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CASE REPORT

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Clinical and Therapeutic Approaches to Nasal Glial Heterotopia in Children

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ABSTRACT

Nasal heterotopias or gliomas are rare congenital malformations characterized by a nasal mass composed of heterotopic neuroglial tissue. They fall within the nosological spectrum of midline dysraphias, presenting a complex diagnostic and therapeutic challenge. A significant concern regarding these lesions is their potential intracranial extension, affecting the anterior base of the skull; MRI is required to rule out a connection to the central nervous system. Nasal glial heterotopia should be considered as a differential diagnosis of nasal masses in children. This report discusses the case of an infant with a paramedian nasal glioma and offers a clinical, radiological, and therapeutic analysis of this condition.

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1. INTRODUCTION

Nasal heterotopias, also known as nasal gliomas, are rare congenital anomalies manifesting as nasal masses comprised of ectopic neuroglial tissue. These entities are situated within the nosological context of midline dysraphias, presenting a complex diagnostic and therapeutic challenge. Approximately 60% of nasal gliomas exhibit an extranasal component, 30% are intranasal, and the remaining 10% display characteristics of both [1]. Externally, these gliomas form a firm mass, variably positioned from the tip of the nose to the glabella, predominantly lateralized, potentially leading to visual field anomalies or compression of the lacrimal pathways on the affected side [2].

A significant concern regarding these lesions is their potential intracranial extension, affecting the anterior base of the skull. Our presentation aims to delve into the clinical features, diagnostic methodologies, and therapeutic management of this pathology through detailed case studies.

2. CASE REPORT

This case involves an infant, born from a smoothly conducted pregnancy, with term delivery devoid of any incidents, who exhibited a left nasal tumor of a violaceous appearance. The tumor, non-hemorrhagic upon contact and adherent to the glabellar skin, showed no anomalies upon nasal endoscopy (Figure 1). Notably, a fissure was observed on the left nostril wing. Radiological evaluations, comprising CT scans and MRI, revealed a small pedunculated solid-cystic mass, the stalk of which was inserted projecting from the base of the nose, without any communication with the anterior cranial fossa, raising the differential diagnosis of an atretic nasal encephalocele (Figure 2).

Surgical intervention, conducted via an external approach, involved the excision of the formation followed by simple closure (Figure 3). For the fissure at the nostril wing, repair was undertaken using a rotation flap and the insertion of a silicone conformer for three months. Histopathological examination confirmed the diagnosis of glial heterotopia. The post-operative

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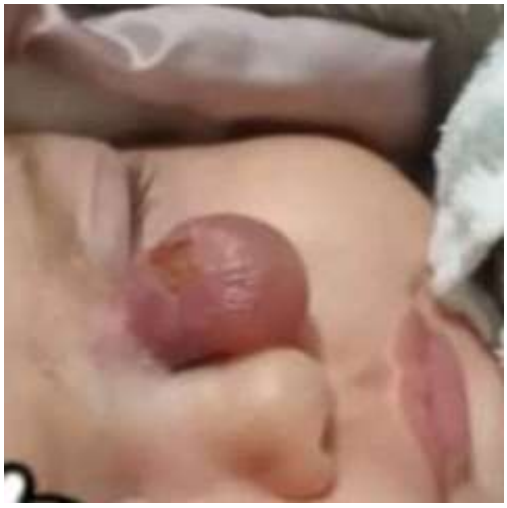

Figure 1. Clinical Presentation of the patient.

course was favorable, marked by the absence of recurrence over a three-year follow-up period (figure 4).

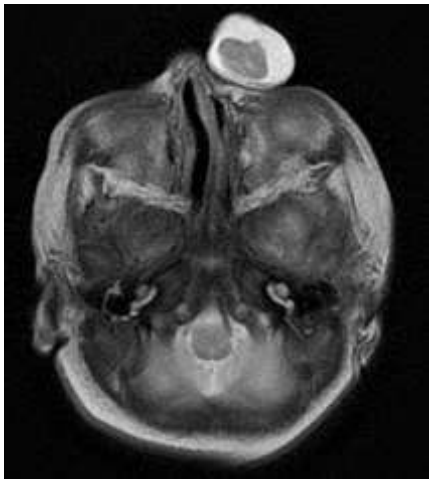

Figure 2. Radiological Assessment on MRI.

3. DISCUSSION

Nasal gliomas were first described by Reid in 1852 [3], with Schmidt being the first to study them, coining the term "glioma" in 1900 [4]. This term, implying a neoplasm, is somewhat misleading since the growth of nasal gliomas is generally slow and comparable to that of normal brain tissue.

Some authors have suggested the terms "encephaloma" or "nasal cerebral heterotopia" [5].Understanding the embryology of the naso-frontal region allows for a topographical classification of midfacial malformations. External nasal gliomas, forming a firm mass between the nose tip and glabella, often

Figure 3. Surgical Treatment.

lateralized, may cause visual field anomalies or compression of lacrimal ducts [2].

Figure 4. Surgical Treatment Outcome.

The root of the nose may appear widened. Internal nasal gliomas present as a firm, pale intranasal mass, potentially causing nasal obstruction through bone or cartilage displacement. In neonates, due to their exclusively nasal breathing, the ventilatory consequences can be significant. Other signs like epistaxis or abnormal nasal secretion may be revealing. Nasal endoscopy is performed to ascertain the mass's location, origin, and extent, and to identify any pulsatile nature. Needle puncture or biopsy is strictly contraindicated before ruling out potential intracranial extension.

Radiological evaluation through CT and MRI in axial, coronal, and sagittal sections is essential to determine the lesion's location, size, and content, and to verify the integrity of the anterior base of the skull. In young children, the absence of ossification at the anterior base may limit the effectiveness of CT scans, making MRI with contrast enhancement a valuable tool for delineating the

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cartilaginous skull base and determining the mass's tissue or fluid characteristics [5].

Therapeutic management of nasal gliomas requires a multidisciplinary approach, involving maxillofacial surgeons, ENT specialists, neurosurgeons, and neuroradiologists. These lesions, characterized by slow growth and no risk of malignant degeneration, necessitate a comprehensive diagnostic evaluation, with a search for any associated congenital malformations

The treatment of choice for nasal heterotopia is surgical excision. Delay in treatment can lead to deformities of the septum or nasal bones. External nasal gliomas typically require an external surgical approach (external rhinoplasty, lateral rhinotomy, median incision, or coronal approach), with conservative treatment of adjacent nasal structures to avoid impacting future growth [1, 2].

4. CONCLUSION

Nasal gliomas present as isolated masses from the brain but can have fibrous attachments to it in 15% of cases. Differential diagnosis primarily includes hemangioma and encephalocele, highlighting the importance of imaging through CT and MRI. The diagnosis is histological, and the treatment is surgical.

Conflicts of interest : None with regard to the article.

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