CASE REPORT
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A Case Report of Nasal Glial Heterotopia

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ABSTRACT

Nasal glial heterotopia (NGH), also known as nasal glioma, is a rare congenital malformation in which a mature glial cell mass is found in a location other than the central nervous system. These benign masses are divided into 3groups based on localization: the intranasal, extranasal, and mixed types. Here we present a case study of a young boy with an extranasal-located NGH who underwent surgical resection. Radiological examination is indispensable to differentiate between other midline masses (hemangiomas, dermoid cysts, and encephaloceles). Magnetic resonance imaging should exclude communication with the central nervous system. Computed tomography is used to verify the integrity of the anterior skull base. The proper management of NGH is a complete resection, often by a multidisciplinary approach involving otorhinolaryngology, neurosurgery, and reconstructive surgery.

Keywords: Children, nasal glial heterotopia, nasal glioma, neural malformation, surgical management

Introduction

Nasal glial heterotopia (NGH) is a rare congenital malformation in which brain tissue is found in an extracranial location. These benign masses are mainly located in the nose and nasopharynx and are therefore also referred to as "nasal gliomas."[1] Like nasal encephaloceles and dermoid cysts, NGH results from abnormal embryonic development. As the embryonic dural diverticulum retracts, remnants of neural glial tissue remain and lose their connection to the subarachnoid space.[1,2] These masses are divided into 3 groups based on location: intranasal (30%), extranasal (60%), and mixed (10%).[2,3]

Due to its rarity, diagnosis may be delayed, and the diagnostic workflow and treatment options may be unclear. Here we present a case of an infant boy with extranasal glial heterotopia. Informed consent has been obtained from the parents.

Case Presentation

A 5-week-old boy was referred to the consultation for pediatric ear, nose, and throat diseases because of a swelling at the level of the glabella. The nodule had been diagnosed prenatally by ultrasound, including prenatal magnetic resonance imaging (MRI), which showed a subcutaneous nodular solid structure. At birth, a pink, non-compressible mass was observed on the nasal dorsum that did not expand with crying (Figure 1). There were no other craniofacial anomalies. There were no complaints of obstructive nasal breathing or difficulty drinking.

The differential diagnosis of midline nasal masses can be narrowed down to 3 main entities namely, nasal dermoid cysts, nasal encephaloceles, and NGH. In addition, one should also keep in mind the possibility of a hemangioma or lymphangioma.[4]

A new MRI showed a subcutaneous nodular solid tissue structure at the level of the glabella, measuring 9 mm × 15 mm × 12 mm, which was locally not clearly demarcated from the nasal bone on the right side (Figure 2). There was no intracranial extension.

Surgical resection was planned at 3 months of age. During surgery, no defects of the (cartilaginous) nasal dorsum were found, and there was no evidence of an intracranial connection. Due to the size of the nodule and the resulting skin defect of approximately 1.5 cm, a glabellar flap reconstruction was performed by the reconstructive surgery team (Figure 3). Histologically, NGH was confirmed, and both the circumferential and deep section surface appeared positive. Due to the subcutaneous location, close follow-up was chosen. At 2 years

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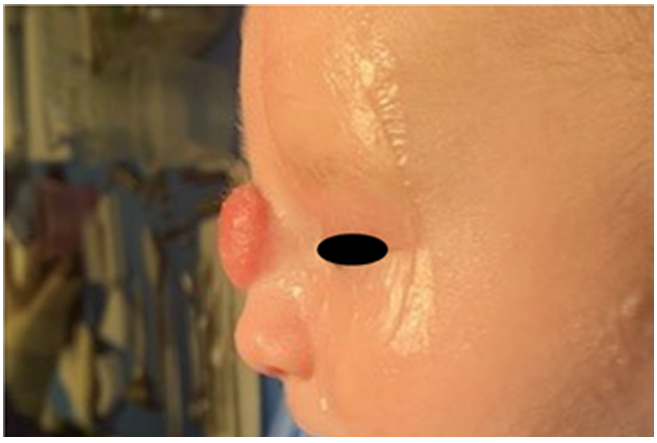

Figure 1. Pink, incompressible mass on the dorsum of the nose.

Figure 2. Magnetic resonance imaging shows a T1-isointense homogeneous soft tissue mass.

post-operatively, there was no clinical evidence of residual tissue or recurrence (Figure 4). This study was approved by Ethics Committee of Catholic University of Leuven. (Approval No: S67970, Date: July 15 2023).

Written informed consent was obtained from the parents of the patient who agreed to take part in the study.

Main Points

- Nasal glial heterotopia is a rare congenital malformation in which mature glial cells are found in a location outside the central nervous system. The major pitfall in the diagnosis is therefore not to include this entity in the differential diagnosis.
- Magnetic resonance imaging plays a crucial role in ruling out any communication with the central nervous system and in determining the surgical approach.
- Treatment consists of a complete surgical resection at a young age.

Figure 3. Surgical outcome.

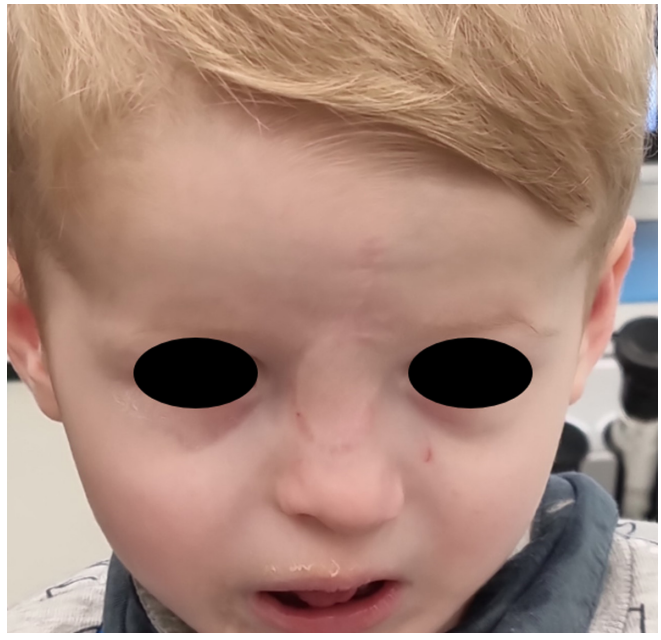

Figure 4. The result 2 years after surgery.

Discussion

Nasal glial heterotopia is a rare benign congenital malformation in which a mature glial cell mass is found outside the central nervous system.[5] The condition is more common in boys (the male to female ratio is 3:2) and has an incidence of 1 in 20 000-40 000 births.[4,6]

Diagnosis is made before the age of 3 in about 90% of patients. Most NGHs are visible at birth, but sometimes the diagnosis is delayed if the mass is too small to be clinically visible.[3]

The extranasal type presents as a hard, non-tumescent, non-pulsatile mass that gradually enlarges as the infant grows. This red or blue mass is usually located at the level of the glabella or nasomaxillary suture. Intranasal NGH presents mainly with

nasal obstruction, resulting in dyspnea, nasal discharge, open-mouth breathing, snoring, or feeding difficulties.

Further diagnostics include nasal endoscopy and imaging to determine the extent of the mass. An MRI is recommended as an initial investigation, mainly to rule out intracranial communication, using a submillimeter slice thickness and heavily T2-weighted sequence. A computed tomography is recommended for the evaluation of bony defect or as a navigational aid for endoscopic surgery. Biopsy is not recommended for midline congenital masses due to the risk of cerebrospinal fluid leakage, hemorrhage, and meningitis.

The extra-nasal type must be distinguished from nasal dermoid cysts, hemangiomas, and an encephalo(meningo)cele. A dermoid cyst usually feels softer, and the distinction is mainly based on histopathology, as NGH contains mature glial tissue and a dermoid cyst contains features of normal skin. The cyst wall consists of keratinized, stratified squamous epithelium and is filled with keratinous material.[2] A hemangioma presents as a soft, purplish-pink nodule that is compressible. An encephalocele is often translucent and, unlike an NGH, will increase in volume with weeping, Valsalva maneuver or compression of the jugular veins (positive Furstenberg sign). Before treatment can begin, an MRI must be performed to check for intracranial communication.

In the intranasal type, the differential diagnosis includes encephalo(meningo)cele and, in older children, nasal polyps.

Complete surgical resection is the recommended treatment. Resection is best performed at a young age to prevent complications such as meningitis and brain abscess in intranasal gliomas and cosmetic complications in extranasal gliomas.[4]

It is important that patients are followed up closely afterwards, as there is a recurrence rate of 4%-10% for NGH.[5] Follow-up for at least 1 year should be planned, with regular clinical review and repeat MRI if in doubt.[1]

Nasal glial heterotopia is a rare, benign congenital anomaly. The major pitfall in the diagnosis is therefore not to include this entity in the differential diagnosis. Magnetic resonance imaging plays a crucial role in ruling out any communication with the central nervous system and in determining the surgical approach. Computed tomography can be used to determine whether bone damage is present. Treatment consists of a complete surgical resection, preferably at a young age. One year of follow-up appears to be sufficient to exclude recurrence.

Ethics Committee Approval: This study was approved by Ethics Committee of Catholic University of Leuven. (Approval No: S67970, Date: July 15 2023).

Informed Consent: Written informed consent was obtained from the parents of the patient who agreed to take part in the study.

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