

TITLE: Evaluation of Congenital Midline Nasal Masses

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Introduction

Congenital midline nasal masses include nasal dermoids, nasal gliomas, and encephaloceles. These are rare congenital anomalies, estimated to occur in 1:20,000 to 40,000 births. Although rare, these disorders are clinically important because of their potential for connection to the central nervous system. Biopsy of a lesion with an intracranial connection can lead to meningitis or cerebrospinal fluid leak. Preoperative knowledge of an intracranial connection is a necessity to allow for neurosurgical consultation and possible planning for craniotomy. The differential of a midline nasal mass includes inflammatory lesions, traumatic deformity, benign neoplasms, malignant neoplasms, and congenital masses.

Embryology

Knowledge of the normal embryological development of the nose is important to understand the development of congenital midline nasal masses. The development of the nose depends on two crucial steps which occur during the first twelve weeks of fetal development. The first is the formation of the neural tube from the neural fold, which occurs at the dorsal aspect of the embryo between the third and fourth week of gestation. Closure of the neural groove begins in the middle of the embryo and extends in a cranial and caudal direction. The neural tube then gives rise to the neural crest cells. As the neural tube is forming by closure of the neural groove, neural crest cells migrate laterally and anteriorly around the eye to the frontonasal process. In most of the body the neural crest cells are involved in forming ectodermal components, however in the face neural crest cells primarily form mesenchymal cells which provide the bone, cartilage, and muscles of the face. The second crucial step in the development of the nose is the correct migration of these cells into the mesenchyme.

The nose is formed from the frontonasal process and two nasal placodes which develop dorsal to the stomadeum (primitive mouth). The nasal cavity is formed from the invagination of the nasal placodes that appear during the third week of gestation, and is completed around the sixth week, separated posteriorly from the oral cavity by a thin nasobuccal membrane. The two nasal cavities are separated by the nasal septum, which is formed from the migration of neural

crest cells into the frontonasal process, growing in an anterior-posterior direction. The ethmoid and sphenoid bones are also formed from the frontonasal process neural crest cells.

The nasal placodes become more prominent and consist of a medial and lateral process. The medial processes approach one another and eventually fuse in the midline. The lateral processes become less prominent as the maxillary process fuses with them. A deep groove in this region, called the nasal-maxillary groove becomes the nasolacrimal duct.

During formation of the skull base and nose, the mesenchymal structures are formed from several centers which will eventually fuse and begin to ossify. Before their fusion there are recognized spaces between these structures which are important in the development of congenital midline nasal masses. These include the fonticulus frontalis, the prenasal space, and the foramen cecum. The fonticulus nasofrontalis is the space between the frontal and nasal bones. The prenasal space is between the nasal bones and the nasal capsule (the precursor of the septum and nasal cartilages). During fetal development these spaces are normally closed by fusion and ossification. Abnormal development of these structures is thought to be involved in the formation of dermoids, gliomas, and encephaloceles of the nose.

Pediatric Anatomic Considerations

Neonates are obligate nasal breathers, due to the high position of the larynx in the neonate and physiologic pattern of breathing and swallowing. The epiglottis abuts the nasal surface of the soft palate, forming an anatomic divide between the airway and digestive systems. Food from the oral cavity is shunted laterally into the esophagus via the pyriform sinuses, whereas air from the nasal cavity is allowed to pass centrally, directly into the larynx. Functionally, this allows the neonate to breathe and feed concurrently. As the child grows, the pharynx elongates and the laryngeal complex descends into the neck, which results in a common pathway that the respiratory and digestive tracts share in the oropharynx. Although oral respiration can now occur, the nasal passages remain the primary airway. It is because of this feature that neonates can suffer from respiratory distress with nasal obstruction.

Evaluation of nasal masses

The evaluation and management of congenital midline nasal masses starts with a complete history and physical exam. Many of these lesions present early in life but adults may also be found who are undiagnosed with these lesions. Dermoids often can present with repeated infection or drainage, a visible sinus tract, and are more solid, noncompressible, and do not transilluminate. Nasal gliomas are also firm, noncompressible, and do not transilluminate but may have overlying telangiectasia. Encephaloceles may be bluish or red, soft, compressible, enlarge with crying, and have a positive Furstenberg test. With intranasal lesions, gliomas arise from the lateral wall while encephaloceles arise more medially. According to Haafiz et al (1995), an intranasal probe can often be passed medial to a glioma but not to an encephalocele. The distinction between glioma and encephalocele is important because while 15% of gliomas have an intracranial connection, all encephaloceles have an intracranial connection.

When a dermoid, glioma, or encephalocele is a suspected diagnosis a biopsy should not be performed before an intracranial connection is ruled out because of the risk of causing

meningitis or CSF leak. The majority of these lesions are found in children, and a high index of suspicion is required, especially for a unilateral intranasal mass. The diagnosis is confirmed by CT and/or MRI imaging. Image findings include soft tissue mass, fluid filled cyst, intracranial mass, enlargement of the foramen cecum, and distortion of the crista galli. CT imaging better delineates bony abnormalities while MRI is valuable to identify an intracranial connection. The findings on CT consistent with intracranial involvement are an enlarged foramen cecum or bifidity of the crista galli. Although these findings are consistent with intracranial involvement they are not diagnostic. According to Pensler et al (1988), these findings are only conclusive if they are absent, eliminating an upward intracranial connection. MRI provides better soft tissue detail and ability to visualize in the sagittal plane. Denoyelle (1997) reviewed thirty-six children with nasal dermoid sinus cysts and recommends an MRI scan to confirm any suspected intracranial extension following a CT scan. In his series two patients had false positive CT scan evidence of intracranial connection which was not found at surgery. Another study by Huisman et al (2004) showed that intracranial extension is equally well detected by CT and MRI using indirect imaging signs (bifid or deformed crista galli, widened foramen cecum, defect in cribriform plate) but that with direct imaging signs (identification of intracranially located lesions or sign alterations) MRI was superior, detecting these findings on 2 patients not detected on CT.

Dermoid Sinus Cysts

Dermoid sinus cysts of the nose present as a midline nasal pit, fistula, or infected mass located anywhere from the glabella to the nasal columella, with the distal one-third of the nasal dorsum being the most common site. If composed solely of epidermal tissue, then dermoid cysts are referred to as epidermoid cysts. The lumen of dermoid cysts is made up of a mixture of keratin and lipid. Nasal dermoid sinus cysts are firm, non-compressible, non-pulsatile masses, and do not transilluminate. Usually, nasal dermoids terminate in a single subcutaneous tract which can sometimes have hair at the opening. They may secrete sebaceous material or pus, become intermittently inflamed, form an abscess, cause osteomyelitis, broaden the nasal root or bridge, lead to meningitis, or form a cerebral abscess. Connection with the central nervous system has been variably reported to occur. In the case of intracranial extension, the sinus traverses either the cribriform plate or the foramen cecum and is attached to the dura or it can extend in the form of a cyst within the falx cerebri or other brain structures. Faulty closure of the anterior neuropore results in a defect in the anterior fontanelle, foramen cecum, cribriform plate, sphenoid and ethmoid bones. Suspicion of intracranial involvement should remain high.

Associated congenital anomalies occur in 5-41% of cases which include aural atresia, mental retardation, spinal column abnormalities, hydrocephalus, hypertelorism, hemifacial microsomia, albinism, corpus callosum agenesis, cerebral atrophy, lumbar lipoma, dermal cyst of the frontal lobe, coronary artery anomaly, cleft lip and palate, tracheoesophageal fistula, cardiac, genital, and cerebral anomalies. There is no known syndromic association of these anomalies.

A widely accepted theory of dermoid sinus cyst development is the prenasal space theory. According to this theory, during normal development a projection of dura protrudes through the fonticulus frontalis or inferiorly into the prenasal space. This projection normally regresses but if it does not the dura can remain attached to the epidermis and result in trapped ectodermal elements. Thus development of dermoids is hypothesized to result from faulty involution of the dural tract.

Gliomas

Gliomas are made of neuroglial elements consisting of glial cells in a connective tissue matrix with or without a fibrous connection to the dura. There is no fluid filled space connected to the subarachnoid space. These lesions usually present as a red or bluish lump at or along the nasomaxillary suture, or as an intranasal mass. They are characteristically firm, noncompressible, do not increase in size with crying, and do not transilluminate. The overlying skin may have telangiectasias. They can be associated with a widened nose or with hypertelorism secondary to growth of the mass. Intranasal gliomas most often arise from the lateral wall of the nose or less often from the nasal septum. Sixty percent are extranasal, 30% intranasal, and 10% are both. Overall, 15% are connected to the dura. The intranasal type is more often associated with dural attachment (35%) than the extranasal type (9%).

The embryological development of nasal gliomas is similar to nasal dermoids. Thus, if glial tissue is also isolated extracranially by fusion of the cranial sutures, it is hypothesized that a glioma results. Abnormal closure of the fonticulus frontalis can lead to an ectopic rest of glial tissue being left extracranially. This is similar to the mechanism for the formation of encephaloceles, however there is not always an intracranial connection to a glioma and there is always, by definition, an intracranial connection to an encephalocele.

Encephaloceles

Encephaloceles are extracranial herniations of the meninges and/or brain which maintain a subarachnoid connection. If it contains only meninges it is termed a meningocele, when it also contains brain tissue it is called a meningoencephalocele. Thus, when a bony defect allows herniation of dura mater and brain tissue extracranially, an encephalocele results. Encephaloceles can be divided into three categories: occipital, sincipital, and basal. Occipital are the most common at 75%. Sincipital are frontonasal lesions which present as a mass over the nose, glabella, or forehead. The intracranial connection is usually anterior to the cribiform plate. Basal lesions make up about 10% of lesions and present as an intranasal or nasopharyngeal mass. Basal lesions herniate either through the cribiform plate or posterior to it which explains their presentation in the nose instead of externally.

They enlarge with crying or the Valsalva maneuver. A characteristic sign is the Furstenberg test, which is enlargement with compression of the internal jugular veins. They also can cause a widening of the nose or hypertelorism. Intranasal encephaloceles originate medially in the nasal cavity as opposed to gliomas which most often originate laterally.

The embryologic development of encephaloceles is the same as that for gliomas. Failure of the fonticulus frontalis to close properly can lead to a herniation of intracranial contents which maintains its connection to the subarachnoid space. This connection with the central nervous system and the possibility of containing brain tissue make encephalocele an important entity to rule out when a midline nasal mass is found.

Developmental Considerations to Surgical Planning

Manning et al (2005) described anatomic and developmental constraints that influence

treatment options for pediatric patients with skull base pathology depending on the age of the child. They discussed that when considering surgical approaches, one must keep in mind the importance of primary growth centers such as tooth buds, nasal septum and palate, and the zygomatic process of the maxilla. The cranial vault is fairly well developed at birth, but the basicranium and facial skeleton are relatively undeveloped and undergo rapid growth in the first few years of life.

The sinuses are relatively undeveloped at birth and do not afford surgical access to the skull base in the first few years of life (although improving technology is pushing back the minimum age for endoscopic approaches). The ethmoid cells are recognizable by the fifth fetal month and may provide some surgical access to the anterior cranial fossa (with 2.8-mm endoscopes) by age 1 year. Sphenoid pneumatization is highly variable, but the minimum age for an endoscopic optical environment to the middle cranial skull base is probably about age 3 years, on average. The frontal sinuses are usually not radiographically apparent before age 6 to 8 years, and approximately 8% of the population has no significant frontal pneumatization in adulthood.

Maxillary sinus approaches to the pterygomaxillary fossa and middle cranial fossa may be limited by small sinus size and by presence of molar tooth buds before the teenage years. (Manning, 2005)

Surgical Treatment

The treatment of nasal dermoids, gliomas, and encephaloceles is by complete surgical excision. Early surgical intervention is recommended to avoid further distortion of the nose or bony atrophy caused by growth of the mass or recurrent inflammation. The entire lesion along with any fistulous tract must be excised in order to prevent recurrence. The key information necessary for surgical planning is the presence of an intracranial connection to the mass.

Ingraham et al (1943) suggested an external rhinoplasty approach for dermoid lesions that do not extend above the glabella and many surgeons still prefer this approach. The transverse rhinotomy is used with small to moderately sized lesions with no evidence of intracranial extension. The benefit of this approach is a favorable scar without the splaying which can occur with a vertical rhinotomy. The fistulous opening is excised within a transverse fusiform segment of skin and the tract is cannulated with a lacrimal probe. A second transverse incision is then made over the lower half of the dermoid, and the entire tract is excised. Medial or lateral osteotomies may be performed as necessary for exposure.

For high lesions, or those with a small suspected intracranial extension based on imaging studies, a midline incision over the dorsum is preferred, according to Manning et al (2005). For lesions with a stalk extending superiorly, the nasal bones are displaced laterally, and the stalk is dissected from inside the nasal septum. With larger lesions, especially in the lower two-thirds of the nose a tripod-eversion rhinotomy approach is used. A transverse incision is made to release the columella, a transfixion incision is made and swept laterally between the upper and lower lateral cartilages. Paraalar incisions will then permit upward rotation of the nose. Any fistulous tract opening is released with a fusiform incision and the tract cannulated with a lacrimal probe. Microscopic visualization and otologic instruments can be of great value in dissection of these lesions. Small CSF leaks can be managed directly, without a craniotomy. A craniotomy is

required for a lesion with extensive intracranial connection, often in a dumbbell configuration with an intracranial cyst above the stalk. With craniotomy approaches to these lesions, however, olfaction must be sacrificed at least on one side.

Meher et al (2005) described a case of the management of a dermoid cyst with intracranial extension, not requiring craniotomy. They described that, after taking neurosurgical opinion, the swelling was excised by a vertical midline incision. The sac of the swelling was found to be going superiorly through a tunnel between the nasal bones and the underlying nasal septum. The nasal bones were removed along with the adjacent anterior part of the frontal bone. Intracranial extension through the cribriform plate was identified. The wall of the sac was incised and, after evacuating the contents, it was removed except for its base where it was attached to the dura. The secretory epithelial surface of the remnant of the sac was destroyed by bipolar electrocautery. The nasal bones were replaced and the wound was closed in layers. They stated that the postoperative period was uneventful and that there was no recurrence during a two year follow-up period.

Manning et al (2005) mention that recent reports in the literature show that an endoscopic intranasal approach for intranasal gliomas even with a central connection is possible, assuming surgeon experience with endoscopic repair of small CSF leaks, this includes studies by (Yokoyama et al (1999), Rahbar et al (2003), and Agirdir et al (2004). This is also limited by the age of the patient. Approach for surgical excision is based on imaging diagnosis of possible CSF connection, location of the pathology, and surgeon experience.

Intranasal lesions are approached via lateral rhinotomy or more recently described endoscopic techniques. Weiss et al (1998) described the use of endoscopic removal of nasal dermoids in two cases. They recommend the use of this technique when the dermoid is located within the nasal cavity and there is little or no cutaneous involvement. They recommend the endoscopic technique even with extension to the anterior cranial fossa only recommending a combine intra-extracranial approach when the mass extends to the falx cerebri. Other authors such as Burkhardt et al (1999) and Dimov et al (1989) have described excision of nasal gliomas isolated in the nasal cavity without evidence of intracranial connection by imaging.

Sincipital (anterior or frontoethmoidal) encephaloceles have a facial component and typically require a combined approach with craniotomy for repair and reconstruction. The rare basal encephaloceles present as a nasal mass with the potential for airway obstruction or meningitis. The two most common imaging findings with basal encephaloceles are foramen cecum defects with variable extension into the ethmoid roof and cribriform or isolated ethmoid roof defects with low-lying funnel-shaped anterior skull base anatomy (Woodsworth et al, 2004).

Traditional approaches to basal encephaloceles involved an anterior craniotomy with repair of the skull base defect with pericranial flaps. Management is shifting in many centers to endoscopic nasal approaches even for large encephaloceles in young patients (Marxhall et al, 2001). The encephalocele itself tends to create more intranasal space by deflecting the septum and middle turbinate, although angled telescopes, possibly including 70° scopes, may be necessary to visualize anterior connections. Generally, the intranasal portion of the encephalocele is ablated progressively with bipolar cautery until the skull base defect is seen. The mucosal cuff is removed circumferentially around the bony defect, which is then repaired

endoscopically.

Conclusion

Congenital midline nasal masses include nasal dermoids, nasal gliomas, and encephaloceles. These are rare congenital anomalies, estimated to occur in 1:20,000 to 40,000 births. Although rare, these disorders are clinically important because of their potential for connection to the central nervous system. Biopsy of a lesion with an intracranial connection can lead to meningitis or cerebrospinal fluid leak. The treatment of these masses is surgical excision. Preoperative knowledge of an intracranial connection allows for neurosurgical consultation and planning for craniotomy. Surgical strategy depends on the location and extent of the lesion, ranging from local excision via an open or endoscopic approach, to a combined intracranial-extracranial approach.

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