Pediatric Head and Neck Lesions

13

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13.1 Embryology: Key Concepts

The structures of the face are formed from five mesodermal processes: the midline frontonasal process and the paired maxillary and mandibular processes (Figs. 13.1 and 13.2).

The nose is formed by the union of two nasal placodes, each comprised of a nasal pit surrounded by a medial and lateral nasal eminence. The medial nasal eminences unite to form the septum, primary palate, and philtrum. The lateral nasal eminences form the lateral nasal walls and nasal ala. These are separated from the maxillary processes by the nasolacrimal groove, within which the nasolacrimal duct develops as a solid cord of cells that later canalizes.

The nasal cavity is separated from the oral cavity by the oronasal membrane.

The maxillary processes give rise to the zygoma, maxilla, and hard palate.

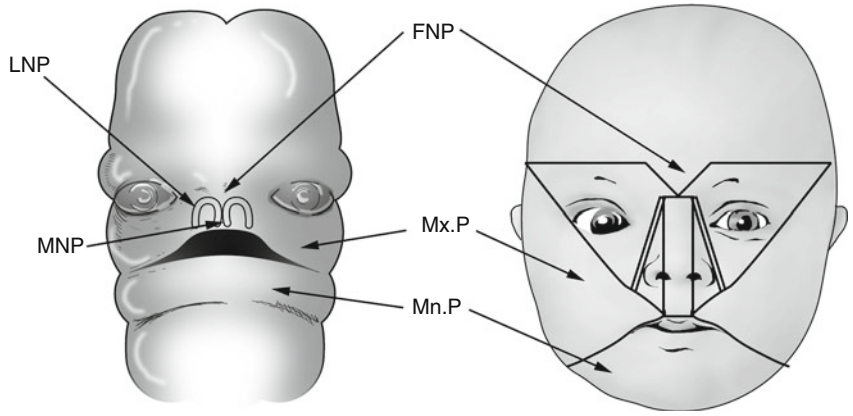

Fig. 13.1 Development of the face. *FNP* frontonasal process, *Mx.P* maxillary process, *Mn.P* mandibular process, *LNP* lateral nasal process, *MNP* medial nasal process

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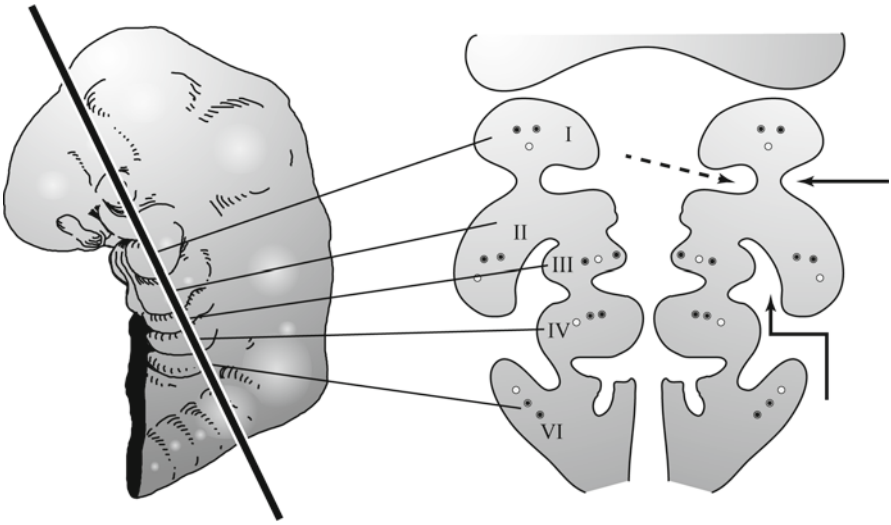

Fig. 13.2 Coronal section through the branchial arches. The branchial arches are paired mesodermal structures in the fetal neck, each with its own artery, vein, and nerve. The *straight arrow* points to a branchial cleft and the *dashed arrow* to a branchial pouch. The clefts are lined by ectoderm and the pouches by endoderm. The *bent arrow* points to the sinus of His, a cleft between the overgrown second arch and the external surface of the branchial apparatus. Second branchial anomalies are believed by some to arise from persistence of remnants of the sinus of His

The developing nasal bones are separated from the frontal bone by a transient fontanelle – the frontonasal fonticulus. The nasal bones are separated from the underlying cartilaginous nasal capsule by the prenasal space, which transmits a dural diverticulum. This diverticulum exits the cranial cavity through the foramen cecum in the midline anterior skull base and extends to the tip of the nose. Both the fontanelle and the diverticulum are obliterated by growth of the frontal and nasal bones.

The branchial arches are five bars of mesenchyme (numbered 1 to 4 and 6) in the developing neck. Each arch contains a nerve, artery, and vein. The arches are separated on their external aspect by ectoderm-lined clefts and on their internal (luminal) aspect by endoderm-lined pouches. The derivatives of the branchial arches are indicated in Box 13.1.

Box 13.1. Derivatives of the Branchial Arches

Branchial arch	Nerve	Muscles	Ligaments	Bones	Pouch derivative	Cleft derivative
I	Trigeminal	Muscles of mastication, tensor palati, anterior belly of digastric, mylohyoid	Lateral ligament of the malleus	Malleus, incus	Eustachian tube, middle ear cavity	External auditory canal

Branchial arch	Nerve	Muscles	Ligaments	Bones	Pouch derivative	Cleft derivative
II	Facial	Muscles of facial expression, stylohyoid, posterior belly of digastric, stapedius	Stylohyoid ligament	Stapes, lesser horn of the hyoid, styloid process		Faucial tonsillar crypts
III	Glossopharyngeal	Stylopharyngeus		Greater horn of the hyoid	Inferior parathyroid gland, thymus	
IV	Vagus	Laryngeal and pharyngeal musculature, except the stylopharyngeus and tensor palati			Superior parathyroid gland, C cells of the thyroid	
V	Spinal accessory	Trapezius, sternomastoid				

13.2 Congenital Anomalies of the Nose and Nasal Cavity

The three most important developmental nasal abnormalities that result in respiratory distress at birth are choanal atresia, congenital piriform aperture stenosis, and congenital nasolacrimal duct mucoceles. These all cause nasal obstruction and are symptomatic because infants are obligate nasal breathers at rest. Midline nasal anomalies include nasal dermal sinuses, dermoid and epidermoid cysts, nasal gliomas, and encephaloceles.

13.2.1 Choanal Atresia

Choanal atresia (Fig. 13.3) results from persistence of the primitive oronasal membrane that separates the nasal cavity from the oral cavity, either unilaterally or bilaterally. When choanal atresia is bilateral, it is symptomatic at birth. In the absence of symptoms, choanal atresia is suspected when the pediatrician is unable to pass a nasogastric tube. The atresia may be osseous, membranous, or osteomembranous and is best evaluated by CT. The most common finding on imaging is thickening of the vomer. It is important to obtain axial images of the nasal cavity in a plane parallel to the hard palate; if this is not done, the posterior choanae may appear artifactually narrowed. It is also important to suction secretions from the nasal cavity prior to imaging in order to be able to assess the cause of atresia

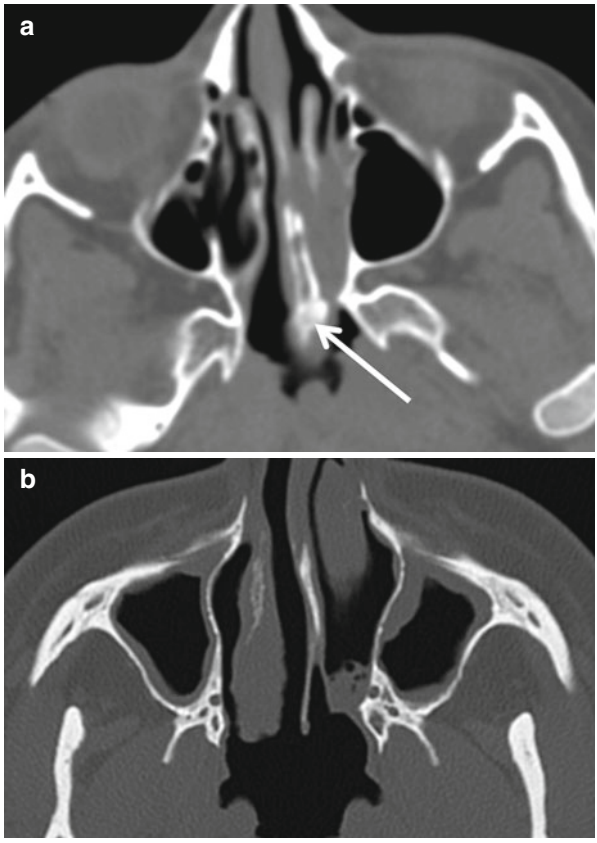

Fig. 13.3 Choanal atresia. (**a**) Osteomembranous atresia. Note the thickened vomer (*arrow*). (**b**) Membranous atresia

accurately. The orbits and temporal bones should be evaluated for coexisting anomalies that may indicate the CHARGE association (coloboma of the eye, heart defects, atresia of the choanae, retardation of growth and/or development, genital and/or urinary abnormalities, and ear abnormalities/deafness). Imaging features of the CHARGE association are discussed in the chapter on the temporal bone and skull base.

13.2.2 Congenital Piriform Aperture Stenosis

Piriform aperture stenosis (Fig. 13.4) arises from abnormal development of the medial nasal eminences which results in failure of formation of the primary palate. The piriform aperture of the nasal cavity is therefore stenotic and the palate is

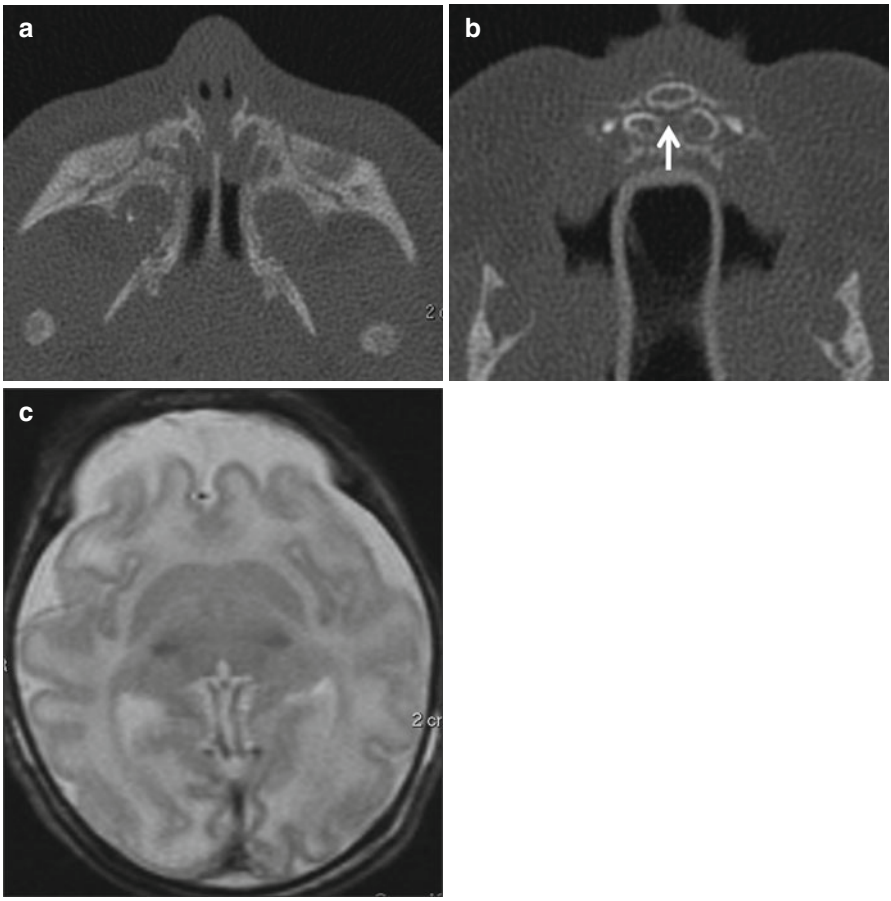

Fig. 13.4 Congenital piriform aperture stenosis. (**a**) The nasal processes of the maxillae approximate each other closely to narrow the piriform aperture. CPAS is considered to be present if the width of each PA is less than 3 mm or if the total width is less than 8 mm. The presence of a single unerupted maxillary megaincisor (**b**) should prompt evaluation for intracranial anomalies (holoprosencephaly – note the forebrain fusion in (**c**) (Images courtesy of C. Douglas Phillips, MD)

narrow. It may be recognized clinically by a complete inability to pass a nasal catheter. This abnormality may be associated with fusion of the central maxillary incisors, forming a single central or mega incisor. Midline abnormalities of the brain including corpus callosal agenesis and holoprosencephaly may coexist. The stenotic piriform aperture is best evaluated with CT; it is useful to obtain an MRI to evaluate for coexisting brain abnormalities.

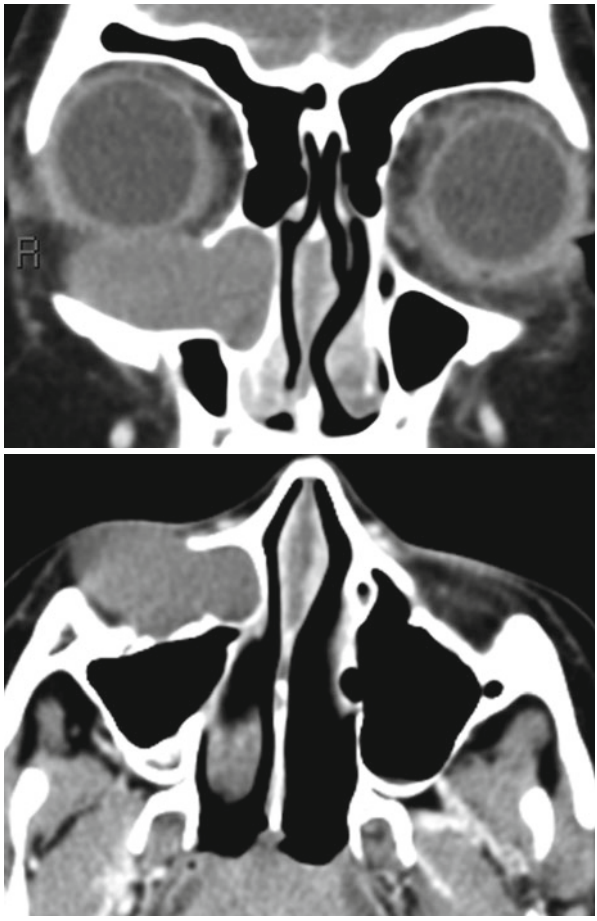

Fig. 13.5 Dacryocystocele. Dacryocystoceles may be unilateral or bilateral, with the latter tending to present at birth with respiratory distress. Unilateral lesions may go undetected until later in childhood. A smoothly expansile lesion, conforming to the course of the nasolacrimal duct is the typical appearance of a dacryocystocele (Images courtesy of C. Douglas Phillips, MD)

13.2.3 Nasolacrimal Duct Mucoceles

These mucoceles (Fig. 13.5) result from obstruction of the nasolacrimal duct by an imperforate valve of Hasner, a membrane that guards the inferior opening of the nasolacrimal duct. Normally, perforation of the valve of Hasner occurs immediately after birth due to increase in pressure within the lacrimal system when the infant cries. If perforation does not occur, a mucocele results. The process may be bilateral, and when the mucoceles are large enough, they may encroach upon the nasal cavity to produce respiratory distress. The lacrimal sac may also be dilated, especially if the valve of Rosenmuller, a membrane that is normally present at the junction of the common lacrimal canaliculus and the sac, is also imperforate. These are best evaluated by unenhanced CT. The most important differential diagnoses for an

intranasal mass in a neonate are encephaloceles, dermoids, teratomas, and hemangiomas. The characteristic low density of the mucocele and its location along the course of the nasolacrimal duct are usually diagnostic.

13.2.4 Midline Nasal Anomalies

Box 13.2 summarizes the embryological mechanisms that underlie the four types of midline nasal anomalies (Figs. 13.6, 13.7, and 13.8) and their typical imaging features. Each of these may present as a sinus or as a midline mass. An encephalocele is likely when the mass increases in size with crying. A firm mass may be a nasal heterotopia or a dermoid. Nasal glial heterotopias (also called nasal gliomas) are nonneoplastic disorganized rests of brain tissue that present as firm midline nasal masses and are best investigated with a combination of CT and MRI. The most important determination is if there is communication with the cranial cavity. With larger encephaloceles, this is usually not problematic and the herniation of meninges, CSF, and brain tissue through the midline frontonasal defect is usually obvious on MRI. Intracranial extension may not be as obvious in the case of dermal sinuses, nasal gliomas, and dermoid/epidermoid cysts. The presence of a widened foramen cecum or of a bifid crista galli is indicative of intracranial extension.

Box 13.2. Midline Nasal Anomalies

Anomaly	Embryological defect	Key imaging findings
Nasal dermal sinus	Patent dural diverticulum, failure of closure of frontonasal fonticulus	Evaluate for intracranial extension which is best indicated by widened foramen cecum or bifid crista galli
Dermoid/epidermoid	Incorporation of ectodermal elements into the diverticulum as it retracts	Well-defined mass with low density on CT and high T1 signal on MRI due to fat; epidermoid cysts are hyperintense on diffusion-weighted imaging
Nasal glial heterotopia	Sequestration of neural elements within the dural diverticulum	Well-defined mass, extranasal or intranasal, usually without direct intracranial extension; variable intensity on T1, hyperintense on T2
Encephalocele	Herniation of intracranial contents through patent dural diverticulum	May contain any combination of CSF, meninges, and brain tissue; contiguity with intracranial subarachnoid space usually apparent

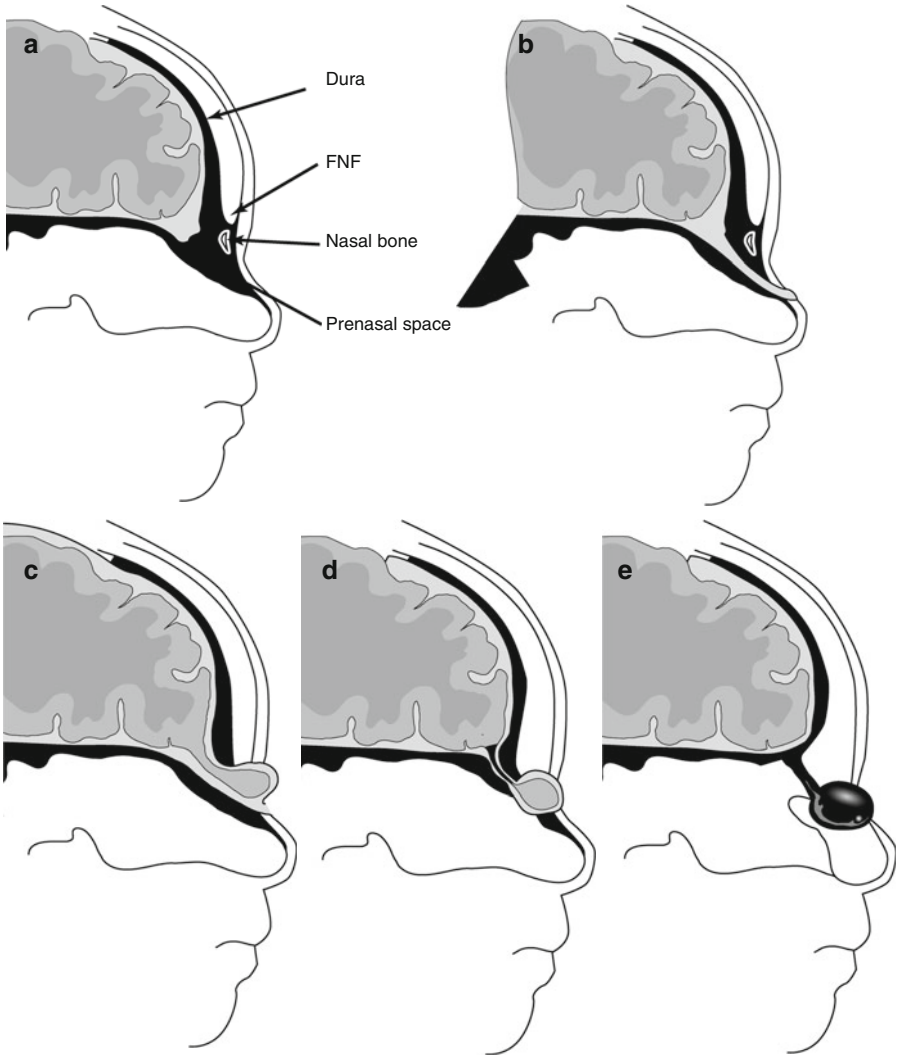

Fig. 13.6 Midline nasal anomalies. (**a**) Normal development. *FNF* fonticulus nasofrontalis – a transient opening that closes during normal development. *FC* foramen cecum. (**b**) Nasal dermal sinus. (**c**) Encephalocele. (**d**) Nasal cerebral heterotopia (nasal glioma). (**e**) Dermoid cyst. Modified from Lowe et al. (2000)

13.3 Branchial Cleft Cysts, Sinuses, and Fistulae

A branchial cleft anomaly may present as a draining sinus or fistula or as a cystic neck mass. First branchial sinuses and cysts (Fig. 13.9) are located adjacent to the angle of the mandible. Second, third, and fourth branchial sinuses and cysts are usually encountered along the anterior margin of the sternocleidomastoid (SCM) muscle in the middle third of the neck.

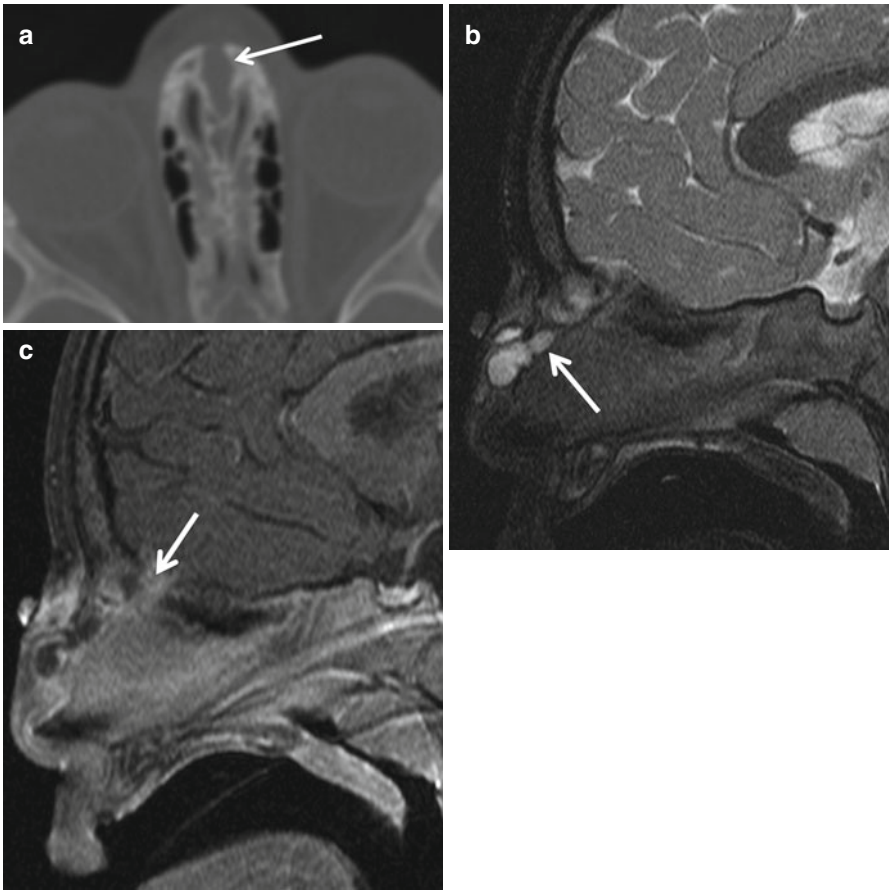

Fig. 13.7 Infected nasal dermal sinus with dermoid. The sinus tract (*arrow*) at surgery was found to extend between the nasal bones and split the crista galli (**a**). Also, a lobulated dermoid cyst, as demonstrated on the sagittal T2 (**b**) and contrast-enhanced T1 (**c**), was found. In (**c**), the intracranial extent of the sinus tract is evident. The enhancement along the sinus tract was due to infection. Dermoid and epidermoid cysts can coexist with dermal sinuses (Images courtesy of C. Douglas Phillips, MD)

The external opening of the first branchial cleft sinus/fistula is usually adjacent to the angle of the mandible although it may occur anywhere from the angle to the external auditory canal (EAC). The internal opening is typically encountered in the floor of the EAC. The sinus tract can be intimately related to the facial nerve within the parotid gland. The relationship with the facial nerve is variable. The tract may lie superficial or deep to it or be entwined with the smaller branches of the nerve. First branchial cysts may be also found in close proximity to the EAC or within the parotid gland.

A type 1 first branchial cyst/sinus lies in proximity to the pinna and may be considered to embryologically represent a duplication of the membranous EAC. Type 1 anomalies lie lateral to the facial nerve. A type 2 first branchial sinus/cyst

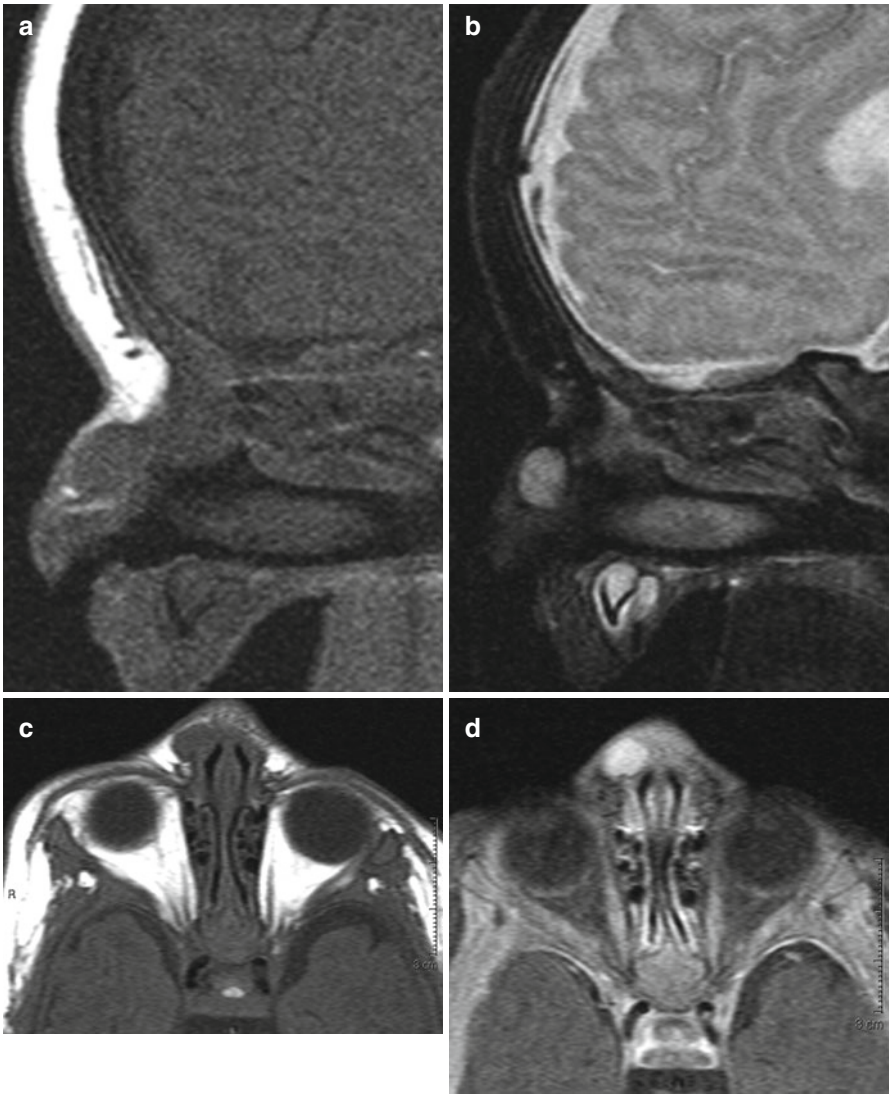

Fig. 13.8 Nasal cerebral heterotopia (nasal glioma). These are heterotopic disorganized dysplastic rests of brain tissue that may be extranasal, intranasal, or both. They present as firm sessile masses that do not change in size when the infant cries. They are isointense to gray matter on T1WI (**a**, **c**), are hyperintense on T2WI (**b**), and may sometimes enhance (**d**), as in this case (Images courtesy of C. Douglas Phillips, MD)

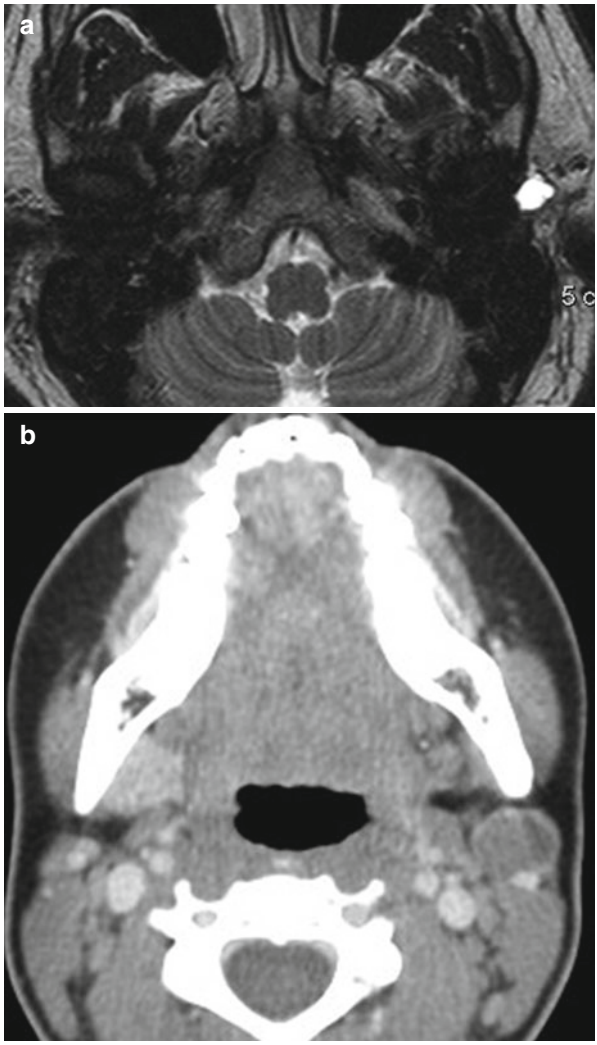

Fig. 13.9 First branchial cleft cyst. (**a**) A Work type 1 cyst in the parotid gland, (**b**) a Work type 2 cyst adjacent to the angle of the mandible

lies adjacent to the angle of the mandible and may lie external or internal to the facial nerve.

The fistulous tract of a second branchial anomaly (Fig. 13.10) passes from the external opening at the anterior margin of the SCM, between the internal and external carotid arteries, and terminates in the tonsillar fossa. Second branchial cleft cysts are usually located between the SCM and the carotid sheath, deep to the submandibular gland.

Third and fourth branchial cysts (Fig. 13.11) are also found along the anterior margin of the SCM in the middle third of the neck. Both fistulae terminate in the

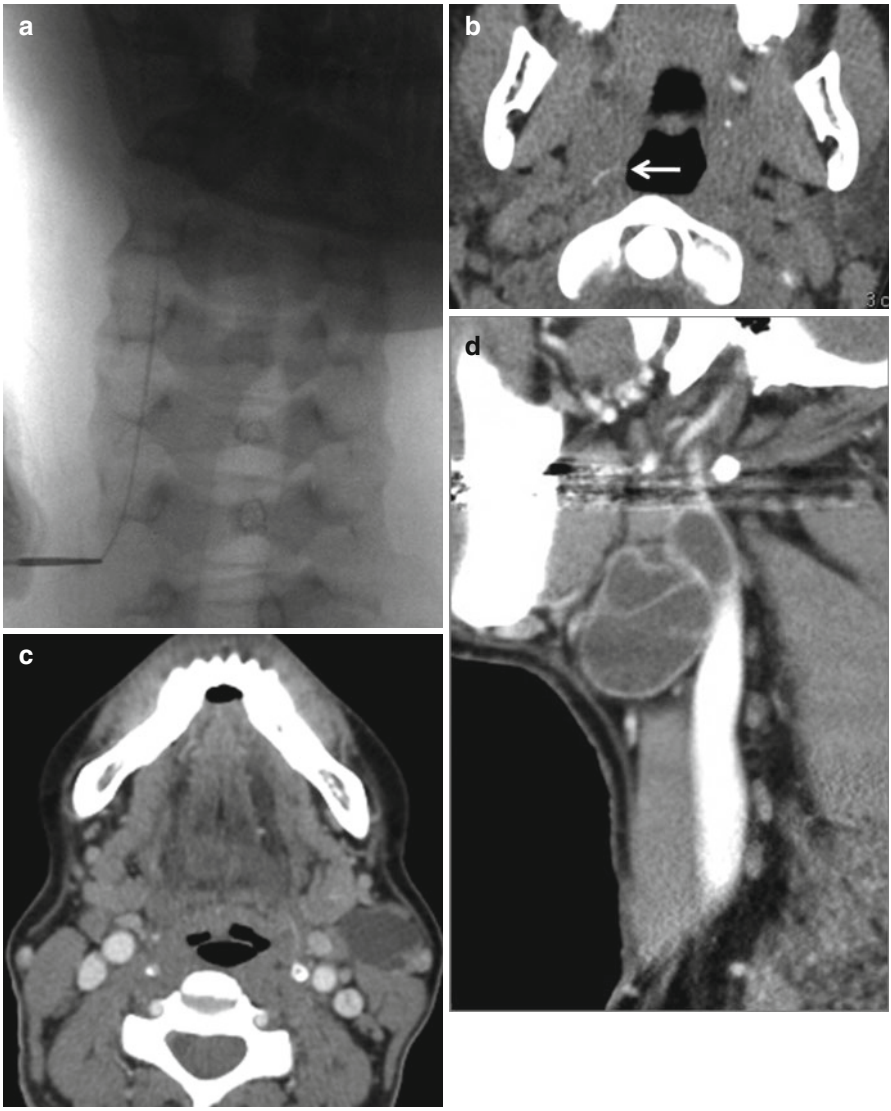

Fig. 13.10 Second branchial anomalies. The fistulogram (**a**, **b**), obtained by injecting dilute contrast medium into the external neck opening through a Rabinov catheter, shows the fistulous tact terminating in the tonsillar fossa (*arrow*, **b**). (**c**) Typical second branchial cyst located anteromedial to the sternomastoid and external to the submandibular gland. (**d**) Shows a multilocular cystic lesion adjacent to the mandibular angle. This lesion in this middle-aged patient was a metastatic lymph node and not a branchial cyst

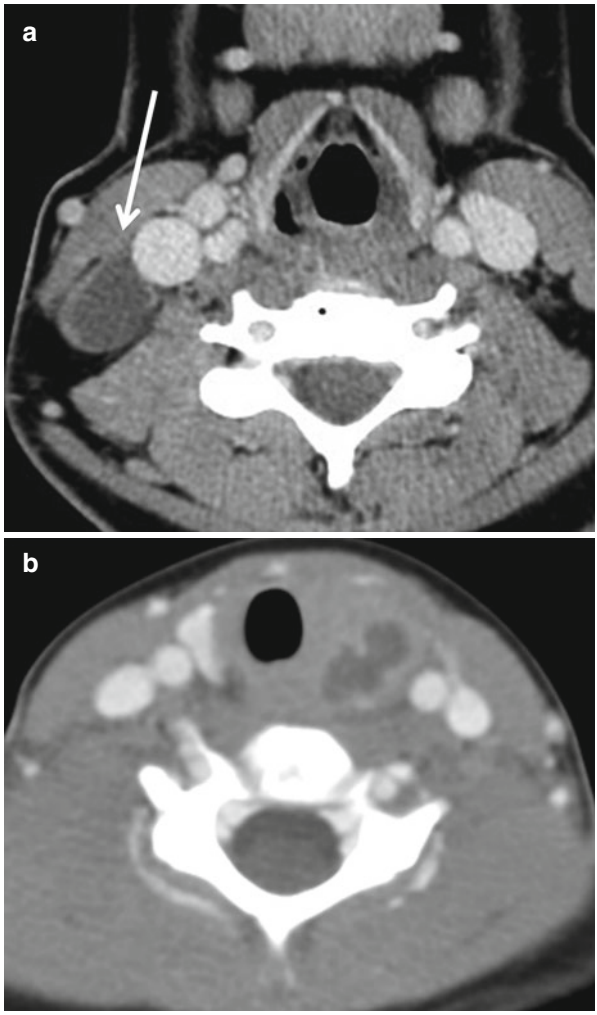

Fig. 13.11 Third (**a**) and fourth (**b**) branchial anomalies. Third branchial cysts are indistinguishable from second branchial cysts but tend to occur lower in the neck (*arrow*). In (**b**), infection along a fourth branchial sinus tract has resulted in a thyroid abscess. Sometimes edema in the ipsilateral (usually left) piriform sinus may be present and is a clue to the presence of the underlying sinus tract

piriform sinus: the third at the base and the fourth at the apex. The proximity of third and fourth branchial cleft abnormalities to the thyroid gland may result in thyroiditis when these are infected. Recurrent thyroiditis in a child must always raise suspicion for a third or fourth branchial anomaly.

The course of the four types of branchial cleft sinuses/fistulae is summarized in Box 13.3.

Box 13.3. Courses of Branchial Sinuses/Fistulae

Branchial anomaly	External opening	Sinus tract course	Internal opening
I	Angle of mandible or preauricular region	May be superficial or deep to facial nerve	Floor of the EAC
II	Anterior border of SCM	Tract passes between the carotids	Tonsillar fossa
III	Anterior border of SCM	Tract passes posterior to the carotid sheath, pierces the thyrohyoid membrane	Base of the piriform sinus
IV	Anterior border of SCM	Tract follows course of the recurrent laryngeal nerve – on the right side, it loops under the right subclavian artery; on the left, it passes under the arch of the aorta before ascending into the tracheoesophageal groove	Apex of the piriform sinus

Branchial sinuses and fistulae are best evaluated with a CT fistulogram (Fig. 13.10).

This procedure involves injection of small amount of dilute water-soluble contrast medium into the sinus orifice under fluoroscopic guidance. It is best not to perform this procedure when active infection is present. After a sufficient amount contrast is injected (usually about 1 cc), CT images are obtained. MR fistulography, performed with fat-suppressed heavily T2-weighted sequences, is an alternative. While no injection of contrast is necessary, the spatial resolution may not be sufficient to map the entirety of the tract adequately.

Branchial cysts are best evaluated with contrast-enhanced CT. Uninfected cysts are unilocular and demonstrate no mural enhancement, nodularity, or septations. When any of these features are present, especially in an adult patient, the diagnosis of branchial cyst must be seriously reconsidered. Metastatic cervical lymphadenopathy from HPV-associated oropharyngeal squamous cell carcinoma can closely mimic branchial cysts, and initial presentation of a branchial cyst in adulthood is rare. It is not infrequent for metastatic lymph nodes to be misdiagnosed as branchial cysts and for appropriate treatment to be delayed. Therefore, a cystic neck lesion with septations, nodularity, and enhancement in an adult must be assumed to represent a metastatic necrotic lymph node until proven otherwise (Fig. 13.11).

A thymic cyst is a closely related abnormality thought to arise from incomplete regression of the thymopharyngeal duct, which is a third branchial pouch derivative. The typical thymic cyst is an elongated, unilocular, low-density structure that straddles the thoracic inlet (Fig. 13.12).

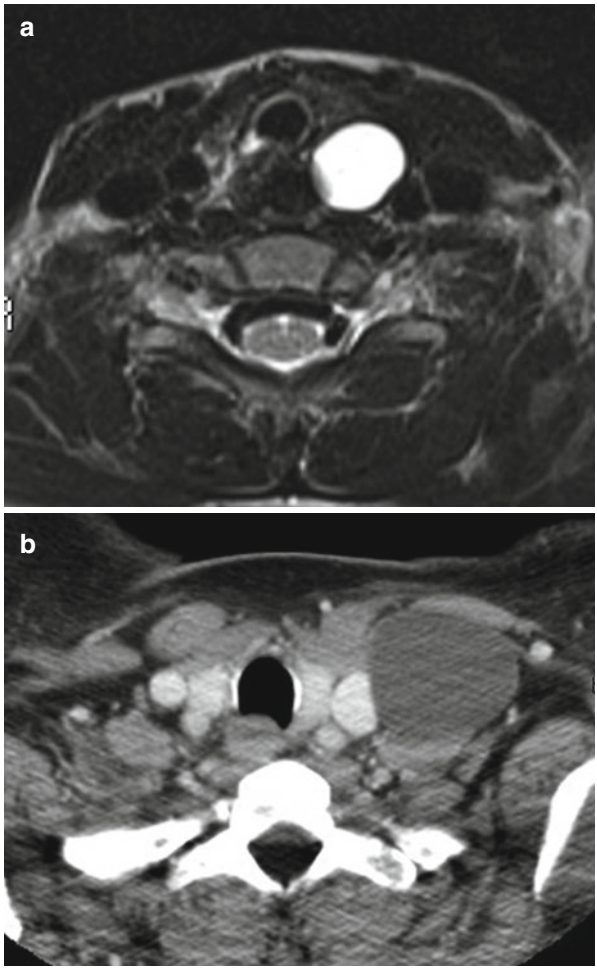

Fig. 13.12 Typical thymic cysts. Thymic cysts are usually left-sided unilocular lesions that straddle the thoracic inlet

13.4 Congenital Skull Base Anomalies

13.4.1 Arrested Pneumatization

This is an asymptomatic variant of the sphenoid sinus, occasionally mistaken for pathology. The sphenoid sinus normally undergoes fatty marrow replacement prior to pneumatization. Incomplete pneumatization results in residual foci of fatty marrow that are easily recognized by their low density on CT and high signal intensity on T1-weighted MRI (Fig. 13.13).

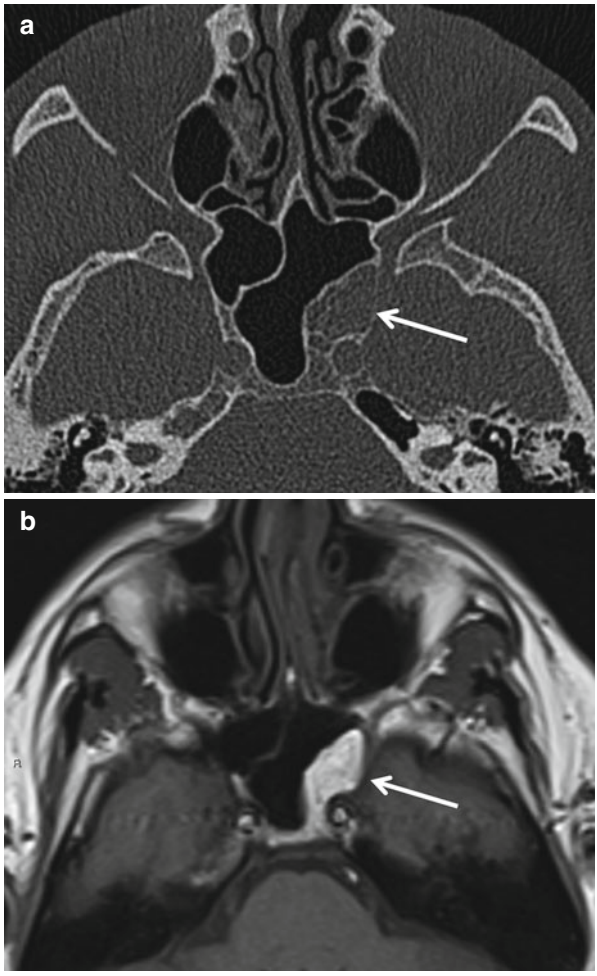

Fig. 13.13 Arrested pneumatization. This is a developmental variation usually encountered in the central skull base, often misdiagnosed as pathology. The fat contained within it (easily identified on T1W MRI) is characteristic of this process. The fat is also detectable on CT with narrow window settings

13.4.2 Encephalocele

Skull base encephaloceles are broadly classified into two types: median (transsphenoidal, sphenoethmoidal, and transethmoidal) and lateral (transalar, sphenoorbital, and sphenomaxillary). These may present with nasal obstruction, CSF rhinorrhea, or recurrent meningitis. There are two theories regarding the origin of these encephaloceles. One holds that they occur due to persistence of embryonic canals such as the pharyngohypophyseal and lateral craniopharyngeal (Sternberg) canals. Another theory, perhaps more plausible, holds that they occur due to herniation of intracranial contents between embryonic ossification centers. Acquired transalar encephaloceles may also arise from gradual rarefaction of bone by arachnoid granulations, possibly due to chronically raised intracranial pressure. Encephaloceles

are best evaluated with a combination of CT and MRI (Fig. 13.14). CT is useful in the demonstration of the osseous defect, while MRI enables evaluation of their contents. Any combination of CSF and brain tissue may be present within them. Although herniated brain tissue is usually isointense to brain parenchyma, it may also appear heterogeneous due to gliosis and encephalomalacia. Rarely, the contents of an encephalocele may enhance.

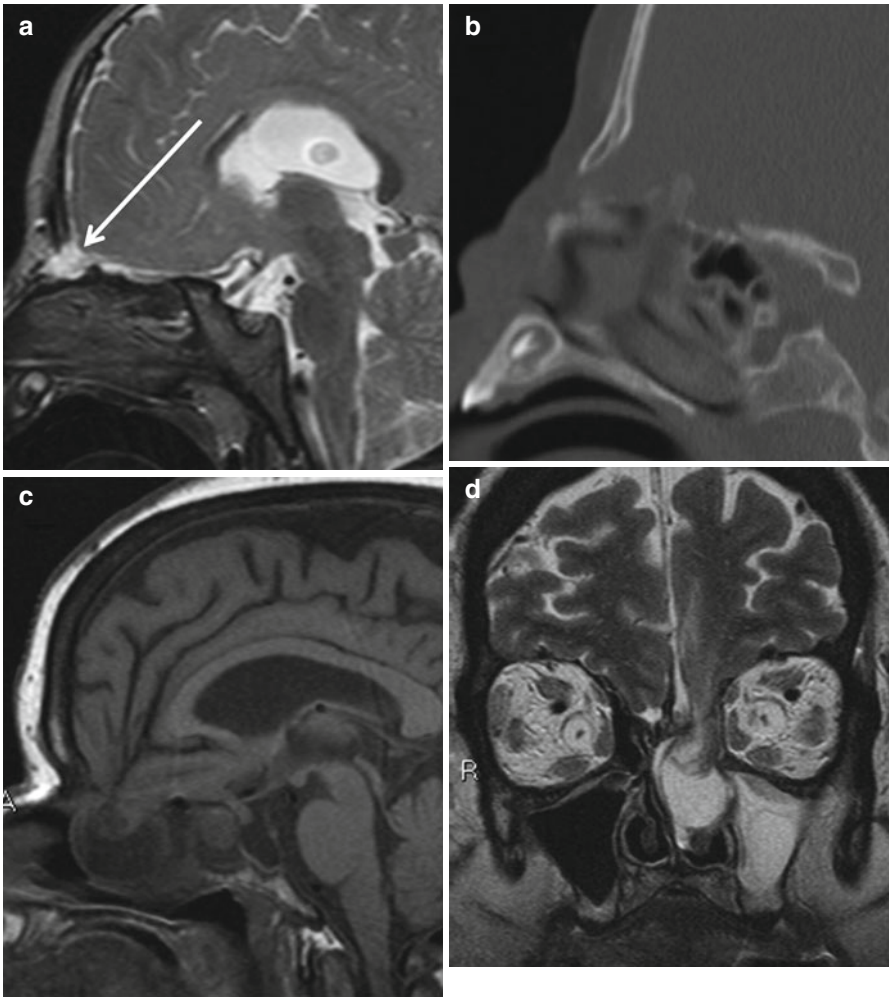

Fig. 13.14 Encephaloceles. (**a**, **b**) Small frontonasal meningocele (*arrow*) in a child presenting with a nasal root mass that increased in size with crying. (**c**, **d**) Sphenoethmoidal encephalocele containing CSF and gliotic brain tissue in an adult. (**e**, **f**) Massive sincipital encephalocele containing hemorrhagic brain tissue (dark areas on the susceptibility-weighted image, **f**). The contents of encephaloceles can be very variable and so therefore can their signal intensities on MRI. It is important to remember that they can present later in life. Indeed, the first question that comes to the surgeon's mind when determining if a nasal mass needs a biopsy is "Am I dealing with an encephalocele?"

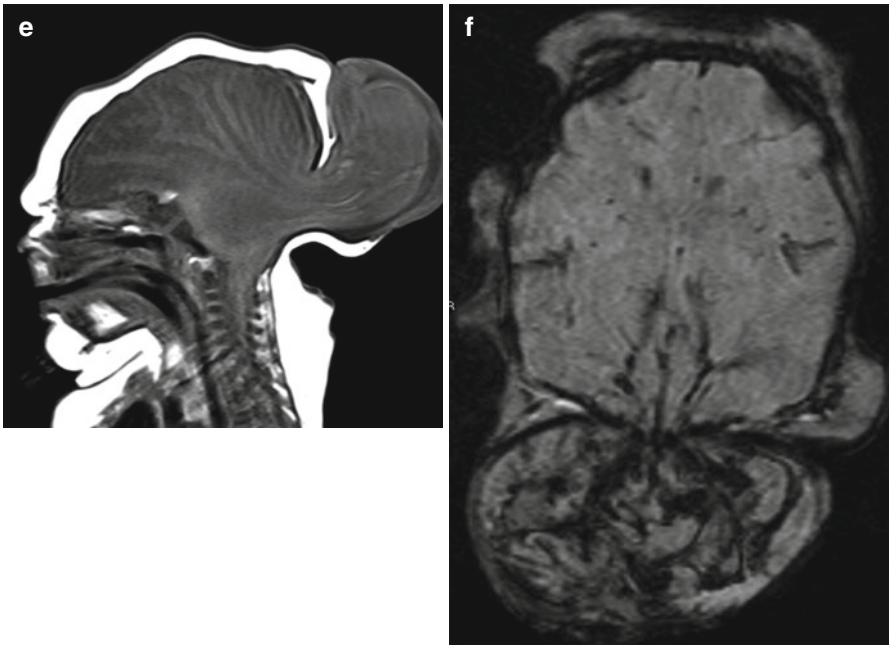

Fig. 13.14 (continued)

13.5 Hemangiomas and Vascular Malformations

The terminology used to describe pediatric head and neck vascular lesions can be confusing. It is important to realize that hemangiomas are true neoplasms, whereas vascular malformations (VMs) are hamartomas. Hemangiomas are present at birth, grow rapidly in the first few months of life, and then tend to involute spontaneously. VMs, on the other hand, grow proportionately with the patient and do not involute. VMs are subclassified into venous, lymphatic, capillary, arteriovenous, and mixed malformations.

13.5.1 Hemangiomas

Hemangiomas (Fig. 13.15) can be broadly divided into two types: infantile and congenital. Infantile hemangiomas are the most common head and neck tumor of childhood, can occur anywhere in the head and neck, and can involve both superficial and deep soft tissues. They can be solitary or multiple. Most hemangiomas of infancy are not obviously visible at birth, but present within the first several weeks of life. These often demonstrate a period of rapid growth followed by quiescence and involution.

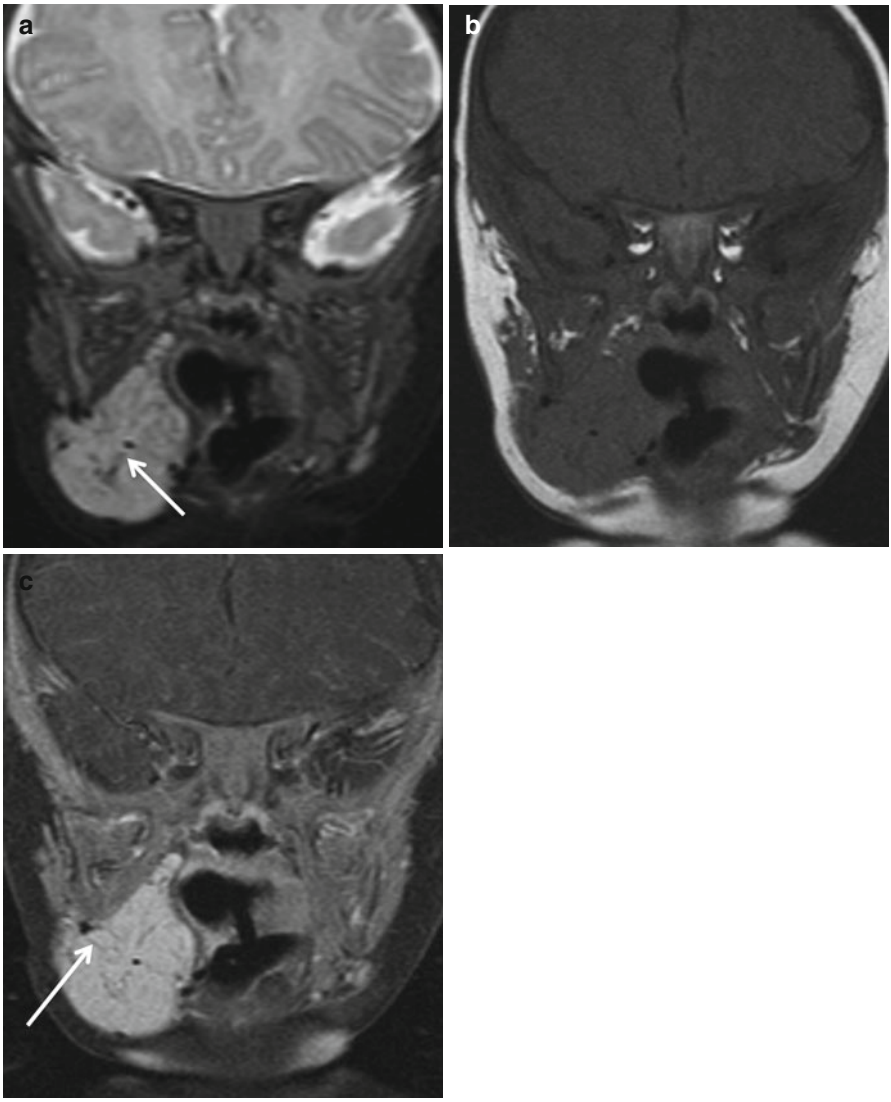

Fig. 13.15 Well defined mass centered in the right parapharyngeal space. The high T2 signal (**a**), flow voids (*arrows*), and intense enhancement (**c**) are typical of a hemangioma

Unlike infantile hemangiomas, congenital hemangiomas complete their proliferative phase before birth. Two distinct forms have been described with the acronyms, the rapidly involuting congenital hemangioma (RICH) and the noninvoluting congenital hemangioma (NICH), based on their clinical behavior.

A combination of ultrasound and MRI is typically used to confirm the clinical diagnosis and assess tumor extent. Ultrasound alone may suffice in cases where the lesion is superficial. MRI is frequently performed to assess the extent of deeper

lesions. On MRI, hemangiomas appear as discrete, lobulated, T2-hyperintense lesions with intense enhancement. On ultrasound, they demonstrate a well-marginated, mixed echogenic appearance, with low-velocity flow on color Doppler.

13.5.2 Vascular Malformations

It is also useful from a therapeutic point of view, to classify vascular malformations based on the presence of arteriovenous shunting; they are thus classified as high flow (arteriovenous malformations) or low flow (venous, lymphatic, and capillary malformations). Treatment of high-flow lesions may require catheter angiography and embolization of arterial feeders, whereas low-flow lesions may be addressed with direct puncture embolization and sclerotherapy or surgical resection.

13.5.3 Venous Vascular Malformations

Venous vascular malformations (VVMs), also referred to as cavernous hemangiomas (Fig. 13.16), are the most common vascular malformation in the head and neck. Although they can be well defined, most are infiltrative in nature and cross fascial and spatial boundaries.

On ultrasound they appear as infiltrative, poorly defined, compressible hypoechoic lesions, with low-velocity flow on Doppler. On CT, VVMs appear as

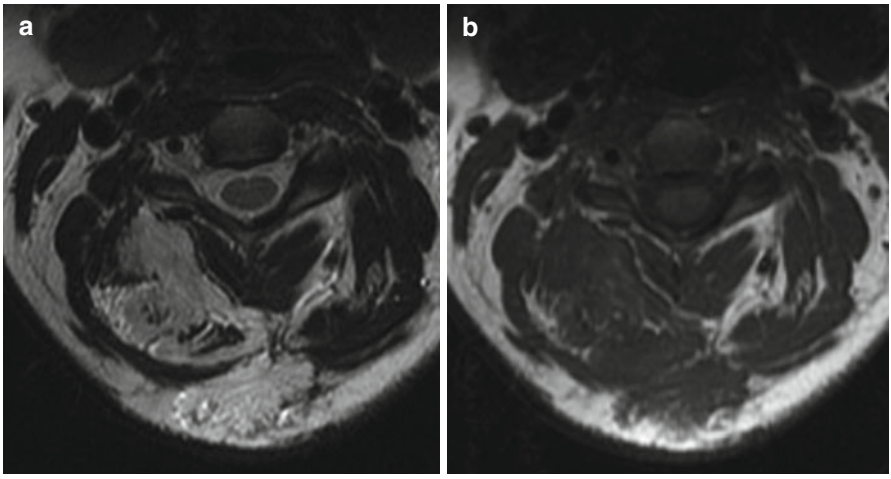

Fig. 13.16 Venous vascular malformation. These are poorly defined trans-spatial lesions (**a**, **b**) that enhance variably with contrast. The presence of phleboliths (**d**) is a typical finding. (**e, f**) are fluoroscopic images obtained during and after percutaneous injection of the sclerosant 3 % sodium tetradecyl sulfate. Note that the lesion has been nearly obliterated in (**f**)

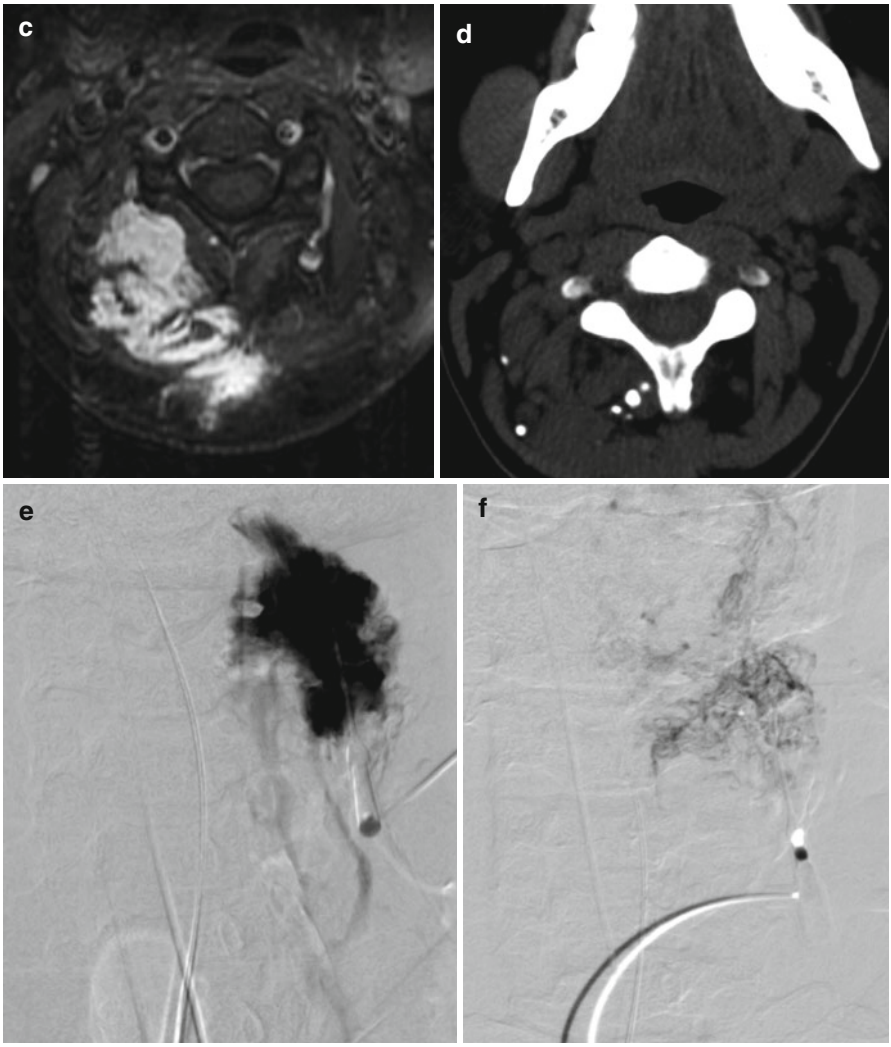

Fig. 13.16 (continued)

lobulated enhancing lesions containing phleboliths that may remodel adjacent bone. Prominent arterial feeders and draining veins are not present. On MRI, an infiltrative contour, transgression of spatial boundaries, and variable heterogeneous enhancement are typical features. When a significant lymphatic component is present, T2-hyperintense cystic foci may be evident, sometimes containing fluid levels from past hemorrhage. Flow voids are typically absent. Phleboliths, which may be evident as discrete areas of signal void, should not be confused for true flow voids.

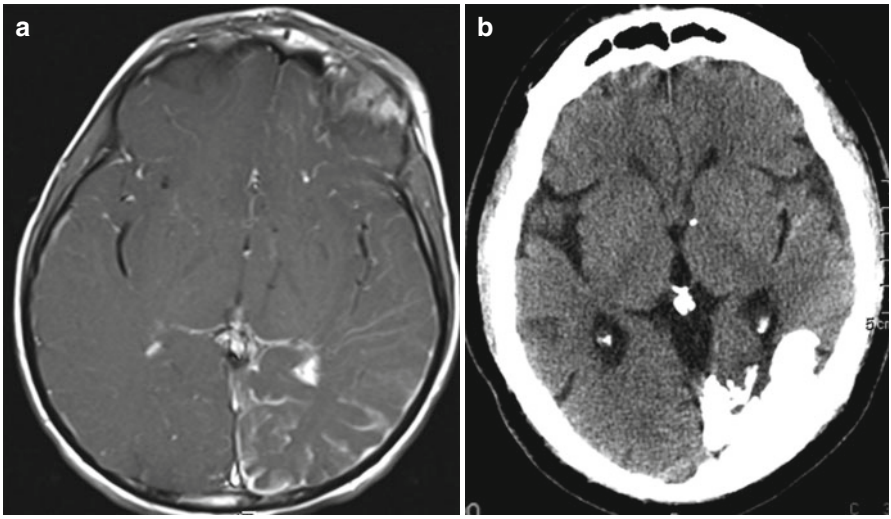

Fig. 13.17 Sturge-Weber syndrome. Capillary hemangiomas manifest as areas of skin discoloration and are usually not imaged. However, when they occur in the distribution of the trigeminal nerve, they may be associated with epilepsy caused by an intracranial pial venous angioma (**a**) and focal parenchymal atrophy and calcification (**b**)

13.5.4 Capillary Malformations

Capillary malformations (nevus flammeus, port-wine stains) are superficial lesions that do not require imaging. However, a patient with such a lesion in the distribution of the trigeminal nerve must be evaluated with MRI for Sturge-Weber syndrome, characterized by epilepsy, ipsilateral cerebral leptomeningeal angiomatosis, "tram track" gyral calcifications on CT, and cerebral hemiatrophy (Fig. 13.17).

13.5.5 Arterial Malformations

These high-flow lesions include arteriovenous malformations (AVMs) (Fig. 13.18) and arteriovenous fistulae (AVF). Histologically, there is a direct connection between feeding arteries and draining veins without a normal intervening capillary network. In fistulae, no nidus of abnormal vasculature is present and the arterial feeders communicate directly with the venous circulation. On ultrasound and Doppler imaging, they present as a loosely defined tangle of blood vessels with rapid flow. MR and CT may be used to assess extent. These malformations often require evaluation with catheter angiography.

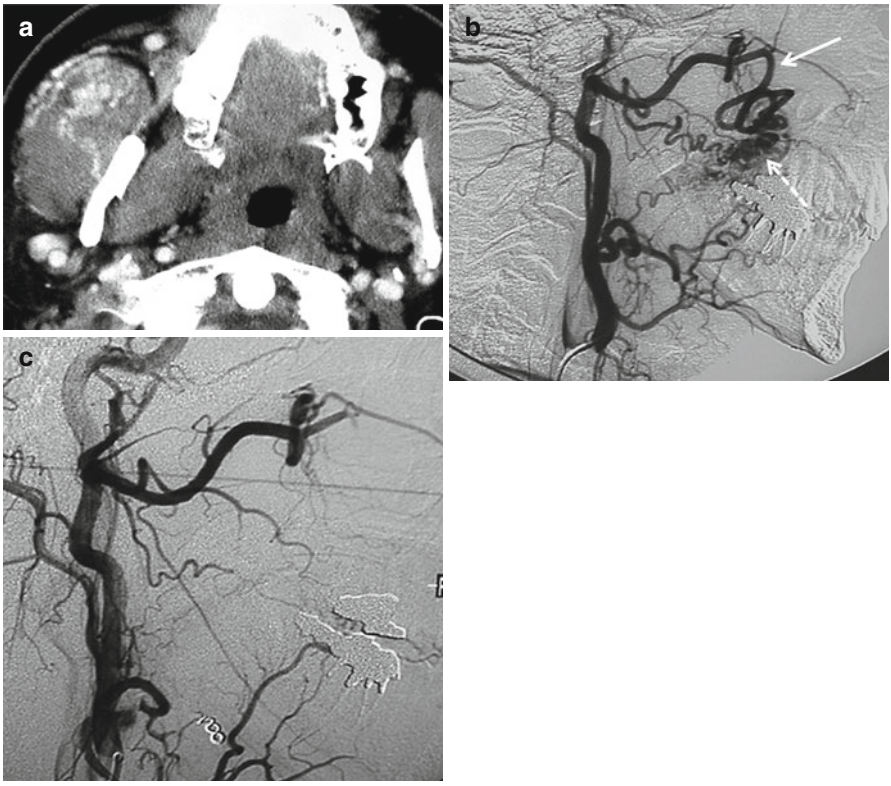

Fig. 13.18 Arteriovenous malformation of the masticator space. (**a**) Depicts tortuous vascular structures in the enlarged right masseter. This is a high-flow lesion as evidenced by the presence of arteriovenous shunting (note the enlarged internal maxillary artery (*arrow*) and the early draining veins (*dashed arrow* in **b**). The post embolization angiogram (**c**) shows that the lesion has effectively been obliterated

13.5.6 Lymphatic Malformations

Lymphatic malformations (LMs) (Fig. 13.19), also referred to as cystic hygromas or lymphangiomas, are most commonly seen in the posterior cervical triangle. Their appearance on imaging depends upon whether they are microcystic or macrocystic in nature and also upon the proportion of a coexisting venous component. On ultrasound, macrocystic lesions are comprised of one or more discrete hypoechoic foci, whereas the microcystic lymphangiomas appear hyperechoic because of the close apposition of small cysts. Uncomplicated LMs demonstrate low density on CT and high T2 signal on MRI and do not enhance significantly. Fluid levels due

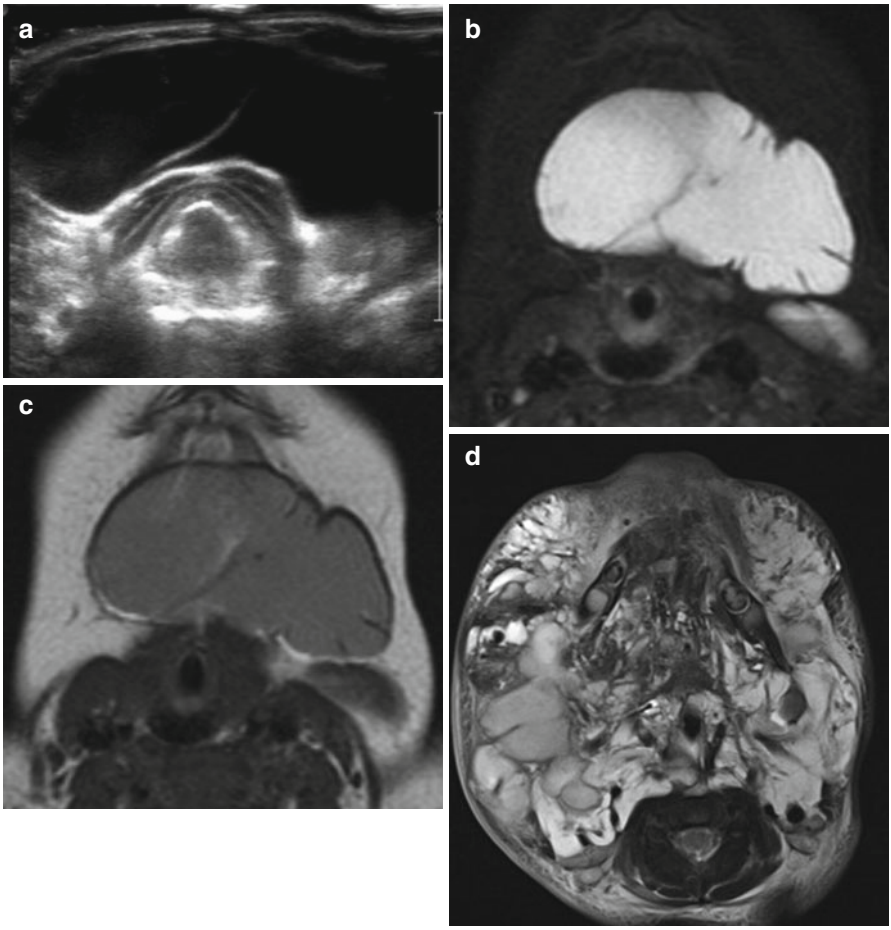

Fig. 13.19 Lymphatic malformations. (**a**–**c**) Depict a large macrocystic submandibular space lymphatic malformation. The large cystic spaces are evident on the ultrasound image (**a**). In (**b**), a T2W MR image, a fluid level, indicated past hemorrhage is present. In (**c**), a T1W image, note that the cyst fluid is of high signal intensity, also likely due to past hemorrhage. The cyst in lymphatic malformations can vary greatly in size. These lesions can also be very infiltrative and transgress spatial boundaries to the extent that complete surgical extirpation can be impossible, as in the example in (**d**)

to hemorrhage and areas of solid enhancement may indicate an associated venous component. Enhancement of the walls and septa may be seen when they are infected. Like VVMs, LMs can be extremely infiltrative in nature. Unilocular lymphangiomas can be mistaken for branchial cysts, thymic cysts, and ranulas.

The imaging features and treatment options for hemangiomas and vascular malformations are summarized in Box 13.4.

Box 13.4. Vascular Anomalies: Imaging Findings and Treatment

Vascular anomaly	Imaging findings	Management
Hemangioma	MR/CT: lobulated, well demarcated, T2 hyperintense, intense enhancement US: mixed echogenicity with low flow on Doppler DSA: intense, persistent stain, enlarged feeding vessels. A-V shunting may be present	Systemic ± intralesional steroids, particle embolization alone or embolization with surgery
Venous malformation	MR/CT: multiloculated, infiltrative, phleboliths (pathognomic), T2 hyperintense with delayed enhancement, no flow voids US: ill-defined variably echogenic lesions with phleboliths; low-flow monophasic pattern on Doppler without an arterial waveform DSA: no A-V shunting or stain; multiloculated pouches that fill with contrast in direct puncture	Percutaneous sclerotherapy with sodium tetradecyl sulfate, ethanol, with surgical excision in selected cases
Lymphatic malformation	CT/MR: uni- or multilocular cystic lesions, may be well defined or very infiltrative; enhancement and hemorrhage with fluid levels may indicate associated venous component; enhancement and thickening of septa when infected US: multiple hypoechoic cysts; may be echogenic when predominantly microcystic; insignificant flow on color Doppler DSA: avascular on angiography. Cysts fill with contrast on direct puncture	Usually surgical excision or direct puncture sclerotherapy
Arteriovenous malformations and fistulae	MR/CT: enhancing lesion with flow voids. (MR) Prominent feeding arteries and draining veins US: biphasic Doppler waveforms suggestive of shunting DSA: high-flow lesions with rapid A-V shunting; AVF shows no nidus, while AVMs have a nidus of multiple small fistulae	Fistulae: embolization at point of fistulization with balloons and coils for larger lesions and liquid adhesive (n-butyl cyanoacrylate) for smaller lesions AVM: rarely cured by embolization alone. Role of endovascular treatment usually to aid surgery

13.6 The Surgeon's Perspective

13.6.1 Choanal Atresia

Because newborns are obligate nasal breathers, bilateral choanal atresia often presents with respiratory distress or cyanosis during feeding. Unilateral choanal atresia may be asymptomatic and only identified with inability to pass a suction catheter to the nasopharynx. For newborns with choanal atresia, CT is the imaging modality of choice. In order to confirm the extent of the atresia (unilateral vs. bilateral) and to determine the type and thickness of obstructive tissue (membrane vs. soft tissue vs. bone), the CT should be performed after careful suctioning of the nasal cavity to clear secretions. Some protocols use a decongestant to further improve the CT evaluation. Choanal atresia is usually repaired via an endoscopic transnasal approach, which results in low recurrence rate and few complications.

13.6.2 Congenital Nasal Malformations

Congenital nasal malformations are best evaluated by a combination of CT and MRI. Biopsy of a congenital nasal mass should be avoided until imaging has been obtained. Often the diagnosis can be made with imaging alone. The treatment of nasal dermoids and gliomas involves surgery to completely remove the tumor and any associated sinus tract. If these extend intracranially, a more extensive craniofacial approach may be required in conjunction with a neurosurgical specialist. For encephaloceles, repair of the bony defect is usually required, which may necessitate a craniotomy.

13.6.3 Branchial Cleft Anomalies

Branchial cleft anomalies are an important diagnostic consideration in the pediatric and young adult population. Branchial cleft cysts are much more common than fistulae or sinuses. Specialized imaging modalities such as fistulograms are useful for defining the tract in fistulae and sinuses to assist with complete surgical extirpation, but are of no utility for the branchial cleft cyst. Branchial cleft cysts can be diagnosed for the first time in an adult without a prior history of a neck mass. However, this is uncommon. Thus, a mass in an adult that appears consistent with a branchial cleft cyst on imaging must be carefully evaluated for malignancy. Cystic metastatic nodes are common with HPV-related oropharyngeal squamous cell carcinoma, which often presents with a subtle primary lesion in young nonsmokers. In an adult with a newly diagnosed cystic neck mass, a thorough head and neck exam including endoscopy should be performed. If no primary lesion is noted, a careful ultrasound-guided fine-needle aspirate biopsy of the rim or any solid component of the cyst should be performed. Even when a diagnosis of cancer is absent and a decision is

made to proceed with surgical removal of the cyst, adult patients should be consented for a completion neck dissection and appropriate staging exam under anesthesia if intraoperative evaluation of the cyst by frozen section demonstrates squamous cell cancer.

13.6.4 Vascular Malformations

It is extremely important to differentiate hemangiomas from other congenital vascular malformations. Hemangiomas can be treated with observation, steroids, or beta-blockers, and parents can be assured that, while these lesions may initially grow, the expectation is complete involution over time. Other congenital vascular malformations are not expected to spontaneously resolve. Management of these lesions is by observation, embolization/sclerosis, or surgical removal. These tumors are often infiltrative and difficult to completely extirpate. Thus, they frequently recur, and careful consideration should be given to nonsurgical therapy when possible. However, when expansion is destructive (e.g., erosion of the mandible), more aggressive surgical intervention may be warranted.

Further Reading

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