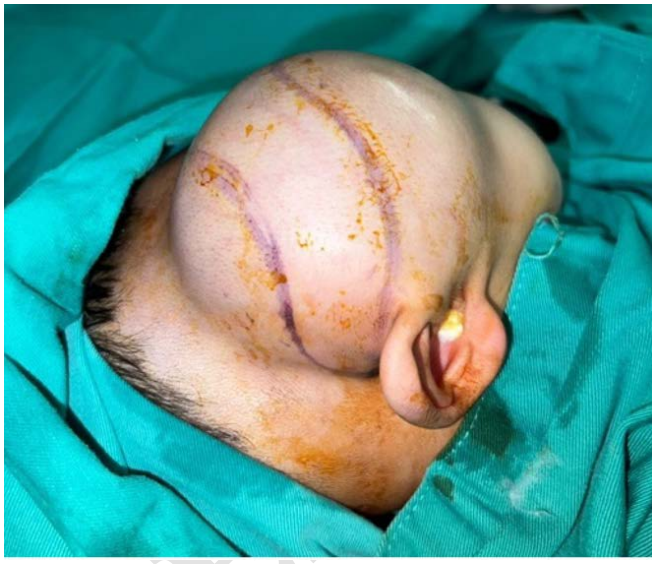

Figure 5. Pre-excision: Lateral view.

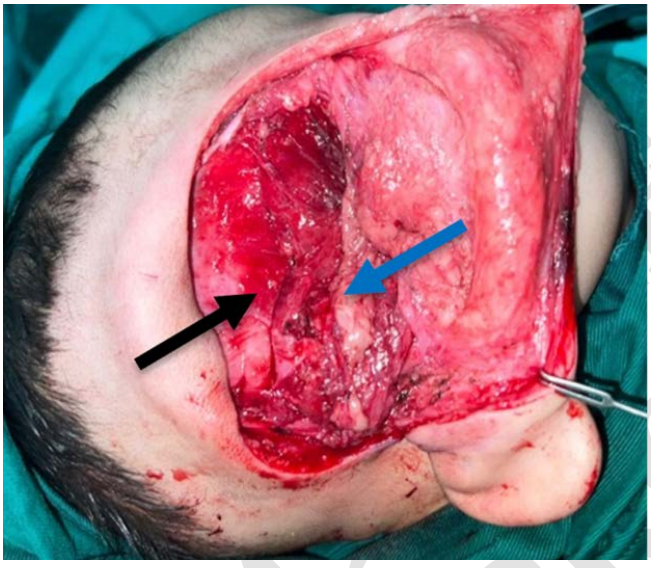

Figure 6. Post-excision: Mass completely removed, indentation over the temporal bone (black) and zygomatic bone (blue).

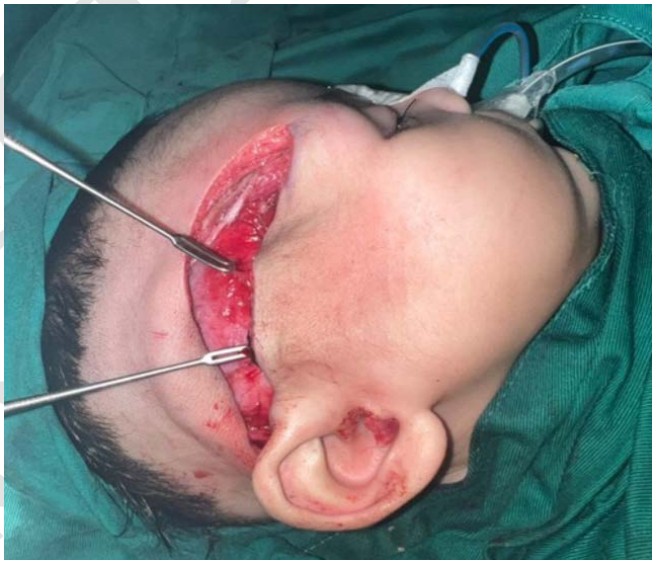

Figure 7. Post-excision: Primary closure possible with no skin tension.

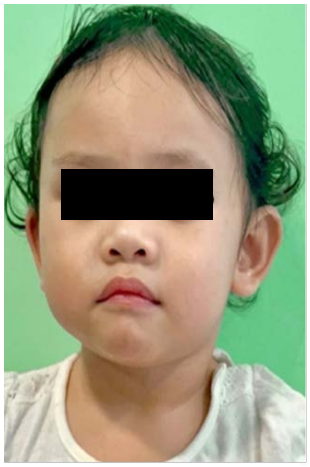

Figure 8. Post-excision (1 year): Front view with no deformity at the previous surgical site.

4. DISCUSSION

Extracranial presentation of mature cerebral tissue is known as nasal glioma or NGH. It is usually encountered during infancy to early childhood.[1] Reid first discovered NGH in 1852, and the term was established by Schmidt in 1900.[1,2] In 1955, the term 'heterotopic brain tissue' was introduced by Lee and McLaurin to describe a lesion containing mature cerebral tissue with no communication to the intracranial cavity.[3]

The nose and the nasopharynx are the most common sites where this rare entity is discovered. However, extranasal presentations do exist. Since intranasal presentations are more common, they are also known as nasal glioma, and

the incidence is 1 : 20,000-40,000 live births.[4] Nasal gliomas may coincide with other anomalies, which include micrognathia, cleft palate, congenital heart defects, and Pierre Robin syndrome. Extranasal sites include the orbit, palate, submandibular region, and overlying the spine.[5]

NGH and nasal encephaloceles, or dermoids, share a similar embryologic development. Two main hypotheses regarding the pathophysiology of NGH have been proposed. The first hypothesis states that at the embryonic dural diverticulum retraction stage, connections to the subarachnoid space are segregated, resulting in remnants of neuroglial tissue. The absence of subarachnoid communication extracranially distinguishes NGH from anterior encephaloceles.[4,6] The second hypothesis suggests that NGH develops from neuroectodermal displacement at the initial stage of embryogenesis, which differentiates into NGH. Radio imaging for this patient showed no evidence of intracranial association.[7]

NGH is present in males and females at a ratio of 3 : 2, with many patients diagnosed and treated by 12 months, including the case in point. Intranasal locations of NGH are reported to be the most frequent (45%), followed by extranasal presentations (36%) and mixed types of NGH (19%).[1]

Evaluation of a congenital mass may require a nasal endoscopy if the location is near the midline. Radio imaging techniques are also used to evaluate the involvement of the intracranial space and assess bony defects surrounding the tumor. Other commodities, such as magnetic resonance imaging (MRI) and X-rays, can also be performed. In this case, an ultrasonogram (USG) of the head and a non-contrasted computer tomography (CT) scan.[1]

The treatment of choice is to perform a curative surgical excision that paves the way for a histopathological diagnostic examination to confirm the diagnosis. Complete and early excision prevents complications, and better cosmetic results are achieved. Extension and localization of the NGH determine the surgical approach and may involve the neurosurgical team if a reasonable doubt arises or if a tract is seen leading to the skull base.[1]

NGH comprises of multiple elements, mainly astrocytes, followed by oligodendrocytes, neurons, functioning choroid plexus, and retinal cells interspersed in fibrous stromal tissue.[8]

The histopathology examination of the tumor in this case revealed a cyst wall lined by benign cuboidal to columnar ependymal cells, accompanied by mature neuroglial tissue with astrocytes in an abundant fibrillary background within the subjacent stroma. Positive yield using glial fibrillary acidic protein (GFAP), Masson's trichome stain, and S-100 protein immunohistochemistry are used to validate the presence of glial cells.[9] The GFAP and S-100 stain confirmed the diagnoses of NGH in this case.

The recurrence rate of 14% has been reported postoperatively, mostly recurring from 5 weeks to 11 months.1 Throughout the 1-year post-operative follow-up of this patient, no recurrence was seen.

5. CONCLUSIONS

(1) Extranasal NGH is a distinctly rare clinical entity.
(2) Diagnosis, primarily using imaging, is essential to differentiate from other soft tissue masses in neonates and to guide management.
(3) Surgical excision remains the treatment of choice and is paramount in diagnosing NGH.
(4) It is recommended that the tumor be investigated with magnetic resonance imaging to limit exposure to radio particles.
(5) Continuous follow-up is also required to monitor for recurrence.

Conflict of interest
None declared.

Funding
None declared.

Acknowledgement
We would like to thank all the surgeons and medical officers from the Department of Plastic Surgery, Hospital Kuala Lumpur, who helped in managing this case. We would also like to show our gratitude towards the Radiology Department and Histopathology Department, Hospital Kuala Lumpur, for allowing us to trace the results and verify the findings according to their expertise.

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