Introduction

This book is primarily a book on MR imaging of the pediatric central nervous system including the spine. However, going through neuroradiology cases, we stumbled on a number of unusual and interesting head and neck cases and decided to include a short chapter on MR imaging of the pediatric head and neck.

The chapter contains congenital malformation, developmental anomalies, and unusual inflammatory neoplastic disease in the pediatric head and neck area. This chapter does not have the ambition of being a complete head and neck chapter, but rather a chapter on miscellaneous conditions that can be well imaged with contemporary techniques. CT is the primary cross-sectional imaging technique for the pediatric head and neck but with newer development and rapid sequences MR imaging will be more and more useful. The lack of radiation is a significant advantage in this group of patients. Degenerative, inflammatory, neoplastic and congenital abnormalities are presented in this chapter.

Multinodular Goiter

Clinical Presentation

An 18-year-old presenting with hyperthyroidism.

Images (Fig. 9.1)

A. Coronal T1-weighted image shows marked enlargement of the left and right thyroid glands with narrowing of the trachea
B. Axial T2-weighted image with fat suppression shows a multilobulated enlargement of the entire thyroid gland with narrowing of the trachea
C. Coronal T1-weighted image with contrast shows enlargement of the thyroid gland without abnormal enhancement

Discussion

Goiter is a clinical diagnosis that implies an enlargement of the thyroid gland. Goiter develops because the thyroid gland compensates for inadequate thyroid hormone output. In goiter the thyroid tissue hypertrophies to achieve an euthyroid state. Often the regulation is not optimal and the patient may show either hypothyroidism or hyperthyroidism. Multinodular goiter can also be evaluated with nuclear scintigraphy and ultrasound. CT and MR imaging are useful in evaluating secondary manifestations of goiter such as compression and displacement of trachea, esophagus and adjacent vessels. It is often important to determine the substernal and mediastinal extension which is best seen on CT or MR images.
Figure 9.1
Multinodular goiter

Suggested Reading


### Thyroglossal Duct Cyst

#### Clinical Presentation

A 3-year-old female presents with a mass at the base of the tongue.

#### Images (Fig. 9.2)

A. Sagittal T2-weighted image shows a cystic lesion at the base of the tongue. It has high signal intensity on the T2-weighted images and is located in the region of foramen sacrum

B. Axial T2 fat-suppressed image shows a high signal in the lesion at the base of the tongue

C. Contrast-enhanced axial image demonstrates no enhancement in the lesion

D. Coronal contrast-enhanced image confirms the location of the nonenhancing lesion to the base of the tongue

#### Discussion

The developing thyroid gland is initially connected to the tongue by a narrow tube called the thyroglossal duct. This duct extends from the junction between the anterior two-thirds and the posterior one-third of the tongue down to the hyoid bone and further to the thyroid bed. In a normal situation the thyroglossal duct begins to degenerate and eventually atrophy between the fifth and sixth fetal weeks. The foramen sacrum at the tongue is the normal remnant of this duct. Thyroglossal duct cyst is the most common congenital neck mass and accounts for nearly 90% nonodontogenic congenital cysts.

It is usually located in the midline and can occur anywhere along the path of the thyroglossal duct. On MR imaging the T1-weighted signal intensity can vary from low to high while the T2-weighted images in general are high signal intensity. The variation in signal intensity reflects the variable protein content of the cyst. Treatment is surgical. It is important to remove not only the cyst but the sinus tract as well to avoid recurrence. The general operation for thyroglossal duct cyst is called the Sistrunk procedure. By this procedure the body of the hyoid bone is removed with the thyroglossal duct and its cyst. This procedure has reduced the recurrence rate from nearly 50% to less than 4%.

The symptoms for thyroglossal duct cyst are often related to infections, but others present with dysphagia or a palpable mass.

#### Suggested Reading


Figure 9.2
Thyroglossal duct cyst
**Tornwaldt Cyst**

**Clinical Presentation**

An 18-year-old presenting with the incidental finding of a Tornwaldt cyst.

**Images (Fig. 9.3)**

A. T2-weighted axial image with fat suppression demonstrates three cysts in the posterior nasopharynx (arrow). This is high signal on T2-weighted images.

B. Contrast-enhanced image demonstrates no enhancement and low signal in the center of the cyst. FLAIR image demonstrate the high signal on the cyst in the posterior nasopharynx.

**Discussion**

Tornwaldt cyst occurs when the pharyngeal bursa ectoderm retracts with the notochord into the clivus. Thus this is a developmental cyst. It is seen in approximately 3% of healthy adults and most of the time is asymptomatic. Occasionally a patient may complain of occipital pain, purulent nasal drainage, ear fullness or odynophagia. A cyst that gets infected causes symptoms that are consistent with infection. The infection can spread inferiorly towards the mediastinum. They are often bright on T2-weighted images, presumably due to high protein content. There is usually no need for treatment. The cyst is named after Gustav Ludwig Tornwaldt (1843–1910).

**Suggested Reading**


Cystic Hygroma in Neck of Newborn

Clinical Presentation

A 3-week-old male presents with apnea and cyanosis. On palpation there was a soft subtle mass in the left neck. This was vaguely visible on inspection.

Images (Fig. 9.4)

A. Sagittal T2-weighted image shows a 5cm high signal intensity multiloculated mass (arrow) in the anterior neck extending from the mandible to almost the thoracic inlet
B. Coronal contrast-enhanced T1-weighted image shows no enhancement but high proteinaceous content of the mass lesion with a fluid-fluid level (arrow)
C. Axial contrast-enhanced T1-weighted image with fat suppression demonstrates again no enhancement but high signal intensity fluid with a fluid-fluid level (arrow)
D. Axial T2-weighted image with fat suppression shows a lesion with high signal in the anterior neck displacing the normal structures. There are multiple fluid levels with low signal indicating blood products

Discussion

Cystic hygroma is the most common form of lymphangioma. It consists of dilated cystic lymphatic spaces. The most common location is in the neck. Cystic hygromas are often isolated with the remainder of the lymphatic system being normal. The differential diagnosis of a large neck mass in a newborn includes lymphangioma, teratoma and epidermoid. An isolated cystic hygroma may develop when one of several potential lymphatic venous anastomoses fail to form. An isolated cystic hygroma can also form if an aberrant bud loses its connection to the primordial lymphatic sac from which it arose.

On imaging cystic hygromas are typically multiloculated masses in the neck. The cyst wall is rarely seen unless the lesion has been infected. Typically the T1 is low and T2 signal intensity is high. If hemorrhage has occurred fluid levels are best seen on MR imaging and a fluid level is characteristic of a cystic hygroma.

Lymphangiomas can be subclassified histologically into cystic hygroma or lymphangioma, cavernous lymphangioma, capillary or simple lymphangioma and vasculolymphatic malformation or lymphangiohemangioma. It is often best to consider these four types as a spectrum of manifestations of the same pathological process. Each case often has combinations and they are often separated by the size of the lymphatic spaces. Pathologically they are very similar since all of them are composed of endothelium-lined lymphatic channels that are separated by connected tissue stroma.

Suggested Reading

Figure 9.4
Cystic hygroma in neck of newborn
Neurofibromatosis

Plexiform Neurofibromatosis of the Neck

Clinical Presentation

A 9-year-old female presents with a painless swelling of the right lower neck.

Images (Fig. 9.5)

A. Axial T2-weighted image with fat suppression shows a multilobulated complex mass in the right lower neck extending across the midline (arrow). It is separating the common carotid artery and the jugular vein and abutting the spine posteriorly.

B. Axial postcontrast T1-weighted image shows the enhancing mass separating the carotid and the jugular on the right (arrow).

C. Coronal contrast-enhanced T1 weighted image shows the multilobulated mass in the carotid sheath on the right (arrow).
Neurofibromatosis of the Orbit

Clinical Presentation

A 30-year-old presenting with a large periorbital mass on the left.

Images (Fig. 9.6)

A. Axial T2-weighted image shows a large expansible mass in the left orbit deforming the globe and extending into the retro-orbital area. There are large flow voids in this highly vascular mass.

B. Contrast-enhanced T1-weighted fat-suppressed image demonstrates marked enhancement of the periorbital mass with compression and deformation of the globe.

C. Coronal contrast-enhanced T1-weighted fat-suppressed image demonstrates the mass expanding the orbit and deforming the globe.
Discussion

Neurofibromas and schwannomas are the most common nerve sheath tumors of the peripheral nerves in the head and neck. There are three types of neurofibromas, namely: localized, diffuse and plexiform. The vast majority of these lesions are localized and may not have association with NF1. Plexiform neurofibromas are associated with NF1. These lesions usually occur in early childhood. Plexiform neurofibromas involve a long nerve segment and its branches and tortuous expansion giving the appearance of a “bag of worms”. Malignant transformation may be seen in 4% of cases.

On noncontrast-enhanced CT images, they are seen as large multilobulated masses of low attenuation related to the fat content of myelin from Schwann cells, water content of myxoid tissue, cystic areas of hemorrhage and necrosis. On T1-weighted images, plexiform neurofibromas show signal intensity similar to that of muscle. On T2-weighted images, the lesions may show the “target sign”. This sign consists of low-to-intermediate signal intensity centrally, with a ring of high signal intensity peripherally. This may be due to a high collagen content located centrally and more myxoid tissue at the periphery. Contrast enhancement is variable. Irregular nodular enhancement with central necrosis is typical of malignant peripheral nerve sheath tumors.

Suggested Reading


Kikuchi Disease

Clinical Presentation

An 18-year-old with renal failure and lupus presents with neck pain and swelling.

Images (Fig. 9.7)

A. Axial T2-weighted image shows bilateral cervical lymphadenopathy in the posterior triangles (arrows). There is also a large retropharyngeal/prevertebral fluid collection
B. Axial T1-weighted image after contrast and with fat suppression shows the bilateral cervical lymphadenopathy. The retropharyngeal/prevertebral fluid collection is not enhancing and there is no ring enhancement to suggest an abscess
C. Axial DW image shows the bilateral cervical lymphadenopathy with high signal intensity. The retropharyngeal/prevertebral fluid collection also has high signal intensity
D. Axial ADC map confirms restricted diffusion in the posterior lymphadenopathy bilaterally. The retropharyngeal/prevertebral fluid collection is not decreased and therefore does not have true restricted diffusion

Discussion

Kikuchi disease, also referred as Kikuchi-Fujimoto lymphadenopathy (KFL) or histiocytic necrotizing lymphadenitis, is a self-limiting benign lymphadenopathy with unknown etiology that predominately affects young females. The cervical lymph nodes are usually involved, more commonly unilaterally, and may be associated with tenderness, fever, skin rashes, weight loss, chills, and sometimes splenomegaly.
The CT imaging appearance of Kikuchi disease may be variable, mimicking various nodal diseases, such as lymphoma, metastasis, tuberculosis, and especially in children, Still’s disease. However, the necrotic areas of the nodes emit relatively lower signal than the non-necrotic portion on T2-weighted MR images, different from the usual bright signal intensity of nodal necrosis seen in other nodal diseases. The speculated reason for this is restricted mobile protons within high protein content in fibrinoid material of necrotic focus. The high signal on DW images and the corresponding low signal on ADC images in the present case further support this.

**Suggested Reading**


Dermoid

Clinical Presentation

An 18-year-old male presents with a doughy swelling of the floor of the mouth clinically resembling a thyroglossal duct cyst.

Images (Fig. 9.8)

A. Axial noncontrast T1-weighted image shows a well-circumscribed 5-cm oval lesion in the center of the floor of the mouth (arrow)
B. The lesion is intermediate to high signal on T2-weighted image (arrow)
C. On coronal T2-weighted image the lesion is high signal and is located in the sublingual space, depressing the mylohyoid muscles
D. Axial post-contrast T1-weighted image shows no enhancement
E. Coronal post-contrast fat-suppressed image confirms the lack of enhancement and the location to the sublingual space

Discussion

Dermoid cysts are the least common of the congenital neck lesions and account for about 7% of all cysts in the neck. The cystic lesions are classified as epidermoid, dermoid and teratoid. Dermoid cysts are commonly used in reference to all three types of lesions without regard to the differentiating histological types. Histologically it is important to distinguish between epidermoid and dermoid/teratoid cysts because the dermoid/teratoid has a malignant potential which the epidermoid does not.

It has been theorized that epithelial rests become enclaved during midline closure of the first and second branchial arteries. The dermoids are frequently located in the floor of the mouth (sublingual, submental or submandibular regions). They can also occur in other locations such as the tongue, lips and oral mucosa. The differential diagnosis is of a cystic mass in this region includes glossal duct cyst, abscess, ranula, and mucocele.

Suggested Reading

Figure 9.8
Dermoid
Schwannoma of the Tongue

Clinical Presentation

A 13-year-old presents with a slowly growing mass of the tongue.

Images (Fig. 9.9)

A. Axial T2-weighted image shows a high signal mass in the anterior tongue (arrow)
B. Axial T2-weighted image with fat suppression shows a well-defined high signal intensity lesion in the anterior aspect of the right body of the tongue. The signal is not decreased after fat suppression indicating that there is no significant fatty component in the lesion
C. Sagittal T2-weighted image with fat suppression demonstrates a high signal mass in the anterior tongue (arrow)
D. After contrast there is marked enhancement
E. Coronal fat-suppressed contrast-enhanced image shows the mass in the right anterior tongue

Discussion

Schwannomas are nervous tissue tumors that arise from Schwann cells. They are uncommon in peripheral nerves and they are rare in the tongue. They often present as a slowly growing mass producing no or few symptoms. Treatment for Schwannomas is exclusively surgical and usually enucleation of the mass is uncomplicated. Malignant transformation of Schwannomas is exceedingly rare.
Suggested Reading


Pleomorphic Adenoma of the Nose

Clinical Presentation

A 13-year-old girl with a history of nasal obstruction and epistaxis. On clinical examination she has a mass in the left nasal cavity.

Images (Fig. 9.10)

A. Axial T1-weighted fat-suppressed MR image shows an overall enhancing mass lesion in the nasal cavity on the left side (arrow). This is contiguous with the turbinates

B. Coronal T1-weighted fat-suppressed image demonstrates the mass in the left nasal cavity (arrow)

C. Axial T2-weighted fat-suppressed image demonstrates the mass in the anterior of the left nasal cavity (arrow). It is a more intense signal than the adjacent nasal turbinates
Discussion

Pleomorphic adenomas (benign mixed tumors) are the most common benign glandular tumors of the oral cavity. They are characterized by the presence of both mesodermal and glandular tissue. The majority of pleomorphic adenomas occur in the parotid gland but 8% occur in the submandibular gland, 0.5% occur in the sublingual gland, and 6.5% occur in the minor salivary glands situated throughout the upper aerodigestive tract.

On imaging, pleomorphic adenomas are well-demarcated, homogeneous and slightly hyperdense to muscle on noncontrast-enhanced images. There is often significant enhancement.

Pleomorphic adenomas in the nose are unusual and the differential diagnosis would be polyp, lymphoma or just an asymmetric turbinate. Treatment is surgical resection.

Suggested Reading


Rhabdomyosarcoma

Clinical Presentation

A 5-week-old female with stridor and a posterior pharyngeal mass.

Images (Fig. 9.11)

A. Axial T1-weighted MR image shows a large mass in the nasopharynx (arrow)
B. Axial T2-weighted image shows the well-circumscribed mass (arrow) in the nasopharynx with linear and punctate areas of low attenuation presumed to be vascular structures
C. Contrast-enhanced T1-weighted image shows enhancement and flow voids (arrow)
D. Coronal contrast-enhanced T1-weighted image with fat suppression shows the large enhancing lesion in the nasopharynx (arrow)

Discussion

Rhabdomyosarcoma is the most common malignancy in the nasopharyngeal region in children and accounts for approximately 8% of childhood cancer. Rhabdomyosarcoma is divided into four histological types: embryonal, botryoid, alveolar and pleomorphic. The embryonal and botryoid types account for about 90% of primary head and neck lesions. The three common sites of predilection in head and neck region are: the orbit, the nasopharynx and paranasal cavities, and the temporal bone. Tumors in the nasopharynx region or sinuses more commonly manifest with airway obstruction, epistaxis, dysphagia, local pain and cranial nerve palsies.
On CT, rhabdomyosarcomas are seen as ill-defined, inhomogeneous, large soft-tissue masses which may have erosion effects on the surrounding bones. Hemorrhage or calcification is uncommon. MR imaging is the technique of choice for evaluation of the tumor site because of superior ability to characterize the soft tissues. On T1-weighted images, the mass is of intermediate signal intensity, and on T2-weighted images the mass is hyperintense to adjacent muscle. The enhancement is variable. Lymphadenopathy when present, tends to be unilateral and small in comparison to nasopharyngeal carcinoma.

**Suggested Reading**

Acinic Cell Carcinoma of the Parotid Gland

Clinical Presentation

An 11-year-old female presents with an asymptomatic swelling of the right parotid gland. This has not responded to antibiotic treatment.

Images (Fig. 9.12)

Ultrasound scan showed a multilobulated noncalcified mass in the right parotid gland (not shown)
A. Axial T1-weighted image shows a well-circumscribed intermediate to low attenuation mass (arrow) in the posterior portion of the right parotid gland
B. Sagittal T1-weighted image demonstrates the mass in the tail of the right parotid gland (arrow)
C. Coronal T1-weighted image demonstrates the mass occupying a large portion of the posterior aspect of the right parotid gland (arrow)
D. Contrast-enhanced axial T1-weighted MR image with fat suppression shows slight enhancement of the mass lesion (arrow)
E. Axial DW image demonstrates high diffusion signal from the mass (arrow)
F. Axial T2-weighted image demonstrates high heterogeneous signal from the mass lesion in the right parotid gland (arrow)

Discussion

Acinic cell carcinoma is a rare form of parotid cancer. It accounts for approximately 1% of parotid cancers. Acinic cell cancer usually grows relatively slowly. This type of cancer has a good prognosis with an almost 90% 10-year survival. It is considered a low-grade malignancy.

Masses in the tail of the parotid gland can be a source of consternation to the radiologist and the clinician. The most common lesion in the parotid gland is pleomorphic adenoma followed by Wharton’s tumor, infectious processes, venous malformation, Sjögren’s disease, lymphatic malformation, lipoma, and HIV related to epithelial lesions. The malignant lesions include non-Hodgkin’s lymphoma, metastatic disease, mucoepidermoid carcinoma, acinic cell carcinoma and undifferentiated carcinomas. Parotid tumors are in general unusual in pediatric patients and acinic cell carcinomas in this patient group are exceedingly rare. The imaging findings are nonspecific and, based on the MR imaging, we would not be able to tell whether this lesion is malignant or benign, or whether it represents a specific type of parotid tumor. There are no soft tissue abnormalities around it to indicate an infection and therefore, from an imaging point of view, it is relatively clear that it represents a neoplasm benign or malignant.

Suggested Reading

Figure 9.12
Acinic cell carcinoma of the parotid gland
Anophthalmos

Clinical Presentation

A 2-year-old female with congenital anophthalmos and seizures.

Images (Fig. 9.13)

A. Sagittal T1-weighted image demonstrates hypoplasia of the orbital region and absence of the orbit. The splenium of the corpus callosum is somewhat small
B. Axial T2-weighted image demonstrates the absence of both eyes
C. Sagittal T1-weighted image through the orbit shows anophthalmos with a malformed congenitally small globe
D. Axial T1-weighted image demonstrates bilateral anophthalmos with micro-optic features

Discussion

Anophthalmos, congenital absence of an eye or eyes, is a rare anomaly that occurs as a result of insults to the developing eye during first 8 weeks of life. Anophthalmos can be of three types: (1) primary anophthalmos which is usually bilateral and sporadic and occurs when the optic primordial does not develop, (2) secondary anophthalmos which is an extremely rare and lethal anomaly that occurs when the entire neural tube fails to develop, and (3) secondary anophthalmos that occurs when the optic vesicle forms but subsequently degenerates. The diagnosis of true anophthalmos can be made when there is complete absence of the ocular tissue within the orbit. Extreme microanophthalmos is seen more commonly in which a very small globe is present within the orbital soft tissue, which is not visible on initial examination. The orbital findings include small orbital rim with absence of extraocular muscles and lacrimal gland. The optic foramen is small and maldeveloped. The globe is completely absent in primary anophthalmos.

CT and MR imaging is performed to assess the presence of an extremely microanophthalmic globe. The optic chiasm and corpus callosum may show agenesis or dysgenesis. Craniofacial anomalies may also be associated.

Suggested Reading

Figure 9.13
Anophthalmos
Wolf-Hirschhorn Syndrome

Clinical Presentation

A 13-month-old male with seizures, developmental delay, sacral dimple and a clinical diagnosis of Wolf-Hirschhorn syndrome.

Images (Fig. 9.14)

A. CT scan through the orbits shows hypertelorism
B. Sagittal MR image shows absence of the posterior portion of the body of the corpus callosum (arrow). There is micronathia with a small mandible and a relatively small maxilla
C. Axial T2-weighted image demonstrates low signal intensity in the basal ganglia bilaterally presumed to be secondary to early iron deposition. There is also decreased signal intensity in the cortical gyri presumed to be from the same etiology

Discussion

Wolf-Hirschhorn syndrome is caused by partial deletion of chromosome 4. Wolf-Hirschhorn syndrome has distinctive facial characteristics – prominent head, wide eyes, and broad beaked nose, collectively described as “Greek warrior helmet” features. Other clinical features include microcephaly, profound mental retardation, growth retardation, muscular hypotonia, seizures, congenital heart defects, coloboma of iris, genital and renal anomalies.

On imaging, findings of absent cavum septum pellucidum, agenesis of the corpus callosum, microgyria, migration defects and hydrocephalus may be seen. In addition, MR imaging shows multifocal white matter lesions.

Suggested Reading

Retinopathy of Prematurity

Clinical Presentation

A 21-month-old ex-premature infant with a history of intraventricular hemorrhage and developmental delay. Impaired left sided vision.

Images (Fig. 9.15)

A. Sagittal T1-weighted image shows a small globe with intermediate signal
B. T1-weighted image shows a small globe on the left side with intermediate signal. The retina is displaced anteriorly and compressed (arrow)
C. T2-weighted axial image shows evidence of retinal detachment on the left side with a small globe (arrow)

Discussion

Retinopathy of prematurity is caused by the vasoconstrictive effect of high blood levels of oxygen used to treat hyaline membrane disease in premature infants. The vasoconstriction results in chronic retinal ischemia which secondarily causes neovascularization. The neovascularization and its following regression cause subretinal exudation, hemorrhage and scarring. The scarring and the subretinal exudate often result in chronic retinal detachment and later on microophthalmia.

Most patients have bilateral imaging findings but often are asymmetrical. MR imaging of acute subretinal fluid will be high on CT images due to the presence of acute hemorrhage. MR imaging of the acute condition will show high signal on T1 and low signal on the T2 images. Chronic detachment will have low signal with no attenuation on CT images, intermediate to low signal on T1-weighted images, and often high or variable T2 signal.

More than 80% of infants weighing less than 1 kg at birth will develop retinopathy of prematurity. The main risk is administration of excessive oxygen. The diagnosis is generally made by ophthalmological examination. The treatment is mainly prophylactic, but severe cases can be treated with cryotherapy and laser to ablate the peripheral avascular retina.

Suggested Reading

Hemangioma

Hemangioma of the Parotid Gland

Clinical presentation

A 9-month-old female presents with a soft asymptomatic swelling of the left parotid gland.

Images (Fig. 9.16)

A. Axial T1-weighted image shows a well-circumscribed mass in the left parotid gland (arrow). There is no reaction in the surrounding fat.
B. Axial T2-weighted image with fat suppression shows the mass having high signal (arrow).
C. Axial T1-weighted image with contrast shows enhancement of the well-circumscribed mass (arrow). There is a rim of nonenhancing parotid gland tissue around the mass.
D. Coronal T1-weighted image with contrast shows enhancement of the well-circumscribed mass. The rim of nonenhancing parotid gland tissue around the mass is well seen (arrow).
Hemangioma of the Face and Orbit

Clinical Presentation

A 4-month-old baby presents with a soft periorbital mass.

Images (Fig. 9.17)

A. Axial T1-weighted image shows a left periorbital and a cheek soft tissue mass (arrow)
B. Coronal T1-weighted image shows a soft tissue mass in the left face (arrow)
C. Axial T2-weighted image with fat suppression demonstrates the precise outline of the soft tissue hemangioma and the left facial structures
D. Axial contrast-enhanced T1-weighted fat-suppressed image shows enhancement of the left cheek mass (arrow)

Discussion

Hemangioma is the most common vascular tumor seen in infancy. The incidence is higher in females and in low-birth-weight babies. Hemangiomas are most prevalent in the head and neck region and constitute 18–38% of head and neck tumors. Approximately 20% of patients have multiple hemangiomas that involve sites such as skin, liver, gastrointestinal tract and brain. Intracranial and intraspinal hemangiomas may also be seen in association with multiple hemangiomas. Diagnosis of hemangiomas is made by a combination of the medical history, physical examination and ultrasound scan. Typical hemangiomas are red, raised and bosselated. Deep hemangiomas have normal overlying skin and may mimic other vascular malformations. Congenital hemangiomas typically show rapid growth and may involute completely.

On CT images, hemangiomas are seen as lobulated solid masses that are isodense with muscle and show intense enhancement. On MR images, they usually show intermediate signal intensity on T1-weighted images, high signal intensity on T2-weighted images, and diffuse intense enhancement. Areas of fatty re-
placement may also be seen. Hemangiomas need to be differentiated from arteriovascular malformations which are also associated with prominent vascularity. However, hemangiomas usually have a lobulated appearance and are not associated with reactive or trophic changes which are commonly associated with arteriovascular malformations.

**Suggested Reading**


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**Langerhans Cell Histiocytosis**

**Clinical Presentation**

A 12-year-old with a history of Langerhans cell histiocytosis presents with decreased vision in the right eye.

**Images (Fig. 9.18)**

A. Axial CT scan shows a mass in the right orbital apex with erosion of the posterolateral wall of the right orbit (arrow)

B. Coronal reformatted CT scan demonstrates the mass filling the entire right orbital apex (arrow). The left orbit is normal

C. Coronal T1-weighted image demonstrates an intermediate to low signal intensity mass filling the right orbital apex (arrow). There is also expansion of the orbital apex

D. Sagittal T2-weighted fat-suppressed image demonstrates a high signal intensity well-demarcated mass in the right orbital apex. This is consistent with a Langerhans cell histiocytosis in a patient with a prior diagnosis of this condition

**Discussion**

Langerhans cell histiocytosis is an uncommon disease characterized by the idiopathic proliferation of Langerhans cells or their marrow precursors. Langerhans cell histiocytosis is classified according to sites of involvement into single or multisystem disease. Single system can be unifocal or multifocal. Bony involvement is seen in 78% of patients and often includes the skull (49%), innominate bone, femur, orbit (11%), and ribs. Extraskeletal involvement is well known. The lesions can be single or multiple. Orbital infiltration presents with pain, swelling and proptosis. Orbital soft tissue involvement without an obvious bony defect is rare. On CT images, they may be seen as a homogeneously hyperdense enhancing masses associated with bony erosion. On T1- and T2-weighted and proton density MR images, they are seen as isointense to gray matter and show enhancement.

**Suggested Reading**


Figure 9.18
Langerhans cell histiocytosis
Osteosarcoma of the Cranium

Clinical Presentation

A 10-year-old presenting with a painless reddish swelling over the right periorbital zygoma region. The patient had a history of retinoblastoma and radiation.

Images (Fig. 9.19)

A. Axial CT image shows an expansile bony lesion in the right orbit
B. Coronal contrast-enhanced T1-weighted image shows the expansile lesion invading both the orbit and the brain
C. Sagittal contrast-enhanced T1-weighted image shows the invasion of the mass into the anterior and middle cranial fossae
D. Axial T2-weighted image shows an irregular low-intensity mass in the right periorbital region invading the orbit and infratemporal fossa. It also extends into the right middle cranial fossa.

Discussion

Osteosarcomas are malignant bone tumors, which commonly affect the long bones of young adults. Primary osteosarcomas of the skull are rare. Secondary osteosarcoma may be seen in patients with long-standing Paget’s disease. The post-radiation osteosarcomas occur in portions of bones at the borders of the radiation field.

Cranial radiographs are of limited value in head and neck osteosarcomas due to superimposed bony structures. CT scanning provides excellent detection of tumor calcification, and cortical involvement. MR imaging is more sensitive in demonstration of intramedullary and extraosseous tumor components.

Suggested Reading

Figure 9.19
Osteosarcoma of the cranium
Fibrous Dysplasia

Clinical Presentation

A 17-year-old presents with asymmetry of the face and skull. There is a painless prominence in the left supraorbital region.

Images (Fig. 9.20)

A. Coronal CT scan shows a dense expansile greater wing of the sphenoid bone on the left side. There is decreased size of the left orbital cavity.
B. Contrast-enhanced T1-weighted MR image shows an isointense lesion in the expanded greater wing of the sphenoid bone. The orbital cavity is compressed.
C. T1-weighted image without contrast shows low signal intensity from the fibrous lesion in the greater wing of the sphenoid bone on the left side consistent with fibrous dysplasia.

Discussion

Fibrous dysplasia is an idiopathic skeletal disorder in which the medulla bone is replaced by poorly organized, structurally, and sound fibro-osseous tissue. It most frequently affects children, teenagers and patients under the age of 30 years. The majority of the cases are mono-osteitic, and this condition affects the ribs, femur and craniofacial skeleton. Albright’s syndrome is a variant that consists of poly-osteitic fibrous dysplasia, pigmented skin pigmentation and sexual precocity. Albright’s syndrome is relatively rare and it has been stated that it occurs 40 times less than mono-osteitic fibrous dysplasia.

Malignant transformation of fibrous dysplasia is very rare (less than 1%). Fibrous dysplasia is typically painless but neurovascular compromise may cause symptoms. Craniofacial fibrous dysplasia is generally not functionally devastating unless in this case there is involvement of the orbit and skull base. There is a potential for optic nerve compression, pituitary dysfunction and compromise of other vital neurovascular structures. The disease is usually self-limiting and
often does not progress after the third decade of life. Surgical treatment is limited to cosmetic debulking and re-contouring of the bone.

**Suggested Reading**


**Bilateral Coronoid Hyperplasia of the Mandible Causing Trismus**

**Clinical Presentation**

A 16-year-old male presenting with gradually increasing trismus. He has no pain and he is able to move his jaw from left to right with a full range of motion, but he is not able to open more than 28 mm (normal 40 mm or more). There is no tenderness over the temporomandibular joints and no clinical suspicion of internal derangement.

**Images (Fig. 9.21)**

A. Three-dimensional CT scan with closed mouth shows elongation of the left coronoid process. The coronoid process is extending above the zygomatic arch

B. Three-dimensional CT scan with open mouth shows the coronoid process behind and interfering with the zygoma. There is normal range of motion in the temporomandibular joint

**Suggested Reading**


Figure 9.21
Bilateral coronoid hyperplasia of the mandible causing trismus
**Dental Radicular Cyst**

**Clinical Presentation**

A 13-year-old with an asymptomatic lucency on a panoramic radiograph was referred for MR imaging.

**Images (Fig. 9.22)**

A. T1-weighted axial image shows expansion of the left body of the mandible with a low signal intensity mass
B. T2-weighted image shows high signal in the expansile lesion in the left body of the mandible
C. Sagittal T2-weighted image with fat suppression demonstrates the lesion inferior to the first molar and premolar
D. Coronal T2-weighted image shows the expansion of the mandible on the left side with scalloping of the inner cortex
E. Coronal T1-weighted image post-contrast demonstrates no enhancement of the lesion but minimal enhancement of the wall of the cystic cavity

**Discussion**

Radicular cyst is the most common odontogenic cyst. The peak incidence is between 30 and 50 years of age, but it may occur in young individuals as in this 13-year-old patient. Caries leading to pulp infection and eventually necrosis is the most common cause of radicular cyst. Radiographically a radicular cyst is a well-circumscribed radiolucency arising from the apex of the tooth and is nearly always bounded by a thin rim of cortical bone. It is not uncommon to see expansion and scalloping of the cortical margins of the jaw. Radiographically a radicular cyst cannot be differentiated from a periapical granuloma. On MR imaging they typically have a low T1 and a high T2 signal intensity with no enhancement. A radicular cyst is usually treated endodontically or by extraction of the involved tooth and surgical enucleation of the cyst. There is no significant recurrence rate.
Chapter 9

Suggested Reading


Mastoiditis with Sigmoid Sinus Thrombosis

Clinical Presentation

A 7-year-old female with a chronic draining right ear.

Images (Fig. 9.23)

A. Axial CT scan demonstrates opacification of the entire right mastoid and the right middle ear cavity without appreciable bony erosions

B. Axial T1-weighted image without contrast shows a soft tissue mass in the right temporal bone (arrow). There is no flow void in the sigmoid sinus on this side
C. Axial contrast-enhanced T1-weighted image with fat suppression demonstrates abnormal contrast enhancement in the area of the right temporal bone (arrow). In the sigmoid sinus there is contrast enhancement in the periphery but a central filling defect suggesting a clot in the sigmoid sinus

D. Axial DW image shows high signal in the clot in the right sigmoid sinus suggesting restricted diffusion secondary to sigmoid sinus thrombosis

E. MR venogram in the axial plane demonstrates lack of flow in the right sigmoid and transverse sinus

F. Coronal venogram demonstrates no flow in the right sigmoid sinus

**Discussion**

Many predisposing factors have been implicated in the development of sinus thrombosis. Trauma, infection, tumors, dehydration, hypercoagulable states such as pregnancy, oral contraceptives and nephrotic syndrome are the most common causes. Approximately 20% of cases are idiopathic. Mastoiditis is a known cause of lateral venous sinus thrombosis. The sigmoid portion of the lateral venous runs on the inner aspect of the mastoid process. Diploic veins and small veins from the middle ear also drain into the lateral sinus. Patients presenting with lateral sinus thrombosis and mastoid congestion should be assessed for the presence of mastoiditis.

Dural sinus thrombosis manifests with diverse clinical findings. Early symptoms include headache, lethargy followed by seizures and focal neurological deficits. Strokes (hemorrhagic) may develop secondary to poor venous drainage. These strokes are often bilateral and outside the normal arterial distribution, reflecting the pattern of venous drainage.

MR venography is the modality of choice for venous thrombosis. The hyperacute thrombus has low signal intensity on both T1- and T2-weighted images. The effect is more pronounced on T2-weighted images. At about 3 weeks, the clot may have low signal on all sequences. Indirect signs of thrombosis include the presence of collateral flow.

**Suggested Reading**


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**Arteriovenous Fistula of Vertebral Artery**

**Clinical Presentation**

A 6-year-old male with right neck bruit.

**Images (Fig. 9.24)**

A. Sagittal T2-weighted image shows an intraspinal vascular malformation with abnormal vessels (arrow)

B. Axial T2-weighted image shows a large abnormal vascular connection between the right vertebral artery and the spinal canal (arrow). There is also an abnormal flow void in the anterior of the spinal canal

C. Contrast enhanced MR angiogram demonstrates the vascular malformation connecting the vertebral artery and the venous system on the right. There is a relatively broad and complex connection between the artery and the vein: arteriovenous fistula

D. FSPGR MR image shows the arterial and the venous side of the arteriovenous fistula but not the connection itself

**Discussion**

An arteriovenous fistula involving the cervical vertebral artery is rare. The etiologies of vertebral arteriovenous fistula include traumatic, iatrogenic, congenital, and spontaneous (neurofibromatosis). Traumatic and iatrogenic injuries appear to be the most common causes, with the greatest incidence arising from firearm and stab wounds. Symptoms include tinnitus and presence of a pulsatile mass with a thrill.

Color Doppler examination is a noninvasive modality which can be used as bedside screening investigation for detection of vertebral arteriovenous fistula. CT angiography and MR angiography demonstrate the anatomical relationship of the fistula accurately. Conventional angiography serves as a diagnostic as well as a therapeutic tool for interventional procedures. The current therapeutic management consists of direct closure of the fistula, either with surgical techniques or percutaneously with detachable balloon occlusion or coil embolization or stent grafts.
Suggested Reading