INTRODUCTION

Congenital nasal masses reflect a small grouping of diseases whose origin stems from an embryological accident. While infrequent, they are an important topic as neonates are obligate nasal breathers and therefore readily demonstrate difficulty in response to the obstruction. In this lecture, we will begin with a review of common causes of pediatric nasal obstruction as well as a review of embryology. Then, a more in-depth discussion of nasal dermoids, nasal gliomas, encephalocele, nasolacrimal duct cysts, and Thornwaldt cysts will follow. Finally, we will conclude with a few brief take home points.

As with any major symptom, the causes can be multitude. A list of differential diagnoses should include congenital, neoplastic, infectious, traumatic, and inflammatory causes. In any young child, a foreign body should also be considered in the differential. The box below from Gnagi SH et al lists a thorough grouping of common causes. In this lecture, we will dwell on the congenital lesions but remembering a thorough differential is key in evaluation of these patients. We will next transition to a review of pertinent embryology.
EMBRYOLOGY

The embryology review begins with the neural tube which forms between the third and fourth week of gestation. The closure of the neural tube occurs from the midline and extends and both cranially and caudally. The neural tube then gives rise to the neural crest cells. As the neural tube closes, neural crest cells migrate anteriorly and laterally around the eyes to the frontonasal process. The nose then forms from the medial and lateral prominence with invagination of the nasal pit. The anterior neuropore (aka the primitive frontonasal region) develops medial to the optic recesses in the third week of fetal life. A transient fontanelle exists between the inferior frontal bone and the nasal bone which is called the fonticulus frontalis.
A funnel-shaped dural projection extends inferiorly and anteriorly through a midline opening anterior to the crista galli of the ethmoid bone. This anterior skull base opening is the foramen cecum. The dural diverticulation extends inferior and posterior to the frontal and nasal bones and superior and anterior the nasal cartilage (known as the pre-nasal space). The apex of the dural diverticulation temporarily approximates the subcutaneous region of the mid-nasal bridge at the osteocartilaginous junction. Failure of involution at these sites where surface and neural ectoderm approximate each other can lead to anomalous development – nasal dermal sinus, encephalocele, and nasal glioma.
NASAL DERMOID CYST

Dermoid cysts are infrequent findings and even rarer in the head and neck. Fewer than 10% of dermoid cysts occur in the head and neck region with nearly 75% of those within the nasoglabellar region. They may be noted immediately at birth or usually within the first few years of life. Very rarely, one may not be noted until the first couple decades of life. Embryologically, the cyst contains squamous cell epithelium with epidermal appendages. These epidermal elements are displaced during intramembranous growth of the nasal bones. Dermoid cysts are typically located in the fronto-temporal region, orbital region, or most commonly the nasoangular region. During development, dura projects thorough the foramen cecum and attaches to the skin. The tissue is supposed to then separate from the nasal skin and retract thorough the foramen cecum. If there is a persistent attachment to the underlying fibrous tissues, nasal capsule, or dura, epithelial elements are trapped and remain in the pre-nasal space.
Up to 50% of nasoglabellar cysts may have a fistula or a sinus tract. The tract traverses via the cribiform plate or the foramen cecum. The tract attaches to dura, falx cerebri, or other intracranial structures. The typical differential diagnosis for a nasoglabellar dermoid cyst is a nasolabial cyst, premaxillary cyst, nasopalatine cyst, and Jacobson organ cyst.

NASAL ENCEPHALOCELE

Nasal encephaloceles consist of extracranial protrusions of meninges, CSF, and neural tissue. Meningoceles are similar except there is no herniation of brain tissue. It may present as an internal or external nasal mass. It is described by the location of dehiscence in the skull base. Nasal encephaloceles present as pale, compressible, pulsatile masses which transilluminate with light. The masses are also positive on Furstenberg’s test which is expansion of the mass with compression of the internal jugular vein. In these children, the mass may also expand by crying or straining.
If the encephalocele protrudes through the fonticulus frontalis, it is termed a sincipital encephalocele; whereas, if it is through the foramen cecum, it is a basal encephalocele. For all these nasal masses, radiological assessment is integral. MRI is the preferred modality both for diagnostic confirmation as well as pre-operative planning, including determining the role for neurosurgical intervention.

NASAL GLIOMA

Nasal gliomas are basically an encephalocele without the intracranial connection. It is also known as a benign congenital nasal neuroectodermal tumor, nasal cerebral heterotopia, and simply as glial heterotopia. While there is no agreement on the exact method by which they arise, several hypotheses exist. Some suggest that they develop from extracranial rests of glial tissue. Some others suggest that there is abnormal closure of the fonticulus frontalis. A third theory is that they are possibly encephaloceles which eventually lost their CSF connection. Nasal gliomas grow along with the child and nearly 15% have a fibrous stalk remnant. The gliomas show dysplastic neuroglial tissue and fibrovascular tissue on pathology. The glabella is the most common site of a nasal glioma although they can be close to the medial canthus. They are typically identified at birth or early in infancy. They are well-circumscribed, bluish or reddish, and telangiectatic on surface. Similar to encephaloceles, they can be sincipital or basal as well. About 30% of nasal gliomas have an intranasal presentation with the lateral nasal wall the most common, followed by middle turbinate and nasal septum. As with the other two midline nasal masses, radiological assessment, preferentially with MRI, is the key, both to confirm diagnosis and better visualize a stalk if present.
NASOLACRIMAL DUCT CYST

Nasolacrimal duct cysts are frequent findings in neonates. Various articles suggest that they may be present in as high as 30% of patients. Most cases tend to resolve by 1 year of age and therefore may not be diagnosed if a child outgrows it. Cysts – Dacrocystoceles – arise from proximal and distal obstruction. Nasal obstruction leads to formation of the cyst as the distal valve of Hasner becomes obstructed. The distal obstruction may even resolve spontaneously with nasal respirations. In approximately 14% of patients with one dacrocystocele, bilateral dacrocystoceles are reported. In a small percentage of patients with nasolacrimal duct cysts, the cyst can become enlarged where it causes symptomatic nasal obstruction.

Above is a picture of a nasolacrimal duct cyst. Neonates with enlarged cysts present with epiphora, facial swelling, and may have a blue to red discoloration inferior to the medial canthus. The cyst is readily noted on anterior rhinoscopy but best evaluated with nasal endoscopy where the cyst can be seen along the inferior aspect of the inferior turbinate. A scope can typically be passed distal to the cyst which confirms the diagnosis. Also, the inferior turbinate is usually displaced superomedially. Imaging can be performed; however, physical exam with endoscopy is all that is necessary to confirm the diagnosis. Management can be conservative with warm compress and facial massage. If further intervention is needed, then endoscopic marsupialization of the cyst with probing of the duct should be performed.
THORN WALDT CYST

Thorwaldt cysts are named after Gustav Ludwig Thorwaldt who published a case series of 26 patients with such cysts in 1885. He described these cysts and midline nasal masses that resulted from an embryological anomaly. The cyst is congenital and lies in the nasopharynx. It results from a communication between the notochord and the nasopharyngeal endoderm. The incidence of Thorwaldt cysts is around 3-4% with a slight male predilection. It is suggested that the frequency may in fact be even higher; however, most cysts are asymptomatic and therefore patients are likely not diagnosed. Symptomatic patients typically present in the 2nd and 3rd decade of life with recent history including an infection, adenoidectomy, or inflammatory process.

The typical presentation of a Thorwaldt cyst is occipital headaches, persistent nasal discharge (more often post-nasal drainage), and halitosis. Less common symptoms at initial presentation include sore or stiff cervical muscles and ear fullness or Eustachian tube dysfunction. On physical exam, nasal endoscopy is required to diagnose this cyst. It appears as a smooth submucosal mass in the nasopharynx. It is superior to the adenoid pad and may have a central dimpling and/or yellow hue if infected or filled with fluid from recent irritation. While history and endoscopy can lead to the diagnosis, radiology is recommended to rule out other possibilities in the differential diagnosis. Below are listed several images on CT and MRI of a Thorwaldt cyst from Skinner LJ et al.

CT:
Thus, the work-up for a patient with a Thornwaldt cyst includes the history with the above symptoms, a physical exam including nasal endoscopy, and finally radiological evaluation. The differential diagnosis for a Thornwaldt cyst includes a Rathke pouch cyst, adenoid retention cyst, Sphenoid sinus mucocele, and nasopharyngeal carcinoma. Treatment of a Thornwaldt cyst includes observation if asymptomatic – many are identified incidentally on radiological work-up for other pathology. If symptomatic, marsupialization or resection is advocated.

CONCLUSION

A few other topics bear mentioning at this time as well. While the above diseases are the most frequent causes of congenital/neonatal nasal obstruction, neonatal nasal tumors should not be forgotten. They occur at a rate of 2-14 cases per 100000 births. The differential diagnosis in this grouping includes teratomas, hamartomas, rhabdomyosarcomas, hemangiomas, neurofibromas, and
lymphatic malformations. In similar fashion to the above pathologies, work-up includes nasal endoscopy and radiological evaluation in addition to a standard history and physical exam. Finally, it is worth repeating that neonates are obligate nasal breathers and therefore will be symptomatic from these pathologies. Radiological findings are what typically clarifies each pathology and often greatly helps in both diagnosis as well as determining the role of surgery and surgical planning. Finally, MRI is typically preferred over CT as it is more beneficial for intracranial assessment.

BIBLIOGRAPHY