

Nasal glial heterotopia: A rare interdisciplinary surgical challenge in newborns

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Abstract

Nasal Glioma (NG) represents a rare congenital abnormality of the neonate, which can be associated with skull defects or even a direct communication to the central nervous system. MRI serves valuable information for differentiation from encephalocele, dermoid cyst and congenital hemangioma. Complete resection remains the treatment of choice. We present two cases of NG, which were both suspected during prenatal ultrasound and MRI. In the first case, postnatal MRI showed a transcranial continuity. Mass excision was performed and the defect was covered by a glabellar flap allowing a good cosmetic result. Postnatal MRI excluded a trans-glabellar communication in the second case. After surgical excision, the resulting skin defect was covered with a full thickness skin graft harvested from the right groin. In cases of NGs complete resection and cosmetic appeal-

ing results can be achieved and might necessitate a multidisciplinary approach.

Introduction

The term glioma refers to benign tumors originating from deposits of ectopic glial cells during embryogenesis. It probably derives, like encephaloceles, from a failed retraction of herniated brain tissue through fetal skull weaknesses during the fourth embryonic week.^{1,2} The typical localization is the facial midline, where they are similar to encephaloceles as they could also have an intracranial communication.³

With an incidence of 1:20,000 – 40,000 births Nasal Glioma (NG) are exceptionally rare benign tumors occurring in the proximal nasal region.⁴ The male-to-female ratio has been described with 3:2 with no familial predisposition.⁵ The tumors can either grow intra-nasally and therefore become evident due to the resulting airway obstruction or extra-nasally and then cause facial deformities.⁶ Intranasal localization is diagnosed in 30%, extranasal in 60% and combined forms in the remaining 10%.²

The lesions represent a diagnostic challenge, in particular because they can be easily confused with other pathologies such as dermoid cysts, encephaloceles as well as congenital hemangiomas due to their purple and bulky surface.⁷ The refinement of prenatal diagnostic tools, including sonography and MRI, frequently allows an accurate antenatal diagnosis.⁸ However, postnatal imaging is still necessary to exactly assess potential intracranial communication of the tumor.⁴ The optimal therapeutic option of NGs consists of meticulous surgical excision and accurate reconstructive surgery, aiming to avoid disfiguring results.⁹ In this report, we present two cases of NG successfully treated in a multidisciplinary approach. Our study adhered to the tenets of the Declaration of Helsinki. The families of both patients agreed to publish the reported photographic documentation.

Case 1

During second trimester anomaly scan at 21+4 weeks of gestation, a moderately hypoechoic solid tumor (9x11x13 mm) originating from the nasal root of the fetus was detected in a 21-years-old pregnant woman following natural conception (Figure 1a). No other anomalies were seen. Fetal MRI confirmed the diagnosis of an exophytic tumor and excluded involvement of the ocular region. However, a communication with the central meningeal structures could not be ruled out. Regular sonographic follow-up

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Key words: Nasal glioma; prenatal diagnosis; congenital hemangioma.

Conflict of interest: The authors declare no conflict of interest.

Availability of data and materials: All data underlying the findings are fully available.

Ethics approval and consent to participate: No ethical approval was required for this case report, because this article does not contain any studies with human participants or animals. Informed consent was obtained from the legal guardians of the patients included in this report.

Informed consent: The patients' guardians gave their written consent to use the patients' personal publication of this report and any accompanying images.

Received for publication: 2 September 2020.

Accepted for publication: 19 March 2021.

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La Pediatria Medica e Chirurgica 2021; 43:240

doi:10.4081/pmc.2021.240

examinations did not show an increase of the lesion's volume. The family refused further invasive diagnostic procedures.

In the 35th week of gestation a caesarean section was performed due to suspected intrauterine asphyxia following premature rupture of the fetal membranes. The female neonate presented with good general condition, a birth weight of 2.530 g and an Apgar score of 5/9/10.

She had an exophytic purplish broad-based mass occupying the right side of the nasal root, mimicking a congenital hemangioma. A feed-and-wrap MRI performed on 7th day of life showed an extension of the mass into the right nasal bone, which was distorted. Furthermore, a stalk-like connection to the dura was described traversing the foramen coecum without any communication with the subarachnoid spaces, in line with the diagnosis of a NG (Figure 1b). At follow-up examinations after discharge home the lesion did neither increase disproportionately in volume nor change its appearance (Figure 1c).

A multidisciplinary surgical approach including pediatric surgery, plastic surgery and neurosurgery was electively scheduled in the third month of life. The tumor was completely removed down to the

glabellar bone surface. The skin defect was covered with a glabellar flap. Histology showed the typical aspect of an ectopic glioma with nests of glial agglomerates surrounded by connective tissue.

On the 2nd postoperative day, the patient was discharged home and two months after surgery the flap was vital with an acceptable esthetical result (Figure 1d). Regular follow-up examinations were performed and three years postoperatively the girl has no signs of relapse.

Case 2

A 29-years-old woman at 29+6 weeks of gestation following natural conception was referred to our prenatal ultrasound unit with a suspected facial mass. Sonography confirmed a moderately hypoechoic solid mass (21x13x14mm) protruding from the nasal root of the fetus (Figure 2a). No further abnormalities were noted. Therefore, a fetal MRI was performed in the 30th week of the pregnancy, which showed a tumorous bulge over the glabella measuring

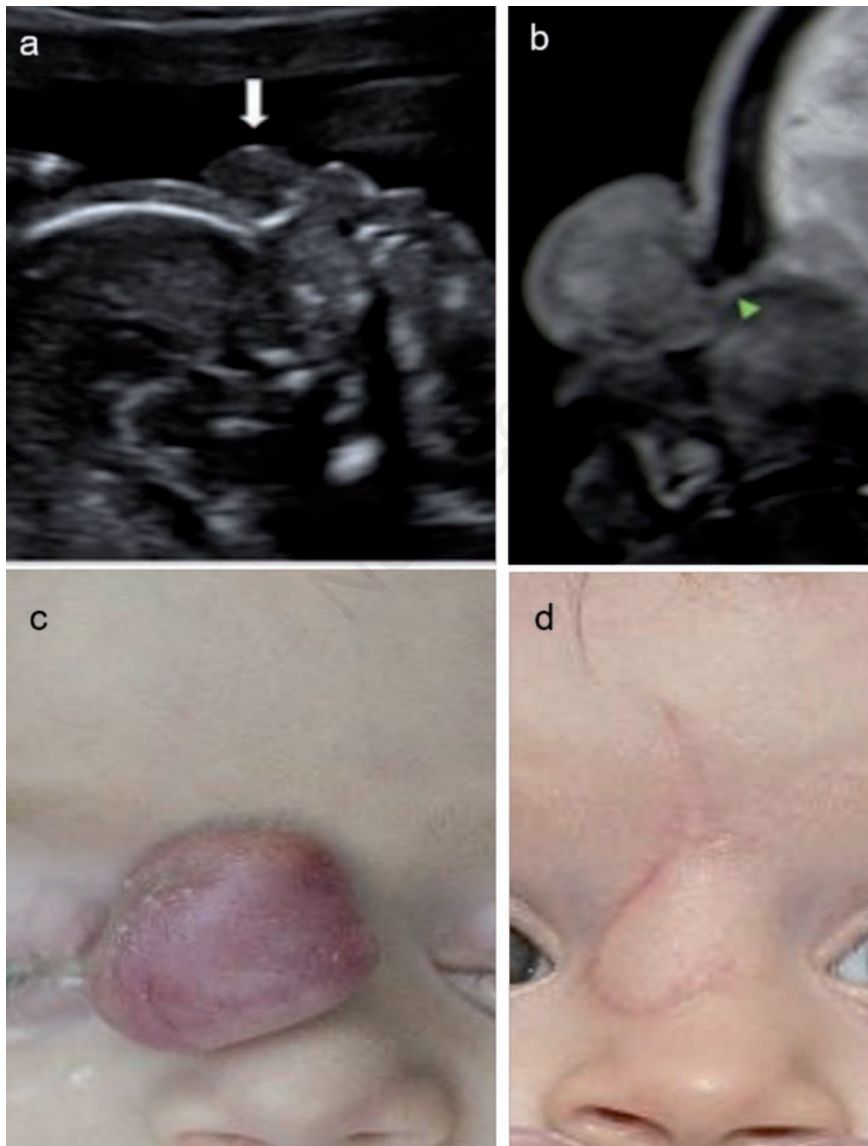


Figure 1. Case 1: a) prenatal ultrasound during the 21st week of pregnancy showing a round exophytic mass (white arrow) with 9x11x13 mm originating from the root of the nose; b) postnatal feed-and-wrap MRI showed a bony defect of the frontal bone and a fibrous stalk (green arrowheads) of the exophytic lesion traversing the foramen coecum; c) the exophytic purplish broad-based prenasal mass occupying the right side of the nasal root during preparation for surgery at 2 months of age; d) outcome two months after mass excision, the postoperative scar was visible but not disfiguring.

14x24x13 mm without any communication to the nervous system (Figure 2b). The family refused further invasive diagnostic procedures. Caesarean section was performed on patient's request with 39 weeks of gestation. At birth, the female neonate had a weight of 2,805 g and an Apgar score of 9/10/10. She had an uneventful post-natal adaptation. A purplish-red mass was observed in the area of the nasal root, resembling even in this case a congenital hemangioma (Figure 2c). The baby was followed up on an outpatient basis in the next months of life and the tumor showed no changes in its clinical appearance. Six months later the performed MRI excluded a bony defect of the frontal bone as well as communication with the central nervous system and validated the suspicion of a NG (Figure 2d).

The patient underwent complete surgical excision of the tumor in the 7th month of life. The resulting skin defect was covered with a full thickness skin graft harvested from the right groin (Figure 2e). Histological examination confirmed the diagnosis of a NG.

The postoperative course was uneventful and the patient was discharged home on the 2nd postoperative day. Follow-up examinations revealed a vital transplant with satisfying esthetical result. At the latest follow up eight months after surgery there was no sign of relapse.

Discussion

NGs - sometimes also referred to as encephalomas, nasal cerebral heterotopias or neuroglial heterotopia - are rare benign congenital tumors derived from a deposit of mature or dysplastic glial tissue.⁵ Most commonly, these lesions are localized extranasally on the nasal surface, followed by the nasal cavity or rarely the nasopharynx, frontal sinus, orbital or oral cavity. Although the lesions have been described everywhere on the nasal surface, the most common localizations is in the glabellar region like in our two patients.²

The etiology of these tumors still remains unclear. The most commonly accepted hypothesis attributes their occurrence to an ectopic trapping of herniated brain tissue due to premature closure of the metopic suture during the first phase of the embryonic period. This process would explain the presence of a communication with intracerebral structures in about 15% to 20% of cases.^{4,5}

Due to an increase of the number and quality of prenatal screening, congenital midline masses are usually diagnosed during routinely performed ultrasound examinations in the second

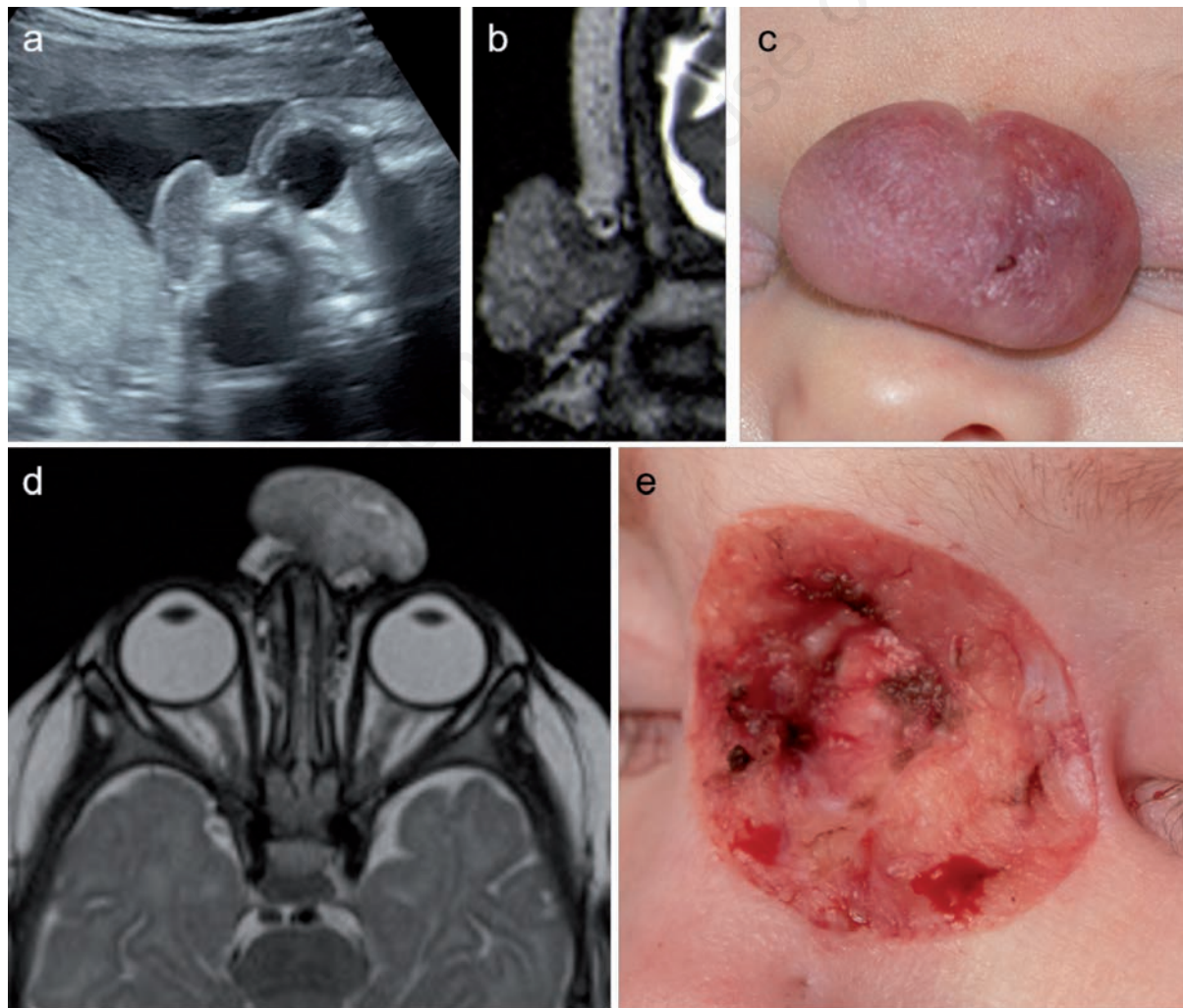


Figure 2. Case 2: a) prenatal ultrasound during the 29+6 week of pregnancy showing a hypoechoic solid mass (21x13x14mm); b) fetal MRI in a sagittal plane showing a well-demarcated exophytic mass on the glabella; c) a purplish-red mass was observed on the nasal root confirming the prenatal diagnosis; d) on the postnatal MRI the high-resolution T2 SPACE image ruled out connection to the intracranial structures; e) at surgery the wide skin defect was covered with a full skin transplant, harvested from the right groin.

trimester of pregnancy. Following detection of the tumor during sonographic evaluation, fetal MRI is generally recommended to aid differentiation from dermoid cysts, encephalocele or vascular anomalies.¹⁰ Due to their similar radiological appearance, the differentiation between NGs and encephaloceles is particularly challenging and correct diagnosis is only possible if an extracranial flow of cerebrospinal fluid can be visualized on MRI which was performed in both of our patients. Due to radiation exposure we did not consider performing CT examinations.

A defect of the skull bone is always present in encephaloceles but can also be found in up to 20% of gliomas.¹⁰ Anterior encephaloceles are more common in the southeast Asian regions and in Russia while posterior ones are more frequent in the western countries.⁵

Vascular tumors, most commonly Congenital Hemangiomas (CH), are reported as possible differential diagnoses of NGs.^{1,7} The differentiation between a NG and a CH which macroscopically may look exactly like NG may be difficult.¹¹ Even though an accurate description of the intrauterine growth of CH is lacking, they are supposed to increase in volume until the end of pregnancy. In our cases of NGs, we did not observe any disproportional increase in size of the mass during prenatal controls. Furthermore, NGs are hard incompressible masses with a homogeneous purple surface, whereas congenital hemangiomas have pale halo borders and a softer consistency.⁹ The incidence of CH is unknown but it is considered an extremely rare tumor. Therefore, it must be considered as diagnosis only if NG was excluded.

The differential diagnosis of a midline craniofacial mass should also include teratoma and retinoblastoma, which in our cases were excluded by the prenatal and postnatal MRI imaging.⁴

In cases of NGs, complete surgical excision is curative and allows pathohistological confirmation of the diagnosis. Early resection is advised in order to avoid the development of craniofacial deformities, particularly concerning the nasal architecture. Additionally, it also protects from visual impairment.⁴ Before surgery, it is of pivotal importance to rule out communication with intracerebral tissue, which occurs in 15%-20% of the cases.⁹ The misdiagnosis of even a tiny communication could lead to an incomplete closure of the meningeal sheets and a subsequent leakage of cerebral fluid and meningitis.⁴

A particular topic of debate is the modality of skin and soft tissue reconstruction after removal of the lesion.⁴ It is essential and challenging to combine an effective complete excision of the mass, the closure of the intracranial communication and an acceptable aesthetic result. In our cases we have chosen two different approaches for the two patients. In the first case, we performed a microscopically assisted closure of the fistulous brain communication, mass excision and then we covered the defect with a rotation flap from the central forehead. In the second case, we preferred a full thickness skin graft due to the larger skin surface removed, for which a rotational flap

would have caused too much tension. In both cases postoperative follow-up results were esthetically satisfying.

The diagnosis of NG is confirmed patho-histologically by the presence of neuroglial fibers intermixed with a fibrovascular connective tissue stroma. Histological confirmation of total extirpation of the tumor is also mandatory because recurrence in case of microscopic residuals is described in up to 10% of the cases.⁹

In conclusion, despite its rare occurrence, NGs should always be considered in the first place when detecting masses protruding from the nasal root of a fetus or neonate. A postnatal MRI is mandatory in order to exclude a communication with the central nervous system and to plan an interdisciplinary surgical approach allowing definitive treatment.

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