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Prenatal detection and postnatal management of an intranasal glioma

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Abstract
Nasal gliomas are rare benign congenital midline tumors composed of heterotopic neuroglial tissue. They have potential for intracranial extension through a bony defect in the skull base. Neuroimaging is essential for identifying nasal lesions and for determining their exact location and any possible intracranial extension. Computed tomography is often the initial imaging study obtained because it provides good visualization of the bony landmarks of the skull base; it is not, however, well suited for soft tissue imaging. Magnetic resonance imaging has better soft tissue resolution and may be the best initial study in patients seen early in life because the anterior skull base consists of an unossified cartilage and may falsely appear as if there is a bony dehiscence on computed tomography. A frontal craniotomy approach is recommended if intracranial extension is identified, followed by a transnasal endoscopic approach for intranasal glioma. A case is presented of a huge fetal facial mass that was shown by ultrasound that protruded through the left nostril at 33 weeks of gestation. Computed tomography of the neonate suggested a transethmoidal encephalocele. Magnetic resonance imaging showed a huge mass occupying the nasopharynx and the nasal cavity and protruding externally to the face but ruled out bony discontinuity in the skull base and, therefore, any intracranial connection. The infant underwent an endoscopic resection of the mass via oral and nasal routes and pathologic examination revealed intranasal glioma.

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1. Case report

A 41-year-old gravida 3 para 2 was referred at 33 weeks of gestation because of an isolated fetal facial mass on ultrasound. It was 5.5 × 3.2 cm in size, mildly heterogeneous, and protruded through the left nostril (Fig. 1). No Doppler flow was present. There was polyhydramnios, but no other associated abnormalities were detected. In particular, the central nervous system (CNS) and the bony structures of the head or face appeared normal. Spontaneous normal delivery of a 1935-g female infant occurred at 35 weeks of gestation at an outside facility. A 6-cm reddish glistering mass was noted protruding through the left nostril of the neonate who was
immediately intubated for mechanical ventilation (Fig. 2). The infant was transferred to our institution on the eighth day of life to be investigated and treated for the facial tumor. Computed tomography (CT) suggested an osseous defect in the ethmoidal portion of the anterior fossa with brain substance herniating to the nasal and oral cavity, leading to the diagnosis of transethmoidal encephalocele (Fig. 3A). Magnetic resonance imaging showed a mass with heterogeneous signal and mild contrast enhancement that occupied the entire left nasal cavity displacing the septum to the right, obliterated the nasopharynx, and extended to the oral cavity. It extended upwards to the ethmoidal portion of the skull base, but there were no osseous discontinuity, intracranial extension, or CNS abnormality. Anteriorly, the mass protruded externally to the face (Fig. 3B). Intranasal glioma was hypothesized. On the 12th day of life, endoscopic excision of the lesion was performed via nasal and oral routes, and the lesion was noted to be attached to the roof of the nasopharynx. Pathologic examination revealed nasal glioma (Fig. 4). In the first postoperative days as the respiratory distress diminished, mechanical ventilation was suspended for 2 days but was then resumed because of a pneumonia that was unresponsive to treatment and progressed to septicemia and death that occurred on the 42nd day of life.

2. Discussion

Congenital nasal masses are estimated to occur in 1 in 20,000 to 1 in 40,000 live births [1]. The main differential diagnosis of congenital nasal masses includes dermoids, hemangioma, glioma, and encephalocele, although in adults, polyps must be considered in intranasal cases [1,2]. Encephaloceles and nasal gliomas are the result of abnormal embryonic development [1]. In encephaloceles, there occurs a protrusion of brain parenchyma and meninges through a bony defect, maintaining a connection with the rest of the brain by a pedicle [3,4]. Nasal glioma, also known as glial nasal heterotopia [3,5], is a congenital midline malformation composed of heterotopic masses of neuroglial tissue in the nose or the nasopharynx [4]. It is thought to be derived from either entrapped neuroectodermal tissue during the closure of the covering of the brain or a nasal encephalocele in which...
the CNS connection has either been absorbed or has become vestigial [2,3,5,6]. Nasal gliomas have thus the potential to extend intracranially by a fibrous stalk [1,6-8].

Approximately 60% of nasal gliomas are extranasal, usually on the bridge or side of the nose; 30% are intranasal; and 10% have extranasal and intranasal components communicating through a defect in the nasal bones [7,8]. Most nasal gliomas are present at birth, but they have been described in patients ranging in age from birth to 81 years [1-3,5]. Fifteen percent to 20% of nasal gliomas may appear with a fibrous stalk extending toward the skull base with an underlying bony defect [1,8]. Imaging studies are required before incision to rule out bony defects and intracranial connection because of the differences in surgical approach involved [4,8,9].

Computed tomography is preferred for visualization of the internal and external bony defects [10]. However, in the first 6 to 8 months of life, the frontal, nasal, and ethmoidal bones are not ossified and have similar CT imaging attenuation as the brain and nasal cartilage, which may give the false impression that there is a bony defect in the frontonasal region [1,8]. Magnetic resonance imaging (MRI) provides more detailed information on the osseous frontonasal region in young infants, the CNS, the tissue characteristics of the mass, and its possible intracranial connection [7,8,10]. Besides, the lack of radiation most importantly in this usually very young patient population [6] and multiplanar imaging capacity favor the use of MRI [7,8]. Encephalocele retains a visible connection with the brain through a bony defect [4] and herniation of meninges alone (meningocele) or brain and meninges (encephalocele) together with cerebrospinal fluid around the lesion is sometimes apparent [1]. Demonstration of a bony defect itself is suggestive of an intracranial connection, which is, however, sometimes not confirmed on surgical exploration [3,7]. It is possible to exclude suspicion of encephalocele by demonstrating absence of bony defect and lack of an anatomical connection between the mass and the CNS [6,11], as occurred in this case.

In patients with a bony defect and intracranial connection of the mass, a frontal craniotomy approach is recommended to prevent cerebrospinal fluid fistula and meningitis [4] followed by a transnasal endoscopic approach for excision of the mass.

**Fig. 3** Postnatal imagings. A, Axial noncontrast CT shows a huge mass occupying the left nasal cavity (*), displacing the nasal septum and protruding externally to the face. Because it was located near the bottom of the anterior cranial fossa, a transethmoidal encephalocele was considered. B, Sagittal T2-weighted MRI demonstrates that the mass occupies the entire nasal cavity (*) and the nasopharynx and extends to the oral cavity. Superiorly, it extends to the ethmoidal portion of the skull base, and anteriorly, it protrudes externally to the face. Thanks to MRI’s higher contrast resolution and multiplanarity, an intracranial connection could be ruled out. Asterisk (*) in B illustrates the equivalent location of asterisk (*) in A.

**Fig. 4** Histopathologic examination showing, under a layer of respiratory epithelium, collagen surrounding the glial tissue (H&E, original magnification ×200).
extracranial-intranasal component of the mass [1]. Biopsy is not advisable because if there is an intracranial connection, it can cause a cerebrospinal fluid leak and meningitis [2-5].

The endoscopic approach is strongly recommended for the removal of intranasal gliomas without intracranial connection because this procedure is associated with less morbidity; it leaves no external wound, and the healing process is short [1,5]. Careful resection is mandatory because recurrences may occur most likely because of incomplete primary excision [4,5].

In reported cases of prenatally diagnosed nasal glioma, the mass arose externally at the side of the nose [6,9,11]. The detection of intranasal glioma was possible in this case because it had extruded through the nostril.

References