Congenital midline nasal anomalies are rare, with a prevalence of 1 in 20,000 to 40,000 births and with 5% to 7% of them being nasal glioma. Differential diagnoses of nasal anomalies include nasal dermoid cysts, gliomas, encephaloceles, nasal polyps, and some other rare anomalies. Due to current medical technological advancements, most of these anomalies are easily correctable, though delaying management may lead to fatal effects. This report describes two cases—one of nasal glioma and one of nevus lipomatosus cutaneous superficialis—that presented as respiratory distress in a newborn. Approximately 10 to 20 cases of these two conditions have been described; notably, this is the second documented case of nevus lipomatosus cutaneous superficialis with nasal presentation.

Two cases involving congenital midline anomalies in neonates manifesting as respiratory distress in newborns are discussed here, one with a diagnosis of nasal glioma and one with a diagnosis of nevus lipomatosus cutaneous superficialis (NLCS) in the nasal cavity. Approximately 20 cases of nasal glioma have been reported in English publications; less than 10 cases of NLCS, with only one other case involving the nasal cavity, have been reported (1–6).

CASE 1
A 40-week and 1-day-old gestational age female infant born in a tertiary center to a 27-year-old mother with good prenatal care and an uncomplicated pregnancy was initially transferred to the newborn nursery for routine care after receiving Apgar scores of 9 and 9 at 1 and 5 minutes of life. Subsequently the infant developed worsening respiratory distress along with increased difficulty breathing and intercostal retractions at 8 hours of life. A nurse reported three consequent desaturations in the newborn nursery; the attending neonatologist was consulted, and the infant was transferred to a tertiary neonatal intensive care unit (NICU) for further management. The infant was placed on nasal bubble continuous positive airway pressure at 4 cm of water for respiratory support with a fraction of inspired oxygen of 21%. A chest x-ray obtained in the NICU showed no obvious abnormalities. A complete blood count with differentials obtained at the time of NICU admission was within normal limits as well. The pediatric otolaryngology department was consulted for evaluation of respiratory obstruction. The infant was found to have a “fleshy mass” in the right anterior nasal cavity via nasal endoscopy (Figure 1a). The patient underwent computed tomography of the face, which showed a 1.2 × 1.1 × 0.7 cm anterior nasal mass with possible tiny central calcification. Magnetic resonance imaging of the
face was also performed. The patient was temporarily treated with phenylephrine for significant nasal congestion and was weaned off oxygen by the third day of life. At the time, she was tolerating appropriate feedings and gaining weight. The patient underwent endoscopic surgical excision of the nasal mass by pediatric otolaryngology without complication. Surgical pathology examination confirmed heterotropic glial tissue (nasal glioma). The patient recovered well with no postoperative feeding or breathing problems.

CASE 2

A 39-week-old gestational age infant was admitted to the NICU immediately after delivery for respiratory distress on continuous airway pressure at 5 cm of water. The NICU team was called to delivery for variable and late decelerations secondary to meconium-stained amniotic fluid. The pregnancy was complicated by maternal obesity, rubella nonimmune status, and oligohydramnios. Physical examination was pertinent for a right upper lid coloboma, preauricular appendage, and large nasal appendage from medial right nares. The infant was gradually weaned off oxygen support and transitioned to all oral feeds. Magnetic resonance imaging of the face demonstrated a 0.9 × 0.7 × 0.8 cm exophytic mass arising from the right nasal fossa along with fat signal. The infant was discharged from the NICU due to stable breathing and feeding and was closely followed as an outpatient by the pediatric otolaryngology department. At 4 months of age, the patient underwent endoscopic surgical excision (Figure 1b) by pediatric otolaryngology without complication. Surgical pathology examination confirmed NLCS. The patient recovered well, with no further breathing or feeding issues.

DISCUSSION

Nasal gliomas account for approximately 5% of all congenital nasal anomalies. The differential diagnoses of midline nasal anomalies that result from atypical embryologic development include nasal gliomas, dermoid cysts, and encephaloceles (7). The word “nasal glioma” is something of a misnomer, as it implies a neoplastic condition (7, 8). In fact, approximately 60% of these nasal gliomas are extranasal; 30% of them are intranasal lying within the nasal cavity, mouth, or pterygopalatine fossa; and 10% are mixed (1, 2). In 20% of cases, gliomas connect to the intracranial space via fibrous stalk (8). Histologically, these tumors are a product of astrocytic neuroglial cells intertwined with fibrous and vascular connective tissue explicitly enclosed with skin or nasal respiratory mucosa.

Midline nasal anomalies should be considered when assessing infants with respiratory distress. Intranasal lesions often present with nasal obstruction, which can sometimes cause life-threatening airway obstruction in infants due to their obligate nasal breathing. When significant nasal obstruction occurs in infants, feeding is also affected; some infants may even require temporary feeding tube support until treatment can take place. A comprehensive physical examination is crucial for diagnosis, and pediatric otolaryngology consultation and appropriate neuroimaging studies should be promptly considered when suspicion exists in order to diagnose and treat the condition effectively. Prompt intervention may be required due to obstructive symptoms.

The treatment of choice is surgical excision, which has a 4% to 10% recurrence rate. A detailed preoperative assessment is essential to demarcate the precise position and extension of the tumor, rule out cranial involvement, and plan an appropriate surgical approach. The traditional approach for an intracranial connection is a frontal craniotomy; other potential approaches may include a transfacial lateral rhinotomy (7). Intranasal endoscopic approaches are increasingly utilized and may be considered alone for intranasal lesions and in combination with other approaches for more complex lesions.

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