9. The above two junior residents need your chiefly expertise. The newborn has bilateral vocal cord palsy. Can you tell them something about congenital vocal cord palsies and its management?

Vocal Fold Paralysis

Epidemiology

Vocal fold paralysis is the second most common congenital anomaly of the larynx, accounting for 15-20% of all cases. No gender difference exists in the prevalence of this anomaly.

Etiology and pathogenesis

Bilateral vocal fold paralysis is usually idiopathic. In certain cases, paralysis may occur secondary to central neuromuscular immaturity. Paralysis may also occur because of lesions in the central nervous system, including Arnold-Chiari malformation, cerebral palsy, hydrocephalus, myelomeningocele, spina bifida, hypoxia, or hemorrhage. Birth trauma that causes excessive strain to the cervical spine may cause transient bilateral vocal fold paralysis lasting 6-9 months. Unilateral paralysis is usually idiopathic but may be secondary to peripheral nerve pathology. Birth trauma causing traction injuries to the recurrent laryngeal nerve may be responsible for a number of cases. Lesions in the mediastinum, such as tumors or vascular malformations, may cause unilateral vocal fold paralysis. Iatrogenic injury to the left recurrent laryngeal nerve can occur during surgery for cardiovascular anomalies or tracheoesophageal fistulas or during neck surgery.

Clinical presentation

Bilateral vocal fold paralysis manifests as an inspiratory stridor at rest that worsens upon agitation in children with near-normal phonation and progressive airway obstruction. Obstruction can progress to a state of respiratory distress that requires airway intervention. Aspiration is common with bilateral vocal fold paralysis, often resulting in recurrent chest infections. On examination, the child with bilateral vocal fold paralysis may or may not be in significant respiratory distress (nasal flaring, supraclavicular or intercostal indrawing, cyanosis). Head and neck examination may reveal other cranial nerve deficits. Flexible endoscopy usually elucidates the diagnosis by demonstrating vocal fold paralysis and no other abnormality. Unilateral vocal fold paralysis may manifest during the first few weeks of life, or it may go unnoticed. The most common symptoms are a hoarse, breathy cry that is aggravated by agitation. Feeding difficulties and aspiration may also occur.

Diagnosis

A history of early inspiratory stridor associated with signs of respiratory distress suggests bilateral vocal fold paralysis. If the child is stable, flexible endoscopy may be performed to demonstrate the paralysis. Perform rigid bronchoscopy to confirm the diagnosis and to assess the airway for other anomalies. If the diagnosis is uncertain, repeat the procedure a week later to confirm the diagnosis. In patients with bilateral vocal fold paralysis, conduct radiographic and sonographic studies to assess for central nervous system abnormalities. Flexible endoscopic examination usually suffices to establish a diagnosis of unilateral vocal cord paralysis. In the presence of respiratory distress, perform rigid bronchoscopy to exclude the possibility of concurrent airway anomalies. Conduct radiographic studies (CT scanning of the mediastinum and neck) to determine if lesions are compromising the function of the recurrent laryngeal nerve. Laryngeal electromyography (EMG) is now used in the evaluation and management of vocal fold mobility disorders.
and to differentiate vocal fold fixation from paralysis. It is also valuable in determining prognosis after the onset of VCP.

Management

Patients with bilateral vocal fold paralysis may need urgent airway intervention, which can usually be achieved by endotracheal intubation. Tracheotomy is necessary to relieve the obstruction and should remain in place for 2 years to allow for spontaneous recovery, which occurs completely in more than half of patients. If recovery does not occur, consider vocal cord lateralization procedures in an effort to decannulate the patient. Arytenoidectomy produces reliable results in maintaining a patent airway that supports decannulation. Transverse laser cordotomy has had early success in allowing decannulation in older children and adults. Supportive measures are also necessary for the patient with bilateral vocal fold paralysis to ensure that adequate nutrition is received while preventing aspiration. Most cases of unilateral vocal fold paralysis can be managed by observation, ensuring that respiratory and feeding difficulties do not develop. Upright positioning is usually sufficient to alleviate aspiration difficulties. Rarely, intubation may be necessary to acquire a patent airway in distressed patients.

VOCAL CORD PARALYSIS IN CHILDREN:

Vocal cord paralysis is the second most common congenital laryngeal abnormality. This must be differentiated from the commonest congenital laryngeal abnormality, laryngomalacia. Stridor is the predominant presenting symptom in both of these conditions. Other symptoms of vocal cord paralysis in children include obstruction, weak cry, dysphagia, and aspiration. The diagnosis can usually be made by flexible endoscopy at the bedside. Once the diagnosis is made, the etiology of vocal cord paralysis must be determined.

The major causes of vocal cord paralysis in children include neurological conditions, birth trauma, and idiopathic causes. The neurological conditions include CNS disease (e.g. cerebral dysgenesis, hypotonia, cerebral palsy, Charcot-Marie-Tooth disease, etc.) and Arnold-Chiari malformation with meningomyelocele.

The connection between Arnold-Chiari malformation and vocal cord paralysis is unclear. The prevailing theories include compression of the brain stem secondary to hydrocephalus or traction on the vagus nerve as the brain stem herniates down into the foramen magnum. Regardless, it has been shown that correcting the increased intracranial pressure early can prevent or relieve vocal cord paralysis. Tracheotomy should be deferred until a shunting procedure is performed.

Management of vocal cord paralysis in children should consist of maintaining an adequate airway. A tracheotomy, at least temporarily, is usually necessary in children with bilateral vocal cord paralysis. Children with unilateral vocal cord paralysis rarely require treatment. Because idiopathic sources constitute a large percentage of the causes of vocal cord paralysis in children, and recovery rates have been reported to be as high as 62.5%, irreversible treatments should be avoided.

10. Your favorite pediatrician consults you for a congenital nasal mass.

Nasal Dermoids

Etiology and embryology

Nasal dermoids are epithelial-lined cavities or sinus tracts with variable numbers of skin appendages, including hair follicles, sebaceous glands, and eccrine glands. They constitute the most common congenital nasal anomaly. Nasal dermoids may arise from clusters of epithelium trapped during
ectodermal processes, or they may signify failure of ectodermal extensions into the fetal nasal septum to disappear as the septum fuses and ossifies.

The most widely accepted etiologic theory centers on the prenasal space and fonticulus, which forms between the anterior wall of the nasal skeleton and the frontal and nasal bones. Should skin remain attached to fibrous tissues of the nasal capsule in the prenasal space or the fonticulus, epidermal appendages (or other components) may be encroached upon and surrounded by the developing bones, thereby forming a tract. Dural attachments also may exist, creating a tract between the nasal skin and the dura that passes through the prenasal space (toward the foramen cecum) or fonticulus.

Clinical presentation and management

These nasal lesions account for 3.7-12% of dermoids in the head and neck and 1.1% of all body dermoids. Nasal dermoids generally occur in the midline, most commonly on the nasal dorsum, in the form of a nasal pit or nasal mass. They may manifest anywhere from the columella base to the glabella. Dermoids may be single or multiple and can manifest as a nasal mass or a fistulous tract. They often manifest with hair and sebaceous material (with or without drainage). They typically manifest within the first month of life, and 73% are diagnosed in the first year of life (see the images below).

A patient with a congenital nasal dermoid. A patient with encephalocele. This MRI depicts a dermoid. The dermoid is intracranial but separate from the brain. (From TL Tewfik, VM Der Kaloustian, eds. Congenital Anomalies of the Ear, Nose and Throat. New York: Oxford UP; 1997, with permission).

Dermoids may extend intracranially and should be differentiated from encephaloceles based on their lack of transillumination and inability to enlarge with crying. Diagnose with CT scan or MR imaging. Biopsy is contraindicated. If infection ensues, it usually is limited to the sinus tract or cyst. Orbital/periorbital cellulitis, osteomyelitis, meningitis, or brain abscess may occur, however.

Several radiologic findings may be noted (eg, fusiform swelling within the nasal septum, widening of the nasal vault, bifid septum, glabellar destruction, bony proliferation above cyst level, large ethmoidal cystic spaces). Enlargement of the foramen cecum with crista galli involvement or deformity may allude to intracranial extension. CT scan reveals the bony defect, and MRI differentiates the dermoid element from brain tissue. Treatment involves complete cyst and sinus tract excision. If an intracranial cyst exists, a combined craniofacial approach with neurosurgical consultation is required. Recurrence is attributed to incomplete excision.

A unique and unusual extension of the dermoid tract into the frontal sinus has been reported. It required an osteoplastic flap to access the frontal sinus floor, combined with a local midline nasal incision at the sinus tract origin.

Previous

Next Section: Embryology of the Nose

Gliomas

Etiology and embryogenesis

Gliomas are unencapsulated collections of glial cells situated outside the CNS. Possible theories of development include the following: (1) sequestration of glial tissue of the olfactory bulb entrapped during cribriform plate fusion; (2) ectopic neural tissue cells; (3) pinched encephalocele; and (4) inappropriate closure of the anterior neuropore (fonticulus frontalis), with failure of mesoderm to enter the region, resulting in inadequate bone formation.

Clinical presentation and management

Gliomas usually present in childhood as intranasal (30%), extranasal (60%), or combined masses (10%). Unlike dermoids, they do not necessarily occur in midline or attach to sinuses or skin. Gliomas form a noncompressible mass that does not increase in size on Valsalva testing and does not transilluminate. Extranasal gliomas are usually located at glabella level, but they may present laterally. Intranasal gliomas are associated most often with middle turbinate or higher structures and may mimic nasal polyps. Combined intra/extranasal gliomas have a typical dumbbell shape with a connecting band.
Fifteen percent of gliomas connect with the dura, either through the foramen cecum or through the
fonticulus. Patients may present with unilateral nasal obstruction, unilateral nasal mass, epistaxis, or
cerebrospinal rhinorrhea. Dystopia canthorum or hypertelorism is more common with extracranial lesions.
Simultaneous bilateral digital compression on the internal jugular veins does not lead to distension of the
mass (Furstenberg sign). Diagnosis is assisted with CT scan, which shows the bony defect, and with MR
imaging, which highlights the soft tissue components. Avoid biopsy. Management consists of surgical
excision of the mass.
Extranasal gliomas can be approached through standard surgical excisions. Intranasal masses that do not
extend intracranially may be approached through lateral rhinotomy. Gliomas that extend intracranially
require neurosurgical intervention. Avoid recurrences by removal of all gliomatous tissue.

Encephaloceles

Etiology and embryogenesis

Encephaloceles signify the herniation of neural tissue through defects in the skull. They may contain
meninges (meningocele) or brain matter and meninges (encephalomeningocele), or they may
communicate with a ventricle (encephalomeningocele). Encephaloceles have an etiology similar to
that of gliomas. No familial pattern has been demonstrated with these lesions. Association with other
diseases (eg, Ehlers-Danlos syndrome, frontonasal dysplasia), however, may suggest a genetic
component.

Clinical presentation and management

Twenty percent of all encephaloceles occur in the cranium. Of those, 15% are nasal. Nasal
encephaloceles can be divided into 2 types: sincipital (60%) and basal (40%).

Further, the sincipital form is divided into subtypes as follows: (1) the nasofrontal (40%), which exits the
cranium between the nasal and frontal bones; (2) the nasoethmoidal (40%), which exits between the
nasal bones and nasal cartilages; and (3) the nasoorbital (20%), which exits through a defect in the
maxilla frontal process. Sincipital encephaloceles typically present as soft compressible masses over the
glabella.

The basal form is divided into subtypes as follows: (1) the transethmoidal, which exits through the
cribiform plate into the superior meatus, extending medial to the middle turbinate; (2) the
sphenoethmoidal, which exits through the cribiform plate, between the posterior ethmoid cells and
sphenoid, to present in the nasopharynx; (3) the sphenoorbital, which enters the orbit via the superior
orbital fissure and may produce exophthalmos; and (4) the transsphenoidal, which herniates in the
nasopharynx via defects posterior to the cribiform plate. Basal encephaloceles may remain hidden
clinically for years.

Both sincipital and basal forms expand with the Valsalva maneuver, have a positive Furstenberg sign, and
transilluminate, thereby distinguishing them from gliomas. Patients may have a history of rhinorrhea or
recurrent meningitis and may have a broad nose or hypertelorism (dystopia canthorum). Broekman et al
(2008) described a case of intranasal encephalocele associated with Beckwith-Wiedemann syndrome
(BWS). This rare congenital syndrome is characterized by gigantism, macroglossia, exophthalmos,
postpartum hypoglycemia, and multiple midline defects such as omphalocele. Nasal masses in these
patients should be carefully differentiated, as complications might be severe.

Both CT scan and MRI are useful in diagnosis, the former for degree of bony defect and the latter for
degree of soft-tissue herniation (see the image below).

CT scan demonstrating encephalocele.

Biopsy is strongly contraindicated due to risk of infection and meningitis. Treatment involves surgical
excision with repair of the bony defect. Often a craniotomy is necessary to approach the encephalocele.