CASE REPORT

Endoscopic Surgery for Nasal Glioma Mimicking Encephalocele in Infancy

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ABSTRACT

Intranasal gliomas are 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 9-day-old male infant in whom an intranasal glioma that mimicked an encephalocele was successfully excised by endoscopic approach after failure to excise it through a craniotomy. After more than a 5-year follow-up, the patient is still free from the disease. An endoscopic intranasal approach provides a safe and effective method for the management of nasal glioma and does not result in postoperative facial scarring or deformity. Endoscopic techniques provide excellent visualization and are preferable to the classic frontal craniotomy to excluding intracranial extension of nasal glioma.

KEYWORDS: Nasal glioma, encephalocele, endoscopic, infancy

Minimally invasive endoscopic surgery has emerged as the standard and preferred technique for several surgical disciplines including urology, general, and orthopedic surgery because of its inherent minimal invasiveness and short recovery time. Such operations are not yet standard practice in much of the head and neck region, perhaps because of its inherent anatomic complexity. Head and neck applications of endoscopy have mainly been limited to cosmetic surgery, thyroidectomy, and sinus surgery. We have advocated several indications for an endo-

scopic approach in the head and neck and skull base for both benign and malignant lesions.¹⁻⁴ In this study, we present the outcome of endoscopic surgery for a 9-day-old male infant with a nasal glioma that mimicked an encephalocele.

Nasal glioma is a rare, benign, congenital abnormality that presents as a mass of extracranial cerebral tissue that is usually not in continuity with the brain, although 10 to 25% may have fibrous tracts connecting to the subarachnoid space.⁵ Approximately 250 cases have been reported in

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Received: March 10, 2008. Accepted: May 9, 2008. Published online: October 7, 2008.

DOI 10.1055/s-0028-1087219. ISSN 1531-5010.

the English literature.⁶ Nasal gliomas are more common in boys than in girls. There is no familial predisposition and no malignant potential. Clinically, nasal gliomas involve the external nose in up to 60% of cases, are restricted to the nasal cavity in 30%, and involve both in 10%.⁷ A lateral rhinotomy or endoscopic approach is recommended for surgical excision of nasal glioma without intracranial extension.^{8,9}

CASE REPORT

A 9-day-old male infant was admitted to our tertiary medical center with nasal obstruction associated with a yellowish nasal discharge and productive cough that had been present from birth. He was a full-term baby and both the pregnancy and delivery had been normal. Initially treated as an infection, his primary care team had experienced difficulty inserting a nasogastric tube through his right nostril. Physical examination revealed a soft tissue mass that was attached to the upper part of the left nasal cavity and skull base. Purulent rhinorrhea and deviation of the nasal septum toward the right caused obstruction of the right nasal cavity. Magnetic resonance imaging (MRI) demonstrated the presence of a soft tissue mass similar in signal intensity to that of gray matter in the left anterior nasal cavity and displacing the septum to the opposite side (Fig. 1). A small bone defect was suspected in the left side of the cribriform plate anteriorly, and this contained soft tissue with a signal intensity similar to that of brain (Fig. 2). The nasal mass was separate from the brain parenchyma in the frontal lobe. The differential diagnosis included glial heterotopia (nasal glioma), meningoencephalocele, and, less likely, esthesioneuroblastoma.

Under general anesthesia with endotracheal intubation, a bifrontal craniotomy was undertaken to repair what was presumed to be an encephalocele. This was unsuccessful as the nasal glioma was not connected to the brain by a stalk and there was no bone defect in the cribriform plate or anterior

Figure 1 Preoperative axial magnetic resonance imaging (T2-weighted, postgadolinium enhancement) shows a soft tissue mass similar in signal intensity to gray matter in the left anterior nasal cavity and displacing the septum to the opposite side.

Figure 2 Preoperative sagittal magnetic resonance imaging (T2-weighted, postgadolinium enhancement) shows a suspicious small bone defect in the left anterior cribriform plate with soft tissue signal intensity similar to the brain.

skull base. Using endoscopic techniques, a reddish, soft mass that did not pulsate was seen in the lateral wall of the left nasal cavity and skull base. This was removed quite simply and without complication endoscopically. No connection to the central nerve system or any cerebrospinal fluid (CSF) leak was found. The patient's nose was packed, and the packing was removed on the first postoperative day. The patient's recovery was uncomplicated, and he was fit for discharge on day 7.

Histopathologic examination of the mass confirmed the diagnosis of nasal glioma. It consisted of mature glial tissue, astrocytes, and fibrosis.

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 extension.⁷ It is frequently diagnosed in the pediatric population, presenting with upper airway symptoms, feeding disturbance, or facial deformity. Rarely is it found in adults. Extranasal glioma usually presents as a noncompressible and smooth mass located on the bridge or side of the nose. The consistency of nasal glioma is firm. Within the nasal cavity, it presents as a polypoid structure.¹⁰ Although intranasal glioma most often arises from the lateral nasal wall near the middle turbinate, some arise from the nasal septum.¹¹

The differential diagnosis of a nasal glioma from an encephalocele is based on the presence or absence of a connection between the mass and the intracranial cavity. Whereas encephaloceles may pulsate and expand when the jugular vein is compressed (positive Furstenberg test), nasal glioma is nonpulsatile and gives a negative response to the Furstenberg test.¹² Biopsy or aspiration is contraindicated because of the risk of subsequent intracranial infection or damage to functional brain tissue within an encephalocele.¹³

Evaluation should include preoperative computed tomography (CT) or MRI to exclude an intracranial extension. However, even with high-resolution CT and MRI, the connection may be very small and not immediately apparent. 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; the absence of bone should not be misinterpreted as a sign of an encephalocele. Chau et al⁹ 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.⁷

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.¹⁴ Surgical approaches should be based on the location and size of the mass, associated cartilage, or bony deformity and the experience of the surgeon. Some surgeons have recommended an initial craniotomy to exclude intracranial extension. Craniotomy can also prevent any postoperative CSF rhinorrhea or intracranial infectious complications.⁷ We advocate transnasal endoscopic excision after excluding the following possibilities: encephalocele, a positive Furstenberg test, and any intracranial extension.

As nasal glioma is not a true neoplasm and recurrence rate is low, an endoscopic approach is advocated for this condition. Endoscopy allows a clear view of each wall of the nasal cavity, and precise excision of intranasal glioma can be achieved with minimal damage to normal tissue. Because angled endoscopy allows visualization of the cribriform plate, intranasal endoscopic surgery is expected to replace craniotomy as the procedure of choice to exclude an intracranial extension of the glioma.¹¹ Close follow-up is also important for these cases because of the possibility of

postoperative CSF rhinorrhea and infection. Endoscopic examination can also be undertaken easily for follow-up purposes. In our case, there has not been any recurrence or complication after a follow-up period of more than 5 years.

CONCLUSION

Intranasal glioma may prove quite challenging because the diagnosis at presentation is not immediately obvious. In addition to CT or MRI, the preoperative diagnosis of a skull defect with intranasal endoscopy is essential to avoid an unnecessary craniotomy. In cases with suspected bony defects, an intranasal endoscopic approach is considered the procedure of choice.

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