A congenital extranasal glioma in a newborn

Nicole A Bailey

Abstract
Nasal gliomas are extremely rare in neonates with an incidence of 1 in 20,000 to 40,000. They often are asymptomatic but can present with respiratory distress depending on the size and location of the tumor. A newborn female was prenatally diagnosed with a left nasal mass. After her birth, she was transferred to a local children’s hospital for subspecialty evaluation and for diagnostic imaging. The mass was resected at 1 year of age. Pathology confirmed a nasal glioma. Several weeks after surgery, a nasal prosthetic device was applied to correct the nasal deformity caused by the pressure effect of the tumor. At almost 1 year of age, there was no evidence of metastasis or recurrence of the nasal glioma. The prognosis and outcome tend to be favorable. The rare case of a neonate with a congenital nasal glioma is presented.

Keywords
Glial tissue, heterotopy, newborn, nasal glioma, congenital nasal disorder, sinonasal tumors

Introduction
Congenital nasal gliomas are rare with an incidence of 1 in 20,000 to 40,000 live births.\(^1\)\(^-\)\(^4\) Their etiology is unknown. However, “the most commonly accepted hypothesis attributes their occurrence to an ectopic trapping of herniated brain tissue.”\(^3\) Nasal gliomas are embryologically related to encephaloceles, but only 20% of nasal gliomas have an intracranial connection like encephaloceles.\(^2\)\(^-\)\(^3\)\(^,\)\(^5\) In this report, the case of a full-term baby born with a nasal glioma is presented.

Case presentation
A newborn female was born via a normal spontaneous vaginal delivery at 40-week and 1-day gestation with a nasal mass. Her mother had limited prenatal care. However, a prenatal ultrasound at 5-month gestation showed the nasal mass. The pregnancy was otherwise uncomplicated. The Apgar scores were 9 and 9 at 1 and 5 min, respectively. Her birth weight was 2980 g (above the 25th percentile). Her length was 47.5 cm (25th percentile), and her head circumference was 32 cm (10th percentile). The patient had no respiratory distress at birth and was subsequently admitted to a newborn nursery at a community hospital. The patient was transferred on day of life 2 to a neonatal intensive care unit (NICU) at a local children’s hospital for subspecialty evaluation.

Physical examination
Upon admission to the children’s hospital, the patient’s temperature was 98.8° F, heart rate was 150 beats per minute, respiratory rate was 40 breaths per minute, and oxygen saturation was 97% on room air. Her physical exam was only notable for a skin-colored soft mass located on the left side of the patient’s nose that was non-mobile and without evidence of pulsation, tenderness, obstruction, or discoloration (Figure 1). There were no other gross anomalies.

Laboratory tests
An initial complete blood cell count showed normal findings—white blood count: 10,000/μL (10.0 × 10¹⁰/L), hematocrit: 51% (0.51), and platelet count: 413 × 10³/μL (413 × 10⁹/L). Her electrolytes and bilirubin levels were also normal.

Division of Neonatology, Nicklaus Children’s Hospital, Herbert Wertheim College of Medicine, Florida International University, Miami, FL, USA

Corresponding Author:
Nicole A Bailey, Division of Neonatology, Nicklaus Children’s Hospital, Herbert Wertheim College of Medicine, Florida International University, 3100 SW 62nd Ave, Advanced Care Pavilion (4th floor), Miami, FL 33155, USA.
Email: nicole.bailey@nicklaushealth.org
Initial imaging studies

An ultrasound of her face showed a solid mass arising from the dorsum of the nose. No significant internal vascularity was seen. Magnetic resonance imaging (MRI) of the face showed a left nasal soft tissue mass measuring $1.9 \text{ cm} \times 2.1 \text{ cm} \times 1.5 \text{ cm}$ that did not extend into the nasal cavity proper. Some mass effect and deformity over the left nasal soft tissue were seen. The nose was not completely occluded. There was no connection to the anterior cranial fossa. No restricted diffusion was seen (Figures 2 and 3).

Differential diagnosis

The differential diagnosis for a nasal mass includes a hemangioma, nasal glioma, encephalocele, fibrosarcoma, fibroma, rhabdomyosarcoma, and dermoid cyst. Imaging and pathology results are helpful in obtaining the final diagnosis.

Clinical course, evaluation, and management

On admission to the NICU, plastic surgery and neurosurgery were consulted who recommended the initial imaging described above. The patient continued to do well and was subsequently discharged from the hospital on day of life 4 with subspecialty follow-up. Over the next several months, the nasal mass grew slightly and was noted to distort the left nasal ala caudally. Since the facial MRI excluded an encephalocele, the plastic surgery team recommended a computed tomography (CT) of the brain with a three-dimensional (3D) reconstruction which was performed at age 5 months for surgical planning. This study was notable for a large area of mass effect along the midline and left side of the nose, in
intimate contact with the cartilaginous septum and deformity of the left sided nasal bones without bony destruction or lytic lesion. Mucosal thickening/fluid was noted in the maxillary and ethmoid sinuses (Figures 4 and 5).

At almost 1 year of age, the nasal mass was resected in its entirety under general anesthesia without evidence of intracranial extension. The extra skin overlying the nose was used to close the defect. The final surgical pathology report was consistent with a nasal glioma (Figures 6 and 7). There were no post-operative complications. The patient was discharged home the following day.

Several weeks after the surgery, a nasal prosthetic device was placed to allow for rehabilitation of the soft tissue and cartilage in the nose and to correct the nasal deformity caused by the pressure effect of the mass. The patient was instructed to wear the device for 24 h a day for 4–6 months.

Nine months following the surgery, the patient continued to do well and no longer required the nasal prosthetic device. Her mother was content with the cosmetic results.
Discussion

Nasal gliomas can be extranasal (60%), intranasal (30%), or mixed (10%),1,3,4 with the intranasal forms mainly causing respiratory symptoms. They are typically benign, do not transform into malignant lesions, and are slow growing.

Given advances in modern technology, the prenatal diagnosis of a nasal mass is possible.6 However, it is important to obtain postnatal imaging to confirm the absence of intracranial connections prior to any biopsy or surgical procedure. In addition, imaging and biopsy samples can assist in differentiating nasal gliomas from other masses such as dermoid cysts, fibrosarcoma, fibroma, rhabdomyosarcoma, and hemangiomas. For instance, MRI will be helpful with excluding intracranial communications and examining soft tissue, while CT is helpful with examining bony deformities associated with the mass.2 Furthermore, an ultrasound may assist with determining whether tissue is cystic, solid, or vascular, the latter of which is more commonly associated with hemangiomas.7 Histologically, nasal gliomas are “composed of large and small islands of glial cells within connective fibrous tissue,”2 while dermoid cysts “are lined by keratinized stratified squamous epithelium, containing skin tissues or dermal appendages.”2 When present, ependymal tissue is more commonly associated with encephaloceles than with nasal gliomas.2 Histological examination can also exclude other tissue malformations in the differential diagnosis such as fibrosarcoma, fibroma, and rhabdomyosarcoma.

The treatment of nasal gliomas is early surgical excision. If not surgically resected, these gliomas can cause bony atrophy and intracranial communications which may predispose the patient to meningitis and other infections.5,6 Potential complications of surgery may include recurrence of the mass with rates ranging from 4% to 10% if the mass is not adequately resected.2,3,4,8

Conclusion

In summary, nasal gliomas are rare masses that are usually benign. They are embryologically related to encephaloceles and can have similar appearances to other nasal masses. Thus, imaging and histopathology are paramount in the diagnosis of nasal gliomas. Surgical excision is the treatment to prevent complications and recurrence. A multi-disciplinary approach is needed to maximize patient satisfaction and outcomes.

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Ethics approval

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Informed consent

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ORCID iD

Nicole A Bailey https://orcid.org/0000-0003-1336-3997

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