

Nasal Glioma: An Unusual Presentation in Adult

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ABSTRACT

The nasal glioma is a rare congenital nasal abnormality, which manifests as a mass of extracranial cerebral tissue unconnected with the brain. The clinical entity is challenging for several reasons. Their diagnosis may not be immediately obvious at presentation. It is important to exclude an intracranial extension, and for this some have suggested a craniotomy. We report a case of extranasal glioma with intracranial extension was removed successfully by combined extracranial and endoscopic approach without craniotomy. Intranasal endoscopic surgery is a less invasive and safe procedure for the management of nasal glioma and does not result in postoperative facial scarring and deformity. Endoscopic techniques provide excellent visualization and are preferable to the classic frontal craniotomy to exclude intracranial extension of nasal glioma. In this report, the management and the benefits of endoscopy in cases with nasal glioma are discussed.

Keywords: Nasal mass, Glioma, Endoscopic sinus surgery, Approaches, Skull base.

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INTRODUCTION

Nasal glioma is a rare congenital nasal lesion which manifests as a mass of extracranial cerebral tissue unconnected with the brain.¹ Synonyms on the basis of etiology include nasal cerebral heterotopia or sequestered encephalocele. A congenital nasal mass is said to occur in 1 in every 20,000 to 40,000 live births.² According to Bradley et al nasal glioma is reported to account for five of 109 congenital nasal masses.³ Clinically, 60% of nasal gliomas are extranasal, 30% are intranasal and 10% are mixed extra- and intranasal.⁴ Extranasal gliomas appear in or just lateral to the midline. Intranasal gliomas usually arise from the lateral nasal wall. Nasal gliomas are more common in boys than in girls.⁵ There is no familial predisposition and no malignant potential. Nasal glioma can potentially extend intracranially with a fibrous stalk in 10 to 25% cases.⁶ Early surgical correction is imperative in view of the potential risk of cerebrospinal fluid (CSF) leak and meningitis caused by the intracranial extension of some of these lesions.⁴

There are recommendations of craniotomy for intracranial extension. However, a thorough work-up, including magnetic resonance imaging (MRI) and intranasal endoscopy could eliminate unnecessary craniotomy. An extranasal approach is usually used for their surgical

extirpation, because such an approach allows visualization of the skull base.^{3,4} A lateral rhinotomy or endoscopic approach is recommended for surgical excision of nasal glioma without intracranial extension.⁷ We describe a case in which extranasal glioma with intracranial extension was removed successfully by combined extracranial and endoscopic approach without craniotomy. In this report, management, especially the surgical procedures employed and the benefits of endoscopy, in cases with nasal glioma are discussed.

CASE REPORT

A 20-year-old boy presented a 2 × 2 cm midline mass present in region of root of nose since childhood (Fig. 1). There was a history of surgical excision at 13 years of age with reappearance of swelling 1 year postsurgery. The mass was firm, noncompressible, nonpulsatile and with negative Frustenberg sign. There was no history of CSF leak or meningitis. Magnetic resonance imaging revealed a well defined oval mass measuring approximately 19 × 15 × 10 mm located in soft tissues overlying the nasal bridge which was hyperintense in both T1- and T2-weighted images. A narrow tract about 1.5 mm wide and 2 cm long was seen extending horizontally and then posterosuperiorly from the lesion along right margin of nasal septum ending in crista galli. Dura appeared to separate the lesion from frontal lobe with no obvious intracranial extension (Fig. 2).

The lesion was completely resected surgically by an external curvilinear incision which revealed a 2 × 2 cm well-encapsulated mass in the soft tissues overlying the root of nose with a defect in between the two nasal bones. Through this defect, a fibrous band was extending intranasally which



Fig. 1: Clinical photograph of the patient showing midline mass present in region of root of nose

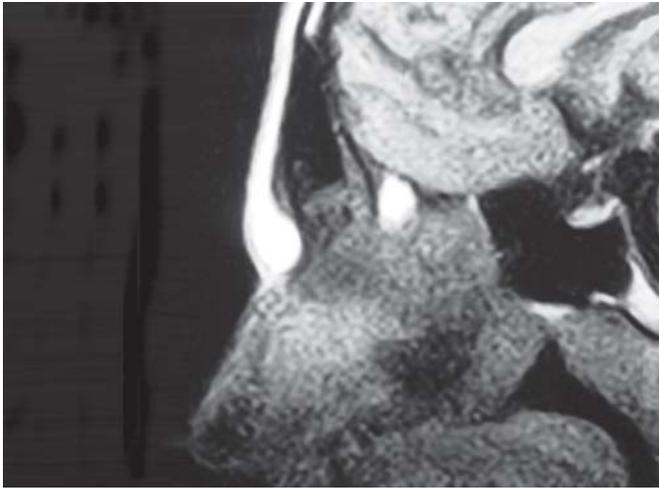


Fig. 2: Magnetic resonance imaging showing a well-defined hyperintense mass located in soft tissues overlying the nasal bridge

was visualized endoscopically in the upper part of right nasal cavity in the region of right cribriform plate (Fig. 3A). There was a skull base defect in the region of right cribriform plate which was repaired in layers using mashed muscle and fascia lata with underlay technique (Fig. 3B). The skull base defect was repaired by cartilage harvested from the nasal septum. The nose was packed with merecele which was removed after 5 days. The postoperative period was uneventful.

Histopathologic examination of the mass confirmed the diagnosis of nasal glioma. It consisted of mature glial tissue, astrocytes, and fibrosis. At 1 year follow-up the patient was doing well with no evidence of residual or recurrence.

DISCUSSION

Nasal glioma or glial heterotopia is nonhereditary congenital lesion composed of mature brain tissue isolated from the central nervous system, which has a potential for intracranial



Fig. 3A: Endoscopic view of the fibrous band extending from the extranasal mass to the upper part of right nasal cavity in the region of right cribriform plate



Fig. 3B: Skull base defect repaired with fascia lata and cartilage

extension.⁸ Nasal glioma is not a real neoplasm. It is considered an embryologic growth abnormality.⁶ It is frequently diagnosed in the pediatric population, presenting with upper airway symptoms, feeding disturbance or facial deformity.⁹ Rarely, it is be found in adults as in our case. Extranasal glioma usually presents as a noncompressible, firm and smooth mass located on the bridge or side of the nose similar to our case. Within the nasal cavity, it presents as a polypoid structure.¹⁰ Intranasal glioma most often arises from the lateral nasal wall near the middle turbinate, but some arise from the nasal septum as well.¹¹

Clinically, a nasal glioma must be differentiated from an encephalocele having meningeal continuity with the brain. Whereas encephalocele may pulsate and expand with compression of the jugular vein (positive Furstenberg test), nasal glioma is nonpulsatile and Furstenberg test is negative. Encephaloceles are frequently associated with cerebrospinal fluid (CSF) leak or history of meningitis, whereas these are rare with nasal gliomas. Biopsy or aspiration is contraindicated because of the risk of subsequent intracranial infection or damage to functional brain tissue within an encephalocele.¹²

In 10 to 25% of cases of nasal glioma, a fibrous stalk connects it to the intracranial space.⁶ Some of these gliomas may be associated with a bony defect in the cribriform plate and CSF leakage during surgery similar to our case.⁴ Intranasal glioma is more likely to have an intracranial extension than is an extranasal or mixed glioma.¹³ Since, CSF leakage results in severe complications like meningitis, exclusion of the possibility of intracranial extension is a most important issue in the clinical evaluation of intranasal glioma.

CT and conventional radiographic studies have limitations in imaging of small bony defects or narrow stalk like connections across the skull base.² MRI is considered

the investigation of choice for detailed evaluation of congenital nasal masses, especially those having intracranial extension.¹⁴ But evaluation of the nasofrontal region is difficult in the neonate because the anterior skull base is largely cartilaginous at birth. The percentage of ossified skull base steadily increases over the first 2 years of life. In the normal neonate, no bone is seen in the region of the crista galli and cribriform plate and the absence of bone should not be misinterpreted as a sign of an encephalocele. Chau et al⁷ concluded that MRI is the preferred method of investigation but advised that preoperative radiological appearances could be misleading. Nasal endoscopy should be performed in all cases to determine the precise location, origin, extent and pulsatility.⁸

The treatment of choice for nasal glioma is surgical resection. Early surgical intervention is imperative in view of the potential risk of facial deformity, CSF leak and meningitis caused by the intracranial extension.¹⁵ Surgical approaches should be based on the location and size of the mass, associated cartilage, or bony deformity and the experience of the surgeon. In cases of nasal glioma with definite intracranial extension, craniotomy must be performed before removal of the mass. Craniotomy is required for excision of the transcranial stalk and adequate repair of the skull base defect.

When intracranial extension has been ruled out, the mass can be removed without craniotomy. Extranasal gliomas are removed with the surrounding skin by external incision.¹⁶ Even for intranasal gliomas, many surgeons have considered extranasal approaches as appropriate for mass removal because of visualization of the skull base.^{2,4} However, extranasal approaches are associated with postoperative cosmetic problems and subsequent abnormal development of the nose and paranasal sinuses.¹⁷ Dupin and Jeune¹⁵ advocated the extracranial excision of nasal gliomas after excluding the following possibilities: Nasal encephalocele, positive Furstenberg test, previous meningitis, cerebrospinal fluid leak and bony defect in the cribriform plate.

According to Yokoyama et al¹¹ an intranasal endoscopic approach has some advantages over extranasal approaches. Endoscopy allows clear visualization of each wall of the nasal cavity, and precise surgery can be carried out with minimum damage to intact tissues. Endoscopy enables complete removal of the mass without postoperative facial deformity. Therefore, the intranasal endoscopic approach is strongly recommended for the removal of intranasal gliomas.¹⁸ As angled endoscopes allow visualization of the cribriform plate, therefore intranasal endoscopic surgery is expected to replace craniotomy as the procedure of choice to exclude an intracranial extension of the glioma.¹¹

According to Chung-Lun Wu et al¹⁹ in cases with suspected bony defects, an intranasal endoscopic approach is considered the procedure of choice.

In our case of extranasal glioma with intracranial connection, craniotomy was avoided by using a combined extranasal and intranasal endoscopic approach. With the help of angled endoscope the skull base defect in the region of cribriform plate with intraoperative CSF leak was identified and repaired successfully.

Close follow-up is also important in these cases because of the possibility of postoperative CSF rhinorrhea and infection. The recurrence rate is low and is reported to be 10%.¹³ This is generally thought to be the result of incomplete excision. Endoscopic examination is recommended for postoperative follow-up along with MRI, because this noninvasive procedure can be performed repeatedly. In our case, there were no intranasal endoscopic or MRI pathologic findings after 1 year of follow-up.

CONCLUSION

The management of nasal glioma may prove quite challenging. Preoperative imaging is mandatory for the diagnosis of an intracranial connection to avoid an unnecessary craniotomy. Intranasal endoscopic surgery should be considered as a procedure of choice for the removal of intranasal glioma and for excluding intracranial extension in cases of extranasal glioma and thus avoiding craniotomy.

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