Midline Craniofacial Masses in Children

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Abstract

Keywords

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- surgical management

Nasal dermoids, encephaloceles, and gliomas are rare congenital lesions that result from improper embryologic development. The differentiation between them and a firm understanding of their pathology is necessary to avoid unnecessary complications. In view of their potential intracranial connection, prompt diagnosis and treatment are paramount. The authors review the embryology, diagnoses, radiologic work-up, surgical management, and complications of these midline craniofacial masses in children.

The evaluation of midline craniofacial masses in the pediatric population yields a large differential diagnosis that includes inflammatory lesions, posttraumatic deformities, benign and malignant neoplasms, congenital masses, and vascular malformations. Congenital midline craniofacial masses include dermoid cysts, gliomas, and encephaloceles.¹ These lesions are rare, occurring in an estimated 1 in 20,000 to 1 in 40,000 births.^{2,3} Most cases require a multidisciplinary team approach, consisting of plastic surgeons, otolaryngologists, neurosurgeons, neuroradiologists, and pediatric anesthesiologists.

To understand the underlying etiology of these congenital masses, a firm understanding of normal embryological development and anatomy is necessary, especially of the frontonasal region. The fonticulus frontalis and the prenasal space are two transient spaces that allow extension of a dural diverticulum from the anterior cranial fossa to the skin. The fonticulus frontalis temporarily separates the frontal and nasal bones. The prenasal space is located between the nasal bones and developing nasal cartilage. With the progressive growth of the frontal and nasal bones, the dural diverticulum and prenasal space regress, and the cribriform plate forms from fusion of the fonticulus frontalis with the foramen cecum, a foramen that passes through the skull base just anterior to the crista galli. Developmental midline craniofacial masses arise due to failure of these processes to occur, causing incomplete regression and separation of the dura from the overlying skin.

Dermoid Cysts

Dermoid cysts are the most common midline congenital craniofacial lesions, making-up 61% of midline nasal masses in chil-

dren. These lesions are benign and present more often in males than females.⁴ Dermoid cysts are subdivided into two groups based on location: those involving the orbital/periorbital area including the midline lower forehead, and those involving the nasal area. They form most commonly in the upper lateral orbit or the upper lateral orbital rim with a prevalence of 80%; they also occur on the forehead 10% of the time.⁵ Dermoid cysts located in the nasal area make up 5 to 10% of dermoid cysts and have a distinct clinical presentation and etiology. Nasal dermoids develop when the dural diverticulum remains in contact with the overlying epidermis and pulls ectodermal elements with it as it regresses.⁶ A sinus tract forms when the misplaced ectodermal elements recede along with the dura and maintain a connection to the overlying skin. An intracranial connection may subsequently develop if the tract extends beyond the foramen cecum. A cyst, on the other hand, forms when ectoderm gets trapped underneath the skin without a formed connection to the skin surface, causing sebaceous material to build within the cyst wall. Dermoid cysts may present with one or both of these elements; thus, the name “nasal dermoid sinus cyst” (NDSC) is used to refer to nasal dermoids. Histologically, they retain features of normal skin, as the cyst wall is made of stratified squamous epithelium filled with keratinous material. Dermal derivatives such as hair follicles, sebaceous glands, and, rarely, sweat glands may be present. Consequently, hair may emerge through the sinus opening and is diagnostic.⁷ If dermis and skin adnexa are not present within the lining of the lesion, the lesion is termed a simple *epidermoid inclusion cyst*.

Associated congenital anomalies have been reported in up to 41% of cases and include hypertelorism, cleft lip and palate, hemifacial microsomia, aural atresia, hydrocephalus,

branchial sinuses, albinism, cardiac, gastrointestinal, genital, and central nervous system anomalies.^{3,8–10} Although a specific inheritance pattern has not yet been identified, NDSCs tend to cluster within families, and an autosomal dominant inheritance has been proposed.^{10–13}

Nasal dermoid sinus cysts are present from birth; however, they may not require attention until inflammation or infection with subsequent drainage occurs. These lesions can present as a small pit or blind sinus, a fistula extending into nasal structures, or a single or multiloculated nodulocystic lesion.⁷ Cystic lesions present as firm, nonpulsating, non-transilluminating, noncompressible, and nonpainful nodules with or without a sinus tract, which is visible as a small opening in the midline. This external ostium or punctum is frequently seen in nasal dermoid lesions, whereas it is an uncommon feature of lesions in the orbital and lower forehead area. Discharge of sebaceous material from the pit is common, resembling a furuncle.¹⁴ Complications often occur if the lesion goes untreated. These include local abscess formation, periorbital and midfacial cellulitis, and osteomyelitis. When infected, NDSCs lead to induration, and at times, erosion of the skin.¹⁴ Additionally, cysts can cause bone atrophy and skeletal distortion, and patients with an intracranial connection are at risk of developing a frontal lobe abscess, convulsions, meningitis, and meningoencephalitis.⁷ Recurrence or meningitis from incomplete excision of the stalk has been reported in up to 15% of cases.¹⁵

On physical exam, hypertelorism may be present, as NDSCs often distort the contour of the nose, resulting in nasal broadening. These masses can range in size from a few millimeters to a few centimeters; most are between 1 and 2 cm in diameter. They tend to increase in size over time, but do not enlarge with crying or with compression of the jugular veins.

Preoperative imaging with a fine-cut computed tomography (CT) scan through the anterior skull base is crucial to determine the location of the cyst and to rule out the presence of a sinus tract and/or an intracranial connection. A patent foramen cecum and bifid crista galli suggest, but do not confirm an intracranial connection. On the other hand, the lack of these findings rules out the presence of an intracranial connection. Standard findings on CT include low attenuation with calcified extracranial components.¹⁵ Central contrast enhancement is absent for dermoid cysts. Magnetic resonance imaging (MRI) is indicated if CT findings are positive. On MRI, NDSCs present with a hyperintense T1-weighted signal representing liquefied cholesterol contents and a hypointense signal on T2-weighted images.¹⁶ The presence of an intracranial mass is diagnostic for an intracranial connection.^{16,17}

Complete surgical excision at an early age is the only successful treatment strategy. Surgical treatment depends largely on the extent of the possibly associated sinus tract. Lesions located in the orbital and periorbital areas are unlikely to harbor an intracranial connection, and surgery may proceed in a straightforward fashion.

Those involving the nasal area are more likely to contain an intracranial connection, as elicited by imaging. The midline location is ominous for this particular anatomy. A formal craniotomy is indicated if the lesion extends intracranially.¹⁸ If MRI findings are unclear or negative, but CT findings are

Fig. 1 (A) Preoperative radiological findings (computed tomography) of a nasal dermoid sinus cyst. (B) Preoperative appearance.

positive, the surgeon should plan for a possible craniotomy depending on intraoperative findings (► **Fig. 1**).

Encephalocele

An encephalocele is an outpouching of meninges or brain tissue through structural weaknesses in the bony structures of the skull. The mass may contain meninges only (meningocele); meninges and brain (meningoencephalocele); or meninges, brain, and ventricle (meningoencephalocystocele). They occur in an estimated 1 in 5,000 to 1 in 40,000 births around the world and develop due to failed retraction of the dural diverticulum.¹⁹ The surface ectoderm fails to separate from the neuroectoderm during the fourth week of gestation, resulting in a persistent subarachnoid connection via a patent fonticulus frontalis.^{10,19,20} Histologically, they are similar to gliomas and contain interweaving strands of neural and fibrous tissue covered by skin or nasal mucosa. Glial cells, cerebral tissue, nonfunctional neural tissue, choroid plexus, and ependymal cells make up these lesions. The presence of cystic structures with ependymal cells or recognizable leptomeninges helps distinguish encephaloceles from gliomas.^{19,21}

The prevailing location of these lesions depends primarily on geographic area: Posterior encephaloceles predominate in Western Europe, North America, Australia, and Japan; anterior encephaloceles occur more commonly in Southeast Asia and Russia. The majority of cases are located in Southeast Asia.¹⁹ The

etiology is unknown, but involves racial, genetic, environmental, and paternal factors. Viral infection, hyperthermia, irradiation, hypervitaminosis, hypoxia, and salicylate therapy in early pregnancy may also contribute to their development.²²

Encephaloceles are named according to their position on the skull: occipital, anterior, or basal. Each of these groups is further subdivided based on the specific location of herniation.^{23–25} The frontoethmoidal group is a subtype of anterior encephaloceles located in the anterior midline between the frontal bone and ethmoid process. The cranial defect in this group is often clinically visible as a mass along the nose due to its location. Basal encephaloceles, on the other hand, herniate posterior to the cribriform plate into the nasal cavity and are usually not visible externally, but may protrude into the oropharynx or nasopharynx.

Encephaloceles are accompanied by other anomalies in at least 60% of patients. These include hydrocephalus (50%), microcephaly, micrognathia, frontonasal dysplasia, cleft lip/palate, polydactyly, vision problems, and mental retardation, as well as heart, renal, and vertebral issues.^{15,19,20,22,26,27} Encephaloceles may also be associated with Arnold-Chiari II malformation and Dandy-Walker malformation.^{19,22}

Frontoethmoidal encephaloceles typically present as nasal broadening and/or a blue, pulsatile, reducible mass near the nasal bridge that transilluminates. They may be seen at the glabella, over the nose, or on the forehead.²⁸ They are often noted early in life due to the resultant deformity and may be misdiagnosed as a cavernous hemangioma, polyp, or hypertelorism.²⁹ Correct diagnosis, however, is crucial, as these lesions may cause cerebrospinal fluid (CSF) rhinorrhea or recurrent meningitis. They tend to enlarge with crying, the Valsalva maneuver, or compression of the jugular veins.¹⁴ A positive Furstenberg's sign suggests an intracranial connection.¹⁷ Basal encephaloceles, unlike frontoethmoidal encephaloceles, often present with upper-airway obstruction. Furthermore, endocrine dysfunction or visual abnormalities may result from herniation in certain subtypes of basal encephaloceles. Snoring or nasal purulent drainage are other common symptoms.¹⁹

Computed tomography in conjunction with MRI is used to evaluate these lesions. CT is useful to detect osseous defects, bone anatomy, and interorbital distance.^{19,30} Three-dimensional CT is superior and demonstrates the entirety of the skull base defect.¹⁹ An enlarged foramen cecum, bifid crista galli, or frontal bone defect indicate an intracranial connection.⁶ MRI helps visualize contents within the sac and reveals brain abnormalities. A soft-tissue mass in connection with the subarachnoid space is seen on MRI if an encephalocele is present. The lesion is isointense relative to gray matter with most MRI sequences, but may be hyperintense with T2-weighted sequences due to gliosis.³¹ As both studies are costly, some recommend using MRI as the initial imaging study. In possible meningoencephaloceles, digital angiography or angioresonance should be ordered to evaluate for the presence of vascular structures.³² Biopsy is always contraindicated due to the persistent intracranial connection.

Treatment is surgical and is always done early in life to allow the developing brain and eyes to remodel the facial deformity as well as to prevent further damage and infection. The location,

extent, origin of the mass, and presence of pulsation may be evaluated by nasal endoscopy.¹⁹ Several surgical approaches and techniques have been successfully employed and usually vary based on the specific type of encephalocele encountered.^{19,33} In every case, it is crucial to obtain a tight dural closure and skin closure to prevent infections and desiccation of brain tissue. Furthermore, the removal of excessive nonfunctional extracranial tissue should be performed. The reconstruction of any craniofacial deformities may then take place.²⁵

Hydrocephalus may occur postoperatively after the resection of a large resorptive area and may result in respiratory arrest. Cardiac arrest has also been reported secondary to associated brainstem dysfunction and apnea from central hypoventilation caused by structural derangement of the medullary respiratory control center or its afferent or efferent pathways. The lack of pharyngeal coordination, a poor sucking reflex, and an absent gag reflex may also result in gastric aspiration.²² The patient must be carefully monitored postoperatively due to these potential complications.

The absence of brain tissue within the herniated sac is the most important factor signifying good prognosis. However, the prognosis also depends on location. For example, frontal encephaloceles are associated with normal intelligence and motor development, and outcomes are generally good. Occipital encephaloceles, however, have higher rates of hydrocephalus and seizure. Finally, gross brain herniation or extruded brain that is damaged and gliotic portends a poor prognosis.³⁴

Gliomas

Gliomas are the least common congenital midline mass.^{28,35} The male-to-female ratio is 3:2 with no familial predisposition.^{21,36} Despite their name, these lesions do not represent true neoplasms and are sometimes referred to as encephalomas, nasal cerebral heterotopias, or neuroglial heterotopias. They resemble encephaloceles histologically, but occasionally contain focal areas of calcification.³⁷ Their embryologic origin is also similar to encephaloceles, except that they lack an intracranial connection in 70% of cases.¹⁶

They are classified into three categories based on location: intranasal (30%), extranasal (60%), and combined (10%).³⁸ Rarely, they may advance into the orbit, frontal sinus, oral cavity, or nasopharynx.²¹ Intranasal gliomas present as a pale mass in the nasal cavity with protuberance from the nostril. The base most often originates from the lateral wall near the middle turbinate and occasionally from the nasal septum. Extranasal gliomas, on the other hand, present as firm, noncompressible, smooth masses that appear anywhere from the nasal tip to the nasal glabella. At times, skin telangiectasias overlie the surface of the mass.³⁹ The nasal bridge may appear widened and the eyes wide-set.²¹ Gliomas do not exhibit pulsation or expansion with crying, coughing, or straining. The Furstenberg test is negative. Complications include nasal obstruction, infection, and deformation of the nose.²¹

Gliomas may be diagnosed prenatally via fetal ultrasound or MRI.³⁴ After birth, CT and MRI studies may be used to differentiate gliomas from other congenital midline masses. The lesion is isoattenuating on CT and can be differentiated

Fig. 2 (A, B) Preoperative radiological findings (magnetic resonance imaging) of a nasal glioma. (C, D) Preoperative appearance. (E) Intraoperative removal of the sinus tract. (F) Postoperative appearance.

from encephaloceles through CT with contrast, as the absence of contrast flow into the nasal mass excludes the possibility of a connection with the subarachnoid space.³⁰ On MRI, gliomas are isointense to hypointense relative to gray matter with T1-weighted images, hyperintense on T2-weighted imaging, and nonenhancing to slightly enhancing with contrast.^{16,29,31} There is an absence of extracranial continuity of CSF flow.⁴⁰

Surgical management is initiated as early as possible to minimize the chances of complications. Techniques involving

an external excision are utilized in the treatment of extra-nasal gliomas.²¹ The surgeon must remain aware that 10 to 25% of nasal gliomas may present with a fibrous stalk extending deep to the nasal bones toward the base of the skull.³⁷ In this case, a nasal osteotomy helps improve exposure. If the initial approach fails to adequately repair the intracranial extension, a frontal craniotomy is indicated.²¹ A transnasal approach is recommended for the resection of intranasal gliomas, as this method allows proper exposure

and complete excision while minimizing any postoperative facial or nasal deformity. In summary, many factors must be taken into account prior to determining the most appropriate surgical approach, including location and size of the mass, associated cartilage or bony deformity, and the experience of the surgeon. Furthermore, surgical treatment should accomplish three goals: complete excision of the mass, exploration of a possible stalk toward the base of the skull, and a good cosmetic outcome.²¹ Recurrence rates range from 4 to 10% after surgery (■ Fig. 2).^{37,41}

Conclusion

Dermoid cysts, gliomas, and encephaloceles make up congenital midline craniofacial masses. Although all three develop in a similar fashion embryologically, the presentation, diagnostic strategy, and treatment method differ for each lesion.

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