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Nasal Glial Heterotopia of the Nasal Root in a Child: A Case Report

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Abstract

Nasal glial heterotopia is a rare, benign congenital malformation characterized by ectopic glial tissue; its clinical diagnosis remains challenging due to overlap with other congenital midline masses such as encephalocele or dermoid cysts.

We report the case of a two-year-old boy presenting with a congenital mass at the root of the nose, which initially appeared to be a dermoid cyst on imaging. Preoperative MRI showed no intracranial communication. Intraoperatively, an approximately 1-cm nasal bone defect was identified, with no communication with the nasal cavity on endoscopic examination. Complete excision was performed via a lateral nasal approach. Histopathological examination revealed mature glial tissue, predominantly astrocytes, within a fibrous stroma. Immunohistochemical staining was positive for glial fibrillary acidic protein (GFAP) and S100, confirming nasal glial heterotopia. The postoperative course was uneventful, with no recurrence during follow-up.

Categories: Otolaryngology, Pathology, Pediatric Surgery
Keywords: child, congenital nasal mass, dermoid cyst, magnetic resonance imaging, nasal glial heterotopia

Introduction

Nasal glial heterotopia (NGH), historically referred to as “nasal glioma,” is a rare congenital malformation characterized by ectopic mature glial tissue outside the central nervous system [1,2]. It predominantly affects children and may present as an intranasal, extranasal, or mixed lesion, with extranasal forms commonly involving the nasal root or dorsum [1-3]. The clinical and radiological features of NGH may overlap with those of other congenital midline nasal masses, particularly dermoid cysts and encephaloceles [3-5]. Magnetic resonance imaging (MRI) is essential for assessing the lesion's extent and, most importantly, its relationship with the intracranial compartment [1,4,5]. However, imaging findings may not be sufficiently specific to establish the diagnosis, and histopathological examination remains definitive [1,6]. We report a pediatric case of extranasal NGH of the nasal root initially considered to be a dermoid cyst on clinical and radiological assessment. The diagnosis was established after complete surgical excision and histopathological and immunohistochemical examination. This case highlights the diagnostic overlap between NGH and dermoid cysts and emphasizes the role of MRI in excluding intracranial communication.

Case Presentation

A two-year-old boy with no significant medical history was referred to our otorhinolaryngology department for a congenital mass of the nasal root. The lesion had progressively increased in size. There was no history of volume fluctuation, recurrent infection, or inflammatory episodes. The child had no nasal obstruction, rhinorrhea, or other nasal complaints.

Clinical examination revealed a firm, well-circumscribed, non-tender mass involving the nasal dorsum and root, slightly deviated to the left and producing a visible deformity of the nasal root. The lesion was mobile and covered by normal-appearing skin without any visible cutaneous changes. There was no pulsation or cutaneous opening. The Furstenberg sign was negative. No intranasal abnormality was identified.

Ultrasonography showed an oval echogenic frontonasal lesion, initially considered suggestive of a dermoid cyst. Brain and facial magnetic resonance imaging (MRI) was subsequently performed. It demonstrated a 15 × 22 mm frontonasal lesion, hypointense on T1-weighted sequences and heterogeneously hyperintense on T2-weighted sequences (Figure *1*). There was no significant enhancement after gadolinium administration. Importantly, MRI showed no intracranial extension, no communication with the intracranial compartment, and no evidence of meningoencephalocele. No intranasal extension was identified. Based on these findings, a dermoid cyst was considered the most likely preoperative diagnosis.

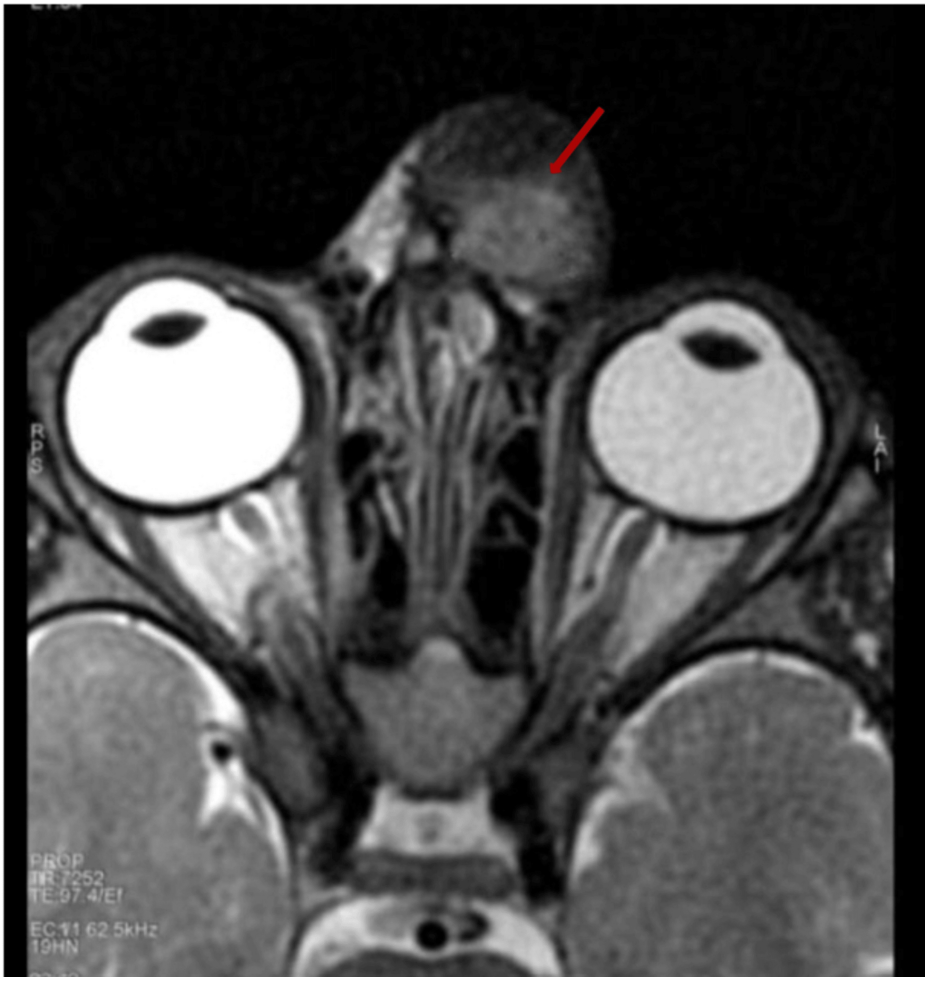


FIGURE 1: T2-weighted axial MRI showing a 15 × 22 mm frontonasal lesion with heterogeneous high signal intensity of the lesion (indicated by the red arrow)

Complete surgical excision was performed under general anesthesia. After local infiltration with lidocaine containing epinephrine, a 2.5-cm vertical incision was made on the left side, approximately 2 cm anterior to the medial canthus (Figure *2A*). The incision was deliberately positioned away from the lesion rather than directly overlying it, to minimize postoperative scar visibility.

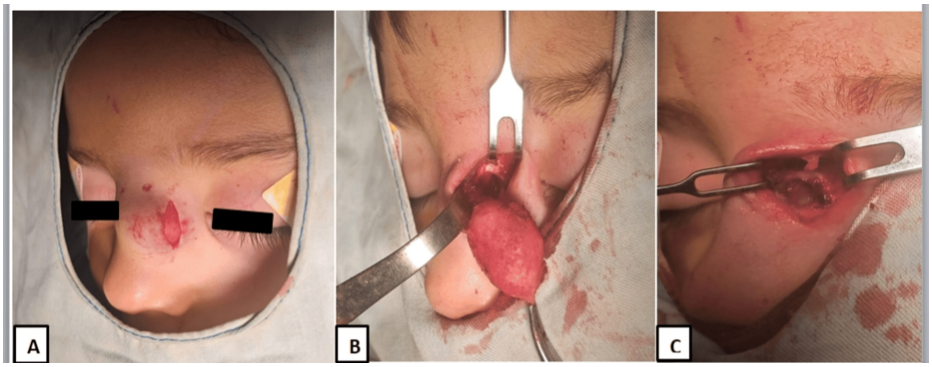


FIGURE 2: Intraoperative findings during surgical excision of the nasal glial heterotopia

(A) Surgical incision and exposure of the lesion. (B) Progressive dissection and mobilization of the mass. (C) Approximately 1-cm osseous defect of the nasal bone after complete excision.

Dissection was performed progressively from the surrounding tissues toward the lesion. The mass was relatively adherent to the surrounding tissues but remained well-circumscribed and encapsulated, with a clearly identifiable dissection plane. Careful dissection allowed complete removal without rupture (Figure *2B*). No vascular pedicle or fibrous tract was identified, and there was no macroscopic relationship with the dura mater. After excision, an approximately 1-cm round osseous defect was noted in the nasal bone (Figure *2C*). The absence of communication between the surgical site and the nasal cavity was verified intraoperatively using a 0° rigid nasal endoscope. Hemostasis was achieved, and the skin was closed using a continuous intradermal suture.

The postoperative period was uneventful. Histopathological examination of the excised specimen revealed a whitish, homogeneous nodule measuring approximately 2 cm. Microscopic examination showed connective tissue composed of collagen fibers associated with mature glial tissue consisting predominantly of astrocytes within a fibrillary stroma. No histological features of malignancy were identified. Immunohistochemical staining demonstrated positivity for glial fibrillary acidic protein (GFAP) and S100, supporting the glial nature of the lesion and confirming the diagnosis of nasal glial heterotopia (Figure *3*).

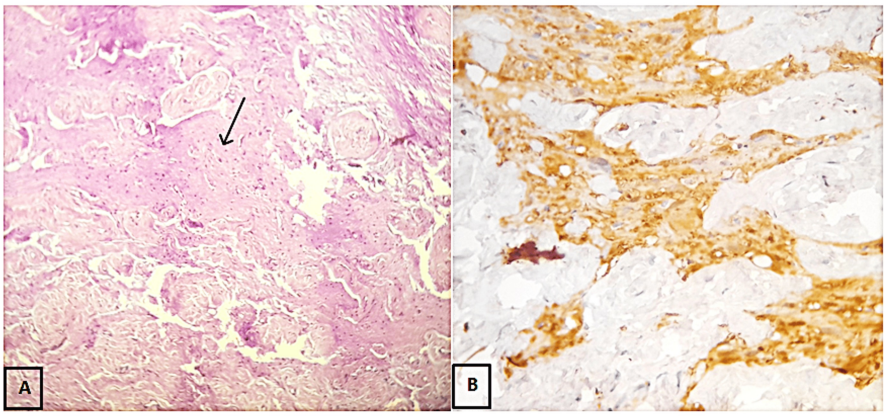


FIGURE 3: Histopathological and immunohistochemical features of nasal glial heterotopia

(A) Connective tissue containing mature glial tissue (arrow) (hematoxylin and eosin staining, ×20). (B) Glial cells showing positive immunoreactivity for glial fibrillary acidic protein (GFAP) (immunohistochemistry, ×40).

At nine months of follow-up, there was no evidence of local recurrence. The surgical scar was supple and slightly pigmented, with no functional complaints or nasal deformity.

Discussion

NGH is a rare, congenital developmental anomaly characterized by the presence of mature glial tissue

outside the central nervous system [1,2]. Its exact embryological origin remains uncertain, with abnormal sequestration of neuroglial tissue during craniofacial development among the principal proposed mechanisms [5].

NGH predominantly affects children. In the systematic review by Gallego Compte et al., which included 152 cases, most patients were younger than 18 years, with only 8% diagnosed in adulthood [1]. Similarly, Yan et al. reported that 90% of patients were diagnosed before the age of 3 years and that 60% were boys [2]. Our patient, a two-year-old boy, therefore corresponds closely to the typical demographic profile reported in the literature.

NGH can occur in extranasal, intranasal, or mixed forms [1,2,5]. Extranasal lesions are commonly located around the nasal root or dorsum and may present as firm, painless subcutaneous masses [1,2,6-9]. Our patient presented with an extranasal mass of the nasal root.

The differential diagnosis of a congenital nasal root mass includes NGH, dermoid or epidermoid cysts, and encephalocele [3,4,10,11]. These entities may have overlapping clinical characteristics, particularly when the lesion is firm, painless, and covered by normal skin. Dermoid cysts are an especially important differential diagnosis because they may occur along embryonic fusion lines and can occasionally be associated with a sinus tract or intracranial extension [3,12,13]. Conversely, encephaloceles may communicate with the intracranial compartment and consequently require specific surgical planning [4,10,11].

The Furstenberg sign may provide a useful clinical clue when evaluating congenital midline nasal masses. Enlargement or pulsation of the lesion with increased venous pressure suggests an intracranial connection and is classically associated with encephalocele [6,11]. However, a negative Furstenberg sign does not reliably exclude intracranial communication [6,11]. In our patient, the Furstenberg sign was negative, but MRI was nevertheless performed to accurately define the lesion's anatomical relationships.

MRI has a central role in the evaluation of congenital midline nasal masses because it provides excellent soft-tissue characterization and allows assessment of the relationship between the lesion and intracranial structures [1,3-5,10]. The principal objective of MRI in this setting is to identify or exclude intracranial extension or communication.

In our patient, MRI demonstrated a frontonasal lesion measuring 15 × 22 mm, without intracranial extension or communication, without evidence of meningoencephalocele, and without intranasal extension. These findings were essential for preoperative planning and excluded an obvious intracranial relationship.

However, MRI did not establish the definitive diagnosis in our patient, and the lesion was considered radiologically compatible with a dermoid cyst. This illustrates an important limitation of MRI: although it is essential for determining the anatomical relationships and intracranial extension of congenital nasal masses, its imaging characteristics may not be sufficiently specific to distinguish NGH from other benign congenital lesions [3,4,14,15]. Shafqat et al. similarly emphasized the variable radiological appearance of nasal gliomas and the potential difficulty of establishing the diagnosis on imaging alone [14].

CT provides complementary information because of its superior spatial resolution for the evaluation of cortical bone and potential osseous defects or transosseous tracts [3,14]. This distinction is relevant in congenital midline nasal masses, as a bony defect may provide an anatomical route for intracranial communication [3,4]. In our patient, CT had not been performed preoperatively, and a small defect of the nasal bone was identified intraoperatively. It therefore represented an intraoperative anatomical finding rather than a diagnostic discrepancy.

Complete surgical excision is the treatment of choice for NGH and is generally associated with a favorable outcome [1,2,7]. Both external and endoscopic approaches have been described, with the choice depending mainly on the lesion's location, size, and extent [1,2].

In the pediatric series reported by Yan et al., treatment was successful in all patients, with recurrence occurring in only 3 of 60 (5%) [2]. In the systematic review by Gallego Compte et al., recurrence was reported in 14 patients, emphasizing the importance of complete excision and postoperative surveillance [1].

In our patient, complete excision was achieved through a lateral nasal approach. The incision was deliberately positioned away from the mass with the aim of optimizing the cosmetic result. The lesion was encapsulated and well circumscribed. Careful dissection permitted complete removal without rupture.

No pedicle, fibrous tract, or dural attachment was identified. Intraoperative examination with an endoscope confirmed the absence of communication with the nasal cavity, further supporting the lesion's isolated extranasal nature.

Histopathological examination remains the definitive diagnostic method for NGH. The characteristic finding

is mature ectopic glial tissue, predominantly composed of astrocytes within a fibrous or fibrillary stroma [1,5,16]. Immunohistochemistry is useful for confirming glial differentiation, particularly through GFAP expression, while S100 may provide additional support [16].

In our patient, histological examination demonstrated mature glial tissue composed predominantly of astrocytes within a fibrillary stroma. Immunohistochemical staining was positive for GFAP and S100, with no histological features of malignancy, confirming the diagnosis of NGH. Gundogdu et al. also emphasized that NGH may be overlooked histologically when the glial component is inconspicuous within a collagen-rich lesion, highlighting the value of appropriate immunohistochemical staining [15].

The patient had an uneventful postoperative course, with no complications, and remained recurrence-free at the nine-month follow-up. The scar was well healed, with a satisfactory cosmetic appearance. This favorable outcome is consistent with the generally good prognosis reported after complete excision of NGH [1,2,7]. The nine-month follow-up is reassuring but insufficient to assess long-term recurrence and cosmetic outcomes. Long-term surveillance is warranted to monitor recurrence, nasal contour, facial growth, and healing of the nasal bone defect, as well as to detect delayed complications.

Conclusions

This case emphasizes a practical diagnostic principle: MRI should be considered essential in the preoperative assessment of congenital nasal root masses because it defines lesion extent and, most importantly, evaluates intracranial communication. However, the absence of intracranial communication does not exclude NGH, and definitive diagnosis requires histopathological examination with appropriate immunohistochemical confirmation.

The combination of preoperative MRI, careful surgical dissection, intraoperative endoscopic assessment, and histopathological examination allowed safe complete excision in our patient. The favorable clinical and aesthetic outcome at nine months further supports complete surgical removal as the treatment of choice for localized extranasal NGH.

Additional Information

Author Contributions

All authors have reviewed the final version to be published and agreed to be accountable for all aspects of the work.

Acquisition, analysis, or interpretation of data: Rihab Lahmar, Ons Menif, Alia Zehani

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Concept and design: Maha Mejbri, Najeh Beltaief

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Supervision: Najeh Beltaief

Disclosures

Human subjects: Informed consent for treatment and open access publication was obtained or waived by all participants in this study. issued approval No. Written informed consent for publication of the case and accompanying clinical images was obtained from the patient's parents. **Conflicts of interest:** In compliance with the ICMJE uniform disclosure form, all authors declare the following: **Payment/services info:** All authors have declared that no financial support was received from any organization for the submitted work. **Financial relationships:** All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might have an interest in the submitted work. **Other relationships:** All authors have declared that there are no other relationships or activities that could appear to have influenced the submitted work.

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