

TITLE: Congenital Midline Nasal Masses

SOURCE: Grand Rounds Presentation, UTMB, Dept. of Otolaryngology

DATE: November 6, 2002

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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. The treatment of these masses is surgical excision. Preoperative knowledge of an intracranial connection allows for neurosurgical consultation and planning for craniotomy. The differential of a midline nasal mass includes inflammatory lesions, traumatic deformity, benign neoplasms, malignant neoplasms, and congenital masses. Dermoid sinus cysts present as a mass on the dorsum of the nose or intranasally, with a pit or sinus tract opening on the nasal dorsum, hair around the external opening, and discharge of pus or sebaceous material. Nasal gliomas are firm masses which are nonpulsatile, present on the nasal dorsum and/or arise from the lateral nasal wall, have telangiectasias of the overlying skin, and do not enlarge with bilateral compression of the internal jugular veins (Furstenberg test). Encephaloceles may present as nasal broadening and/or as a blue, pulsatile, compressible mass near the nasal bridge which transilluminates, enlarges with crying or with bilateral compression of the internal jugular veins, or as an intranasal mass arising from the cribriform plate. To understand the development of congenital midline nasal masses, knowledge of the normal embryological development of the nose is important.

Embryology

The most critical period in the embryology of the face is during the first twelve weeks of fetal development. Between the third and fourth weeks of fetal growth the neural fold develops and forms the neural tube on the dorsal aspect of the embryo. Closure of the neural groove begins in the middle of the embryo and extends in a cranial and caudal direction. A key element in understanding development of the face, including the nose, is the importance of 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 neural

crest cells in the face primarily form mesenchymal cells which provide the bone, cartilage, and muscles of the face.

The nose is formed from the frontonasal process and two nasal placodes which develop dorsal to the stomadeum (primitive mouth). 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. As the external nose is developing, other neural crest cells migrate through the frontonasal process to form the posterior septum, ethmoid bone, and sphenoid. The nasal septum develops around week five from the frontonasal process, growing in an anterior-posterior direction.

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 elements 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. This will be discussed with each particular anomaly later in this paper.

Dermoid Sinus Cysts

Nasal dermoid sinus cysts are the most common of the congenital midline nasal masses. Many present at birth but some are not found until later in childhood or even adulthood when they become symptomatic. They can occur as an isolated cyst or with a sinus tract opening to the skin. They constitute 1-3% of all dermoids but 3-12% of dermoids of the head and neck. In one study dermoid and epidermoid cysts together accounted for 61% of nasal masses in a review of 109 cases. Dermoids are related to epidermoid cysts but contain both ectodermal and mesodermal elements (adnexal structures such as hair follicles and sebaceous glands).

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. 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 from 4-45%. 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.

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%). They are more common in males by a 3:1 ratio although the significance of this has not been established.

The embryological development of nasal gliomas is similar to nasal dermoids. 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 by definition an intracranial connection to an encephalocele. This will be further discussed in the following section.

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. Ingraham and Matson (11) divided encephaloceles 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 cribriform plate. Suwanwela and Suwanwela (21) divided nasal encephaloceles into nasofrontal, nasoethmoidal, and naso-orbital lesions based on the projection of the mass between the nasal and frontal bones, along the side of the nose, or into the medial orbit. He reported that nasofrontal encephaloceles occur directly anterior with short necks and could possibly be excised via an external approach while nasoethmoidal and nasoorbital have long necks and necessitate intracranial closure. Basal lesions make up about 10% of lesions and present as an intranasal or nasopharyngeal mass. Basal lesions herniate either through the cribriform plate or posterior to it which explains their presentation in the nose instead of externally. They rare at 1:35,000 live births in Western Europe, America, Australia, Japan, China, and India but more common at 1:6000 live births in Southeast Asia and Russia. Encephaloceles are often bluish, soft, compressible masses which

can be transilluminated. 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 subarchnoid 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.

Evaluation

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, 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, an intranasal probe can often be passed medial to a glioma but not to an encephalocele(9). 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., these findings are only conclusive if they are absent, eliminating an upward intracranial connection(15). MRI provides better soft tissue detail and ability to visualize in the sagittal plane. Denoyelle 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(6). Bloom et al. reviewed ten patients with nasal dermoids, reporting CT results falsely positive for intracranial extension in one of six cases and indeterminate in one of six cases. Due to the increase cost of two tests, delay in diagnosis, and added risk of additional anesthesia for additional imaging, they recommend MRI as the initial imaging study (1). Schlosser et al., reported three cases of the use of preoperative three dimensional CT scanning. They found it useful in a case of an encephalocele with a large anterior cranial floor defect to demonstrate the full extent of the defect and to provide images to

use in counseling of the parents more easily understood than conventional two dimensional CT (19).

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. Other complications are abscess formation, osteomyelitis, and meningitis with those lesions with an intracranial connection. The entire lesion along with any fistulous tract must be excised in order to prevent recurrence. Denoyelle et al. reported a recurrence rate of 5.5% (two of thirty-six patients) for nasal dermoids in their series, both with an external rhinoplasty approach(6). The key information necessary for surgical planning is the presence of an intracranial connection to the mass.

Pollock(16) reviewed the surgical treatment of the nasal dermoid cyst and recommended four criteria for a surgical approach. First, the surgical approach should permit access to all midline cysts and should readily permit medial and lateral osteotomies, if required. Second, the surgical exposure should favor the rapid repair of cribriform defects, should they be present, and would permit control of CSF rhinorrhea, if it develops. Third, it would allow reconstruction of the nasal dorsum, if it is required. Fourth, the approach should offer the probability of acceptable scar formation. The approaches recommended are the transverse rhinotomy, the vertical zig-zag rhinotomy, and tripod-eversion rhinotomy. 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. 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. The operating microscope is used to improve visualization. Very large lesions, those in adults in whom underlying bone and cartilage have been damaged by prior surgery or erosion, and in patients with known intracranial extension a longitudinal zig-zag rhinotomy is used for wide exposure. The incisions are designed with limbs extending superiorly at angles greater than forty degrees but less than ninety degrees. Any fistulous opening is excised by fusiform excision. Scar prognosis is better than with a straight incision because the zig-zag are at less than ninety degrees to the relaxed skin tension lines running horizontally across the nose. Rohrich et al recommended an open rhinoplasty approach with a stair-step columellar incision in the majority of cases for the following reasons: ease of exposure, wide exposure of the nasal dorsum, controlled external osteotomies, ease of dorsal reconstruction, and wide exposure of the upper lateral cartilages and septum(18).

Intranasal lesions are approached via lateral rhinotomy or more recently described by endoscopic techniques. Weiss et al (26) 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. This technique can be combined with a small external midline excision of a cutaneous punctum. 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 have described excision of nasal gliomas isolated in the nasal cavity without evidence of intracranial connection by imaging (3,7,26).

Midline masses which have an intracranial connection will require a combined approach with a neurosurgeon. A frontal craniotomy is performed, the intracranial portion of the mass is excised and the bone-dura defect is repaired, and the extracranial mass is removed.

Some authors have advocated the use of intraoperative frozen section examination of the stalk of dermoids where it cannot be followed to the base. If only fibrous tissue is found, the stalk is ligated, if dermoid tissue is found an intracranial approach is performed (6).

Conclusion

The diagnosis of congenital midline nasal masses requires a high index of suspicion. These are rare anomalies, however the life threatening complications of those with an intracranial connection is important to remember. When a patient presents with a mass consistent with nasal dermoid, glioma, or encephalocele, especially a child, biopsy of the lesion should not be performed before imaging is obtained. CT can be used initially with follow up MRI for inconclusive results or evidence of intracranial connection or MRI may be used as the primary imaging study. Treatment of these lesions is surgical excision via an external or intracranial approach or both.

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