

Case Reports

Nasal Gliomas

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Nasal encephaloceles and gliomas present along a broad spectrum of severity and deformity. A combination of transfacial and intracranial approaches is often required for the safe and definitive treatment of these anomalies.

As a group, congenital midline nasal masses are unusual lesions, having an incidence of 1 in 20,000 to 40,000 live births.¹ Nasal gliomas, both intranasal and external, represent the least common subset of these patients with less than 170 cases described in the literature.² The vast majority of these lesions are diagnosed at birth or in early childhood. Only a handful of gliomas have remained undetected until adulthood.²⁻⁵ They are thought to share a common embryogenesis with encephaloceles. These lesions are clinically relevant primarily as a consequence of their potential for communication with the central nervous system. Histologically, gliomas are characterized by unencapsulated collections of astrocytic eosinophilic glial cells in a connective tissue matrix that may retain some fibrous attachments to the dura. Encephaloceles represent herniation of the meninges (with or without associated brain parenchyma) through the skull base. Unlike most gliomas, encephaloceles will communicate with the subarachnoid space cerebrospinal fluid compartment.

In this article, a previously undescribed lesion with two distinct, well-defined tracts leading from the floor of each anterior cranial fossa into a single intranasal glioma presenting in adulthood will be used to highlight the diagnosis and treatment of this clinical entity.

Case Report

HC is 42-year-old African-American male who presented to the Otolaryngology and Facial Plastic Surgery service with a long-standing history of right-sided nasal

obstruction. The patient had no antecedent history of facial trauma and was in good overall health. Initial intranasal examination revealed the presence of a 3-cm smooth, soft submucosal mass. Rigid endoscopic examination revealed that the mass was not separable from its attachment to the septum or the undersurface of the nasal bones. The lesion was distinct from the lateral nasal wall. The mass was not compressible and did not appear to transilluminate or change in size with Valsalva maneuvers. There was no evidence of present or past cerebrospinal fluid leakage or meningitis. The possibility of intracranial communication or origin was entertained at this point and confirmed by subsequent computed tomography (CT) and magnetic resonance imaging (MRI) scans (Fig. 1). A dehiscence in the floor of the right anterior cranial fossa in continuity with the intranasal lesion was noted on these studies.

Thus, the patient was brought to the operating room for definitive resection of this lesion. An initial frontal craniotomy confirmed the presence of a 1.5-cm defect in the floor of the right anterior cranial fossa with a fibrous connection extending from the dura to the intranasal lesion. A transfacial lateral rhinotomy was then used to gain access to the nasal mass (Fig. 2). Due to its intimate attachment to both the posterior septum and the undersurface of the nasal bones, osteotomies were performed at these levels to allow for complete and safe removal of the lesion. Once fully mobilized, the tract leading to the floor of the right anterior cranial fossa floor was identified and

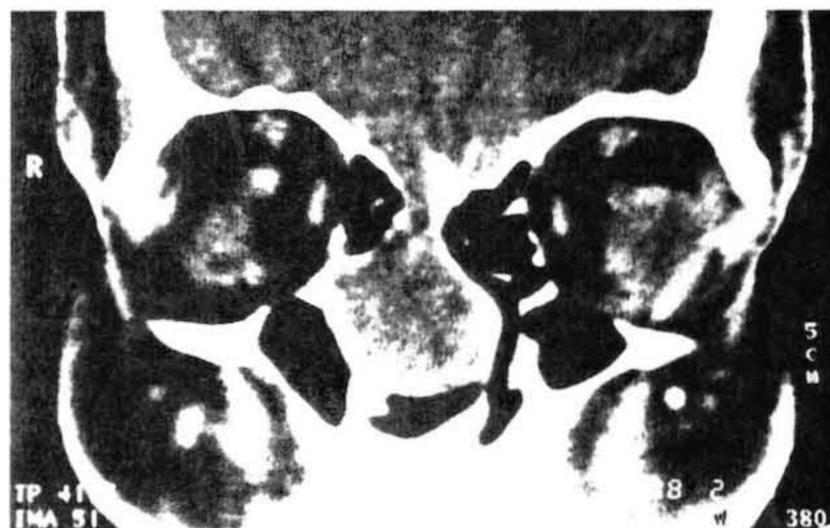


Figure 1 Coronal CT scan demonstrating intracranial communication between nasal mass and floor of right anterior cranial fossa.

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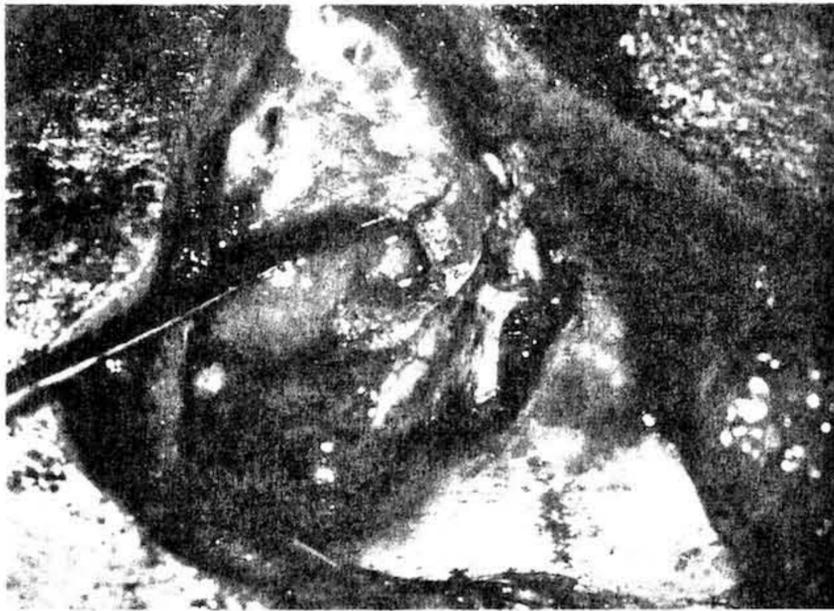


Figure 2 Intraoperative photograph taken via lateral rhinotomy approach. Note elevator between caudal margin of nasal bone and nasal mass.

removed in continuity with the main specimen. Further dissection more posteriorly revealed the presence of a second distinct tract measuring 1.0 cm in diameter leading to the floor of the left anterior cranial fossa (Fig. 3). This, too, was removed in continuity with the nasal lesion after intracranial exposure of the left side was obtained (Fig. 4). Pathologic analysis revealed the presence of benign astrocytic neuroglial cells in a fibrous stroma, pathognomic of intranasal gliomas. The patient's nasal dorsum was reconstructed with a split calvarial bone graft. The patient has done well postoperatively with no evidence of intranasal disease or cerebrospinal fluid leakage (Fig. 5).

Discussion

Gliomas and encephaloceles appear to be developmentally related, with gliomas likely representing encephaloceles that were amputated at the level of the skull

base during development. Failure of normal migration of neural crest cells into the anterior cranial region would result in an area devoid of the mesenchyme necessary for skull base formation, resulting in a protrusion of intracranial contents, forming an encephalocele. If neural crest cell migration is delayed, subsequent osseous skull base formation would result in amputation of the intracranial communication, resulting in the formation of a nasal glioma. Incomplete skull base closure may give rise to persistence of a fibrous stalk that may be intimately associated with the dura.

Nasal gliomas are generally noted in infancy or early childhood and have only very rarely been diagnosed in adulthood. There is no recognized familial tendency, malignant potential, or sex predilection and, for the most part, they are not associated with other developmental anomalies.⁶ They are generally firm, noncompressible masses that do not transilluminate or expand with Valsalva or crying. Sixty percent of these lesions present externally, 30% present intranasally, and 10% have both components.⁷ External gliomas may, most often, be found at the level of the glabella. Intranasal gliomas are almost universally noted to arise along the lateral nasal wall at or above the level of the middle turbinate. Overall, only 10 to 15% of gliomas have a connection to the dura.⁸ It should be noted that intranasal gliomas are two to three times as likely to have such a connection as compared to their external counterparts. Communication with the intracranial space may lead to meningitis, on occasion.

The possibility of nasal glioma may be entertained based upon clinical evaluation and the use of adjunctive studies (CT and MRI scans); however, definitive diagnosis is often only possible after complete surgical excision. Incisional biopsies should be performed with caution, if at all, under controlled circumstances in the operating room, due to the risk of serious cerebrospinal fluid leak



Figure 3 Following disarticulation and inferior displacement of nasal root (with glioma attached to its deep aspect), two distinct tracts leading to the anterior cranial fossa are demonstrated with elevators.

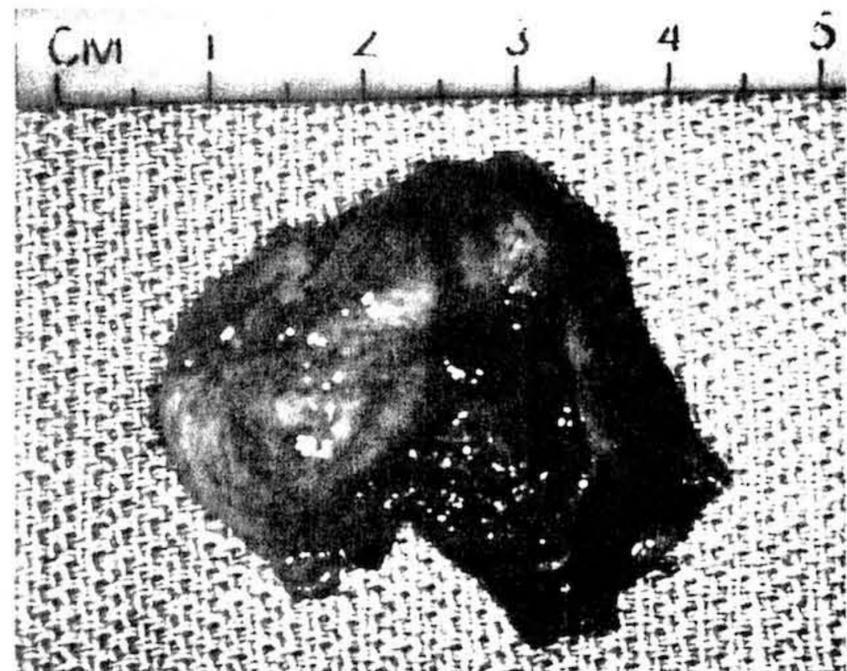


Figure 4 Surgical specimen.

with subsequent intracranial infection risk. A CT and/or MRI scan should be obtained in each patient to determine the presence of intracranial communication. These imaging modalities are often complementary when used in the evaluation of gliomas and encephaloceles.

Adequate surgical treatment of gliomas is associated with a recurrence rate of less than 10%.⁶ If there is no evidence of intracranial communication, external/transfacial approaches are adequate. Neurosurgical consultation may occasionally still be necessary if a previously unrecognizable tract is identified intraoperatively to be coursing to the skull base. Otherwise, external gliomas are generally excised in an elliptical fashion directly over the lesion's centre. An external rhinoplasty approach may provide for excellent visualization of the lesion as well as direct access to the nasal bones, which may be transgressed by fibrous stalks connecting to the dura. Median osteotomies at this level (if required to gain transfacial access to a fibrous stalk) will allow for controlled, complete lesion excision. Intranasal gliomas, if small and lacking intracranial communication, may be excised endoscopically. Larger lesions or those with intracranial communication are often best approached via a lateral rhinotomy as in the illustrative case presented. In case of suspected intracranial communication, based either on clinical examination or radiographic examination, initial frontal craniotomy is suggested to allow for safe, subsequent transfacial excision.

The case presented in this article is unique in a number of ways. It is unusual for a glioma to have remained undetected and to have presented in adulthood. Furthermore, its attachment to the septum and undersurface of nasal bones and lack of any distinct origin at the level of the lateral nasal wall is unusual. The presence of two distinct stalks arising from a single glioma and extending to the floors of both anterior cranial fossae has been a previously unrecognized variant of this lesion. This dual communication is significant in that it was not identifiable on preoperative imaging or clinical examination. Diligent circumferential dissection of these lesions is necessary to allow for clear visualization of any tracts communicating with the dura. One should maintain such vigilance even in the presence of what initially appears to be a strictly extracranial glioma, as imaging studies are not infallible and the presence of multiple tracts is possible. Failure to adequately excise all such tracts would, in theory, increase the risk of recurrence of these lesions.

Conclusion

Nasal gliomas generally present as congenital masses that are detected in early infancy or childhood. Presentation of a unilateral, firm, noncompressible intranasal mass in an adult should also lead one to consider the



Figure 5 Patient's 2-month postoperative appearance.

possibility of this entity. Although CT and/or MRI scanning is suggested to determine the presence of any intracranial communication, the absence of such a connection does not rule out the presence of single or multiple tracts leading from the lesion to the dura. Such tracts should be excised with a combined intracranial-transfacial approach.

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