Principles and Practice of Radiological Investigations for the Diagnosis of Pediatric Head and Neck Diseases

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Introduction

The head and neck location includes the face, eye and orbit, nasal cavity and paranasal sinuses, ear and temporal bone, oral cavity, jaw, and neck. Head and neck pathologies in pediatric population differ from adults by the types and nature of occurrence.

Neck pathologies in children commonly have an inflammatory, infective, or congenital cause. Neoplastic lesions are less common comprising of only 5% of childhood cancers arising in the head and neck region [1]. Imaging is necessary to characterize and assess the extent of head and neck mass lesions. In case of malignant disease, it helps to analyze the metastatic spread and in follow-up to detect the response to treatment. Imaging can also be used to guide fine-needle aspiration (FNA) or needle biopsy. In this chapter, we describe various imaging techniques used to assess lesions of the head and neck in children, guidelines and indications of their utility, radiation and contrast-related concerns specific to children and then review imaging findings of ten important subsets of head and neck pathologies in pediatric population.

Important clinical features that need to be considered when interpreting imaging include [2]: age of the child, duration and nature of onset of the mass, location of the mass, characteristic imaging features (e.g., mineralization, vascularity, intensity of enhancement, cystic areas, diffusion restriction) and whether the child has a known mutation.

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Why Is Pediatric Imaging Challenging?

Imaging kids is challenging as compared to the imaging of adults. This is because of the following:

- It requires dedicated imaging protocols for image acquisition with special considerations for signal to noise due to smaller sizes.
- There is need for sedation or general anesthesia for procedures like MRI, which takes longer time.
- Specific training is required for the healthcare personnel involved. Good knowledge and expertise need to be applied for evaluating the images.
- It also requires an understanding of optimum radiation exposure, if ionizing radiation is being used.

Preferences in Pediatric Imaging

Environment

The objective of acquiring good quality images in children involves gaining a child's trust and co-operation before and throughout the examination. Children are irritable and not comfortable around strangers and unfamiliar environments. To encourage improved patient experience, the external environment in the radiology department should be made more child-friendly, for example, walls can be painted with colorful characters (Fig. 2.1), and there can be children's books in the waiting room. Support from parents is always appreciated. It may be sometimes necessary to sedate the child or use immobilizers for longer studies like MRI. Commonly used sedatives are diazepam, midazolam, and ketamine. Techniques and equipment need to be hired to minimize the need for sedation as it has its own harmful effects. Where sedation or anesthesia is required, there should be dedicated pediatric specialists who help in recovery. Child life specialists and Innovative methods like dog therapy and virtual reality are also successful in

P. Campisi et al. (eds.), *Pediatric Head and Neck Textbook*, https://doi.org/10.1007/978-3-030-59265-3_2

Fig. 2.1 Showing the colorful walls leading to MRI at SickKids Hospital, Toronto

avoiding sedation and anesthesia in appropriately selected subjects [3]. The emotional needs of parents should also be addressed when considering any pediatric imaging service.

Equipment and Protocols

Dedicated pediatric imaging department with dedicated pediatric technologists may result in greater compliance with the pediatric protocols and significantly reduced patient dose. Imaging needs to be child-focused and must be tailored according to the age of the child. The standardization of the techniques and protocols is also important. Child-appropriate protocols need to be implemented for all the modalities.

Training

It must be ensured that only the staff with appropriate training in Pediatric imaging are employed and their performance should be reviewed regularly. Also, radiologists reporting pediatric cases must have a good knowledge and expertise, as pathologies afflicting children are different and peculiar than that occur in adults. Also, because of the dynamic anatomy in children, normal variants look like pathology. Proper knowledge of these variants helps avoid making such mistakes. The pediatric radiologist also decides the appropriate modality of choice.

Quality Assurance

Regular audits and quality checks for the equipment need to be ensured for optimum performance and calibration for pediatric use. There should also be quality assurance with respect to technique and reporting.

Radiation Protection

- Considerable advances in technology have reduced the amount of radiation required to achieve images of diagnostic quality. Although stochastic effects have not been demonstrated at radiation doses encountered in diagnostic radiology, the risk for cancer induction is thought to be greater in children than adults [4]. Significant efforts must be made to ensure that delivered radiation doses are low.
- Measured application of ionizing radiation remains of critical importance and must be catered to clinical appropriateness and diagnostic task of the examination.
- Development of a comprehensive quality assurance and quality control program is essential; ensuring that X-ray equipment is functioning within specific tolerances, delivering the exposures expected and appropriate, and is compliant with acceptable standards of installation and design. It is critical that radiologists, radiographers (medical radi-

ation technologists), and physicists develop standards for each institution.
- Creation of institutional diagnostic reference levels (DRL) are necessary to ensure that radiation dose metrics are known, expected, and within specified and reasonable tolerances.

Concerns which need to be considered while using conventional radiographic X-ray equipment for imaging in children include the following: [4]

- Appropriate imaging protocols: must be applied for the diagnostic task, the body part to be imaged, and patient habitus
- X-ray tube and generator: must operate at a high frequency, have sufficient power to facilitate short exposure times, and a large dynamic range of tube current and tube current-time product to accommodate a large range of body habitus.
- Automatic exposure control (AEC): should be used in appropriate circumstances
- Spectral filtration: the addition of aluminum and copper filters improves beam quality and reduces the proportion of lower-energy photons contributing to skin dose while improving penetration
- Antiscatter grid: use of a grid may improve image contrast by reducing the proportion of scatter reaching the image receptor
- Beam collimation needs: primary radiation must be limited to the area of interest
- Immobilization devices should be used where applicable to reduce patient motion
- Use of computed or digital radiography is recommended

Concerns which need to be considered while using computed tomography (CT) for imaging in children include the following: [5]

- Dedicated CT protocols for pediatric imaging must be implemented for all examinations to ensure appropriate image quality and radiation dose. This involves customization of tube voltage, tube current, tube current-time product, tube-current-modulation, image thicknesses, convolution kernel, and reconstruction technique (filtered back projection, iterative reconstruction, etc.)
- Limit scan range to indication
- A lower tube voltage can reduce radiation dose when used in studies with iodinated contrast
- Shorter rotation times can reduce examination time and minimize the likelihood of patient motion
- Patients must be centered in the gantry to ensure appropriate tube-current-modulation
- Limit number of phases for IV contrast examinations and consider noisier technique for select phases if appropriate

Advantages and Limitations of Each Imaging Modality in Head and Neck Location: [6]

The indication for diagnostic imaging in the anatomically complex head and neck region should be established for a specific type of imaging modality on the basis of a thorough clinical examination. Main advantages and limitations of imaging modalities in the head and neck region are listed below.

1. Ultrasonography
   - **Advantages**
     - Widely available
     - No exposure to ionizing radiation
   - **Limitations**
     - Limited to superficial regions
     - Diagnostic value is examiner-dependent

2. Nuclear Imaging/PET CT
   - **Advantages**
     - Whole-body evaluation
     - Functional, not merely anatomical, evaluation
   - **Limitations**
     - Limited structural information
     - Cancer-specific diagnostic value of positron emission tomography (PET) not generally accepted

3. Conventional X-rays
   - **Advantages**
     - Low cost
     - Low radiation exposure
   - **Limitations**
     - Poor risk–benefit profile, owing to diagnostic uncertainty from projection effects
     - Therefore, restricted to certain specific indications (e.g., dental diagnosis)

4. Computed Tomography (CT)
   - **Advantages**
     - 3D sectional imaging technique with high diagnostic value
     - Widely available
     - Best risk–benefit profile for standard care
   - **Limitations**
     - Low-dose protocols have not yet come into use in all centers

5. Cone Beam CT (previously also known as Digital Volume Tomography)
   - **Advantages**
     - 3D sectional imaging technique
     - High spatial resolution
     - Usually low radiation exposure (but depends on apparatus and examiner)
   - **Limitations**
     - Cannot be used to examine soft tissues, incl. tumors

6. Magnetic Resonance Imaging (MRI)
   - **Advantages**
     - 3D sectional imaging technique with the highest diagnostic value
     - Best modality for imaging the soft tissues
     - No exposure to ionizing radiation
   - **Limitations**
     - Requires expensive special equipment

7. Cerebral Angiography
   - **Advantages**
     - Evaluates arteries of the head and neck before surgery
     - Provides additional information on abnormalities seen on MRI or CT of the head, such as the blood supply to a tumor
     - Basis for treatment, such as embolization of a neoplasm
     - In preparation for minimally invasive treatment of a vessel abnormality
   - **Limitations**
     - Invasive; procedure-related complications
     - Involves ionizing radiation
     - Contrast-related risks

Indications of the various imaging modalities in head and neck pathologies (6):

Conventional Radiographs (Plain X-rays):

Conventional Radiographs now have a limited value for imaging in the head and neck area. They have been almost entirely replaced by cross-sectional imaging, except for a few specific indications like diagnostic assessment of the teeth and jaws.

- *PNS (Paranasal Sinus)*

Similarly, conventional X-rays of the paranasal sinuses are not indicated for screening purposes, for example, to evaluate headache, cystic fibrosis in children, asthma, or allergies; or for the detection of an infectious focus in patients with unclear inflammatory symptoms or as an exclusion of a particular disease in persons with an elevated risk.

- *Skull*

Some indications for conventional skull films still indicated are as follows: to exclude isolated fractures of the zygomatic bone, maxilla, mandible, or nasal bone; to diagnose congenital anomalies and premature synostoses; to demonstrate pneumocephalus after intracranial procedures; to detect metallic foreign bodies before MRI; and to check the setting of a programmable ventriculoperitoneal shunt.

Plain films of the skull were once commonly obtained in the evaluation of trauma cases. However, skull fractures that were recognizable on projection views poorly correlated with intracranial injuries, while the clinically relevant entity is not the skull fracture itself, but rather the intracranial hemorrhage that is associated with it. Jend et al. in their study reported that only 40% of patients with a skull fracture had an associated intracranial injury as well; on the other hand, 44% of patients with an intracranial injury demonstrated no skull fracture [7]. Thus, CT is clearly the modality of choice.

- *Orbit*

Convention radiographs were used in the past almost exclusively for the evaluation of trauma cases. Conventional orbital X-rays have now been replaced by tomographic imaging for nearly all indications. As an exception, they can still be used to rule out the presence of metallic foreign bodies before MRI. Conventional orbital X-rays are not mentioned in the current AWMF guidelines; they are obsolete for orbital diagnosis.

- *Temporal Bone*

The main conventional X-ray views of the temporal bone are Stenvers and Schüller. Stenvers view is still used today to document the position of the electrode carrier for cochlear implantation. However, it is obsolete for all other indications. Schüller view yields some information of the degree of pneumatization of the mastoid bone but does not permit any validity whether diminished pneumatization is due to a congenital anomaly, tympanic sclerosis, or chronic inflammation. Schüller views are still occasionally obtained in patients with suspected mastoiditis or otitis media; however, this has no medical justification.

Conventional temporal bone X-rays are not approved to be taken preoperatively to demonstrate anatomical relationships, as an aid to surgery: projection effects make them unreliable for the identification and quantitative measurement of surgically relevant anatomical variants. Temporal bone X-rays are also not indicated for the assessment of trauma, malformations, and tumors of the temporal region. They have been replaced by sectional imaging—CT, DVT, or MRI, depending on the indication.

- *Dental and Maxillary Region*

The primary imaging modality for assessment of the teeth and jaws is still conventional radiography: specifically, intraoral dental views or an (extra oral) panoramic tomographic view (orthopantomogram, OPG). Intraoral dental views pro-

vide the assessment of endodontal and periodontal disease with high local resolution and minimal radiation exposure. OPG, on the other hand, enables a comprehensive survey of all of the teeth and the underlying bone, with a low radiation exposure.

Dental views and OPG are currently used for evaluation of inflammatory diseases, orthopedic evaluation of the jaws, trauma assessment, evaluation of unclear symptoms, and planning of dental implantation procedures. Conventional lateral views are used (optionally) in orthognathic surgery.

Tomographic imaging is indicated for the evaluation of large cysts, benign or malignant lesions of the jaws and for the evaluation of trauma with potentially extensive midface involvement. It is also used to plan implantation procedures. Compared to conventional X-rays, it gives a more accurate assessment of the bone substance, exact measurement of the height and width of the jaws, three-dimensional localization of the mandibular nerve canal, and an assessment of the topography of the maxillary sinuses and of inflammatory processes than may affect them.

Cone Beam CT (CBCT) is superior to conventional X-rays (dental views and OPG) for the evaluation of dental trauma. The preferred methods of tomographic imaging are CT and CBCT. Both modalities are subject to the same restrictions with regard to radiation safety.

Ultrasonography

Diagnostic ultrasonography of the head and neck is mainly used to assess organs and lesions that lie near the surface, including the salivary glands, the thyroid gland, the major vessels, enlarged superficial lymph nodes, and other superficial pathologic lesions.

Superficial palpable masses of the head and neck are also common in the pediatric population, with the vast majority of the lesions ultimately proven to be benign. Duplex ultrasonography (US) is the first-line imaging modality for the evaluation of superficial pediatric masses. Without utilizing radiation, iodinated contrast material, or sedation and/or anesthesia, US provides a means for quick and cost-effective acquisition of information, including the location, size, shape, internal content, and vascularity of the lesion. US is usually performed in an array of common and uncommon pediatric head and neck masses that include neonatal scalp hematoma, craniosynostosis, dermoid and epidermoid cysts, Langerhans cell histiocytosis, lymph nodes and their complications, fibromatosis colli, thyroglossal duct cyst, branchial cleft cyst, cervical thymus, congenital goiter, thyroid papillary carcinoma, parathyroid adenoma, hemangioma, lymphangioma, jugular vein phlebectasia, Lemierre syndrome, acute parotitis and parotid abscess, leukemia and/or lymphoma, neurogenic tumor, and rhabdomyosarcoma. Finally, in situations where the head or neck mass is too large and deep, or if it is hyperechoic to be fully assessed within the US field of view, or if malignant or a high-flow vascular lesion is suspected, then further evaluation with cross-sectional imaging becomes necessary [8].

Computed Tomography

CT is the most commonly used imaging modality for all indications in the head and neck region. The spiral CT mode currently involves the acquisition of only one thin-section axial volume data set. From this data set, tomographic images in all of the required planes can be computed without any further radiation exposure or loss of image quality.

Intravenously administered contrast media improve the delineation of soft-tissue pathologies and are in-dispensable in the diagnosis of malignant tumors and inflammatory complications.

Low-dose CT is the imaging modality of choice for chronic rhinosinusitis. The preoperative CT reveals the site and extent of chronic inflammatory changes that have not responded to conservative treatment; it also documents any anatomical variants that may have contributed to the causation of sinusitis or that might be danger areas for the current state-of-the-art, minimally invasive, endoscopic surgical approach.

The imaging modality of choice for trauma involving the orbit, midface, and skull base is again thin-section CT. Multiplanar and three-dimensional reconstructions of the CT data set yield the details that are needed for the comprehensive assessment of complex fractures affecting the entire midface or any part of it. The proper imaging study to evaluate visual disturbances is an MRI or CT of the whole cranium, orbits included.

CT is the preferred modality for the assessment of trauma, aggressive inflammatory diseases and other extracranial processes, and conductive or mixed hearing loss, as well as for the planning of cochlear implantation and other surgical procedures in the temporal region.

Cone Beam CT

CBCT is a sectional imaging modality similar to CT that was used initially only for dental diagnosis because of the restriction to small volumes. Technical advances have made CBCT applicable in larger volumes, and it can now be used as an alternative to CT for evaluation of the craniofacial and temporal high-contrast structures.

The advantages of CBCT are high spatial resolution, low radiation exposure (in the same range as low-dose CT), and decreased metal artefact. It is unsuitable for soft-tissue diag-

nosis as the image-noise is too high. Apart from clinical use in the diagnosis of high-contrast structures like teeth and jaws, a definite judgment, mainly as an alternative method to CT, is not possible as yet. Therefore, the current guidelines designate CBCT as a possible alternative to CT in individual cases but provide no specific recommendations regarding its use.

Magnetic Resonance Imaging

MRI is currently the imaging modality that yields the most detailed view of the soft tissues. Its main advantage, in comparison to CT, is the absence of ionizing radiation; its main disadvantage is the much longer time during which the patient must keep still during the study. Patients who cannot cooperate may need sedation or even general anesthesia. In the head and neck, MRI is mainly used for pre- and postoperative tumor imaging and to evaluate suspected intracranial complications of sinusitis.

MRI is the imaging study of first choice for the evaluation of orbital tumors or endocrine orbitopathy, after ultrasonographic examination by an ophthalmologist. CT can be particularly useful for the demonstration of calcifications or bony changes.

MRI is the method of choice to assess anomalies of the inner ear, sensorineural hearing loss/deafness, dizziness, and intracranial processes.

Other rare indications include congenital anomalies of the temporal bone, as well as the preoperative assessment for cochlear implants. MRI can be used instead of CT as the primary imaging modality whenever ionizing radiation is to be avoided, for example, in children who need imaging of the paranasal sinuses before surgery.

Nuclear Isotope Imaging

Nuclear isotope imaging plays a key role in the evaluation of thyroid disease. Bone scanning can be used to evaluate craniofacial or other skeletal involvement by chronic inflammatory or neoplastic entities. Positron emission tomography (PET) gives the details of whole-body functional imaging with good spatial resolution. [18F] Fluorodeoxyglucose ([18F] FDG) PET has a wide range of applications in staging oncological disease and monitoring response to treatment. Moreover, the increasing availability of PET-CT allows improved localization and definition of disease activity. The role of PET and PET-CT in childhood malignancies continues to evolve, but it is widely used in the staging and follow-up of lymphoma and also have a role in soft tissue sarcomas.

Contrast Media in Children

We address specific areas in which pediatric use of contrast material differs from adult use.

Iodinated Intravascular Contrast Media

Unique Considerations in Children

Contrast Agent Osmolality

Osmolality is an important physical property of contrast media. A variety of adverse effects attributed to intravascularly administered iodinated contrast agents seem to be related, at least in part, to this physical property, including physiologic side effects, allergic-like reactions, complications following contrast medium extravasation, and fluid shifts.

Contrast media osmolality is of particular importance in neonates and small children. These patients are thought to be especially susceptible to fluid shifts and have a lower tolerance for intravascular osmotic loads when compared to adults. Intravascular administration of hyperosmolar contrast medium may result in migration of fluid from extravascular soft tissues into blood vessels, consequently expanding blood volume [9, 10].

If the fluid shift is large, cardiac failure and pulmonary edema can result; children with significant preexisting cardiac dysfunction may be at particular risk.

Contrast Media Viscosity

Viscosity, a measure of fluid resistance to stress, is another important physical property of contrast media. As viscosity increases, the pressure associated with an intravascular contrast medium injection increases. This physical property is especially important for pediatric patients due to the use of small gauge angiocatheters in tiny blood vessels. Contrast medium viscosity and angiocatheter size are important factors in determining maximum injection rates. If a rapid injection rate is desired through a small angiocatheter and if contrast medium viscosity is high, two problems can potentially result: First, the desired injection flow rate may not be achieved. Second, high pressure may cause catheter failure and/or vessel injury.

Additionally, contrast medium viscosity is not directly proportional to the concentration of iodine iscosity of contrast media is affected by temperature. As temperature increases, viscosity decreases, allowing for increased flow rates at lower pressures. A study by Vergara and Seguel [11] that included both adult and pediatric patients showed that warming contrast media resulted in fewer adverse events following injection when compared to contrast media adminis-

tered at room temperature. In another study of 24,826 intravenous (IV) contrast material administration in children and adults [12], warming of iopamidol-370 to body temperature reduced the extravasation rate, but warming of iopamidol-300 to body temperature had no effect. The authors concluded that higher viscosity agents may benefit more from warming than lower viscosity agents.

Other Unique Concerns in Children

Several additional issues complicate the administration of intravascular contrast media to neonates and children, including the use of small volumes of contrast medium, the use of small gauge angiocatheters, and unusual vascular access sites. First, very small volumes of contrast media are typically administered to neonates and infants (typically 1.5–2 mL/kg) [13]. As a result, timing of image acquisition with regard to contrast medium administration may be important when performing certain imaging studies, such as CT angiography. In some instances, a slower injection rate (compared to that used in older children and adults) may be useful to prolong intravascular enhancement. Second, small-gauge angiocatheters (e.g., 24-gauge) located in tiny peripheral veins (e.g., in the hand or foot) are commonly utilized in neonates and infants.

A study by Amaral et al [14] showed that 24-gauge angiocatheters in a peripheral location can be safely power injected using a maximum flow rate of approximately 1.5 mL/sec and a maximum pressure of 150 psi. When access is thought to be tenuous, hand injection of contrast medium should be strongly considered to minimize the risk of vessel injury and extravasation. Since many currently used central venous catheters are not approved for power injection, one should always verify in advance that any catheter to be utilized for bolus contrast material instillation can tolerate the anticipated injection. It is also important to ensure that the pressure used does not exceed the catheter's pressure rating.

Particular attention should be paid to the injection sites of neonates and infants, as such individuals cannot effectively communicate the possibility of an injection site complications. Extravasation rates in children appear to be similar to those of the adult population. An extravasation rate of 0.3% was documented in a study of 554 children in which a power injector was used to administer iodinated contrast medium [14]. Most extravasations in the pediatric population resolve without untoward sequelae. A study by Wang et al [15] showed that 15 of the 17 cases of contrast-medium extravasation in children were mild in severity with minimal or no adverse effects.

Physiologic Side Effects in Children

Although most minor physiologic side effects to IV contrast medium administration in adults are of minimal significance, such events are often of increased importance in children. For example, local warmth at the injection site and nausea, generally regarded as physiologic side effects to contrast medium administration, may cause a child to move or cry. Such a response to contrast medium injection may result in the acquisition of a nondiagnostic imaging study, necessitating repeat imaging and additional exposure to contrast medium and radiation. There may be differences between the various nonionic low-osmolality iodinated contrast agents with regard to the incidence of injection-related side effects [16].

Incidence of Allergic-Like Reactions

There are several difficulties in interpreting the available literature on the incidence of allergic-like reactions to IV iodinated contrast media in children. First, many studies have failed to discriminate between physiologic side effects and allergic-like reactions and have used heterogeneous definitions of what constitutes mild, moderate, or severe reactions. Second, there is a lack of controlled prospective pediatric studies on the topic. Prospective investigations are difficult to perform because allergic-like reactions to contrast media in children are rare, and large numbers of patients would be needed to acquire statistically meaningful results. Also, much of the existing literature is retrospective in nature, for which it is difficult to ensure that all adverse reactions are appropriately documented.

It is generally agreed, however, that the incidence of allergic-like reactions in children is lower than that in adults [11, 16, 17]. A very large retrospective study by Katayama et al of more than 100,000 contrast medium administrations [17], when stratified by age and the use of non-ionic iodinated contrast media, showed that patients less than 10 years of age and the elderly have the lowest rates of adverse reactions. A study by Dillman et al. [18] retrospectively reviewed more than 11,000 IV injections of low-osmolality nonionic iodinated contrast media in children and neonates and documented an allergic-like reaction rate of 0.18%. Of the 20 reactions documented in their study, 16 were mild, one was moderate, and three were severe [17]. A similarly performed study by Wang et al. [19] in adult patients from the same institution over a similar time period revealed an adult reaction rate of approximately 0.6%. A study by Callahan et al. [20] of 12,494 consecutive patients up to 21 years of age revealed a 0.46% incidence of adverse reactions to ioversol, the majority of which were mild. A smaller study by Fjelldal et al. [21] documented five allergic-like reactions to iohexol following a total of 547 injections, for a rate of reaction of 0.9%. Although fatal reactions to contrast media in children are extremely rare (and may be due to co-morbid conditions in some cases), infants and young children require close observation during and immediately following IV contrast medium administration, as they are unable to verbalize reaction-related discomfort or symptoms.

Prevention of Allergic-Like Reactions

General guidelines for the prevention of allergic-like reactions in children are similar to those used for adult patients. Allergic-like reactions following premedication may still occur, although the frequency of such reactions is unknown [18]. It should be noted that there has been no prospective, controlled investigation performed to assess the efficacy of premedication for the prevention of allergic-like reactions to iodinated contrast media in children.

Treatment of Allergic-Like Reactions

General guidelines for the treatment of allergic-like reactions in children are similar to those used for adult patients. Pediatric medication dosages are significantly different from adult dosages used in the management of such reactions.

It can be helpful to have a pediatric medication chart with weight-based dosages placed on the emergency cart or posted in the rooms where intravascular contrast media is to be injected into children. Dedicated pediatric emergency resuscitation equipment (including various sizes of supplemental oxygen face masks) also should be available in all such locations. A separate box of pediatric airway equipment attached to the emergency cart may be useful in areas where both children and adults receive contrast media.

Contrast-Induced Nephrotoxicity in Children

There has been no large prospective investigation dealing with the possible nephrotoxic effects of intravascular low-osmolality iodinated contrast agents in children. Consequently, the effects of contrast media on the kidneys are generally assumed to be similar between children and adults. A few key differences are mentioned below.

Measurement of Renal Function in Children

Serum creatinine concentration reflects the balance between creatinine production and excretion. Creatinine is a breakdown product of skeletal muscle, and its rate of production is proportional to muscle mass. Muscle mass depends on a variety of factors, including patient age, gender, and level of physical activity. Normal serum creatinine concentrations, thus, are quite variable in pediatric patients, even in the presence of preserved renal function. It is important to recognize that normal adult creatinine concentrations cannot be applied to the pediatric population. Normal pediatric serum creatinine concentrations increase with age, with the upper limits of normal always less than adult values. Age-based normal serum creatinine concentrations may vary slightly from laboratory to laboratory.

There are problems with using serum creatinine concentration as the sole marker of renal function. First, a normal serum creatinine value does not mean that renal function is preserved. For example, an increase in creatinine from 0.4 mg/dL to 0.8 mg/dL in a 10-year-old patient would be clinically significant and suggest some degree of renal impairment, even though both measurements may be within acceptable limits for patient age. Serum creatinine concentration may not become abnormal until glomerular filtration has decreased substantially. Second, it may take several days in the setting of acute renal failure for serum creatinine concentration to rise. A patient, therefore, may have impaired renal function and a normal serum creatinine concentration. Measurement of blood urea nitrogen (BUN) concentration is a poor indicator of renal function. BUN concentration depends on numerous variables in addition to renal function, including daily dietary protein intake, hepatic function, and patient hydration. A popular manner by which to express renal function in children is the estimated glomerular filtration rate (eGFR). It is important to note that the formula used to calculate pediatric eGFR (see below) is different from that used in adults. eGFR calculation in children requires knowledge of patient serum creatinine concentration and height. In addition, the assay used to measure serum creatinine concentration must be known.

GFR Calculator for Children

There is no perfect manner of estimating the GFR in children. The National Kidney Disease Education Program, an initiative of the National Institutes of Health, provides an online calculator for estimating purposes and has published the following information regarding the estimation of GFR in children. Currently, the best equation for estimating GFR from serum creatinine in children is the Bedside Schwartz equation. This formula is for use with creatinine methods with calibration traceable to isotope dilution mass spectroscopy (IDMS). Using the Original Schwartz equation (which is no longer recommended) with a serum creatinine value from a method with calibration traceable to IDMS will overestimate GFR by 20–40%.

Equation: Bedside Schwartz Equation

$$\mathrm{GFR}\left(mL/\min/1.73\,m^2\right)=\frac{\left(0.41\times\mathrm{height}\right)}{\mathrm{serum\ creatinine}}$$

- Height in cm
- Serum creatinine in mg/dL

Although other methods of estimating GFR exist (such as cystatin C measurement or nuclear medicine GFR study), the Bedside Schwartz equation remains the most readily available and easiest to use in pediatric patients.

Prevention of Contrast-Induced Nephrotoxicity in At-Risk Children

Risk factors for contrast-induced nephrotoxicity (CIN) in children are thought to be similar to those in adults. Unfortunately, there are no established evidence-based

guidelines for the prevention of CIN in children with impaired renal function. As no pediatric-specific measures for the prevention of CIN have been established in the literature, strategies described in adults should be considered when using IV iodinated contrast media in children with renal dysfunction. A noncontrast imaging examination should be performed if the clinical question can be answered without IV iodinated contrast media. In addition, the use of alternative imaging modalities, such as ultrasound and MR (with or without gadolinium-based contrast medium, depending on the exact degree of renal impairment and the clinical question to be answered), should be considered.

Gadolinium-Based IV Contrast Agents

There are only a few published studies that address adverse reactions to gadolinium-based IV contrast media in children. The guidelines for IV use of gadolinium-based contrast agents are generally similar in both the pediatric and adult populations. A few pediatric-specific issues regarding these contrast agents are discussed below.

Osmolality and Viscosity

As with iodinated contrast media, there is a significant range in osmolality and viscosity of gadolinium-based MRI contrast agents. These physical properties, however, potentially are less important when using gadolinium-based contrast agents in children compared to iodinated contrast agents. The much smaller volumes of gadolinium-based contrast agents typically administered to pediatric patients likely result in only minimal fluid shifts. The slower injection flow rates generally used for gadolinium-based contrast agents result in lower injection-related pressures and decreased risk for vessel injury and extravasation.

Allergic-Like Reactions and Other Adverse Events

Though rare, allergic-like reactions to IV gadolinium-based contrast media in children do occur. A study by Dillman et al [22] documented a 0.04% (48 reactions/13,344 injections) allergic-like reaction rate for these contrast agents in children. A more recent study by Davenport et al that included 15,706 administrations of gadolinium-based contrast media in children (under the age of 18 years) documented only eight allergic-like reactions, for a reaction rate of 0.05% [23]. Although mild reactions are most common, more significant reactions that require urgent medical management may also occur [23]. Pediatric allergic-like reactions to gadolinium-based contrast media are treated similarly to those reactions to iodinated contrast agents.

While no investigation has studied the efficacy of corticosteroid and antihistamine premedication regimens for the prevention of allergic-like reactions to gadolinium-based contrast agents in children or adults, regimens, such as those presented in Table A below, are thought to provide some protective benefit.

A variety of physiologic side effects may also occur following administration of gadolinium-based contrast media, including coldness at the injection site, nausea, headache, and dizziness. There is no evidence for pediatric renal toxicity from gadolinium-based contrast media at approved doses. Extravasation of gadolinium-based contrast media is usually of minimal clinical significance because of the small volumes injected.

Nephrogenic Systemic Fibrosis and Asymptomatic Gadolinium Deposition in Tissues

There are only a small number of reported cases of nephrogenic systemic fibrosis (NSF) in children. As of September 2012, there were 23 unique pediatric NSF cases, and all patients were 6 years of age or older [23]. Seventeen of these children had documented exposure to gadolinium-based contrast material. Thirteen of 13 children with available clinical data pertaining to renal disease had substantial renal dysfunction (acute kidney injury and/or chronic kidney disease), and 10 were on hemodialysis or peritoneal dialysis (or both). In 10 children, renal status was unknown. A few early cases were described prior to this condition's known apparent association with gadolinium-based contrast media (24–30). Only 10 children (all older than 8 years of age) with biopsy-confirmed NSF have been reported to the Yale Registry, with no new cases reported between 2007 and 2013 after guidelines were published in 2007 limiting the use of gadolinium-based contrast media in children with impaired renal function [24].

As there are no evidence-based guidelines for the prevention of NSF in children in particular, adult guidelines are to be followed for identifying at-risk patients and administering gadolinium-based contrast media in the presence of impaired renal function [25]. Children at risk for renal impairment should be identified (chronic kidney disease or acute kidney injury) and screened for impaired renal function. As in adults, gadolinium-based contrast media should be avoided in the setting of acute kidney injury or chronic kidney disease with an eGFR.

High-osmolality iodinated contrast agents should be avoided in children who are at risk for aspiration. Aspirated hyper osmolality contrast medium may cause fluid shifts at the alveolar level and chemical pneumonitis with resultant pulmonary edema (33–36). Aspiration of large volumes of both barium-based and iodinated oral contrast agents rarely may be fatal [26].

Gadolinium deposition has been observed in pediatric patients [27]. The gadolinium deposition is seen in patients with normal renal function. Multiple studies have confirmed increased signal intensity in the dentate nucleus and globus

pallidus on unenhanced T1-weighted image that was correlated with previous exposure to GBCAs (Gadolinium-based contrast agents). It was also confirmed that the hyperintensity noted in brain nuclei was caused by gadolinium deposition on autopsy studies [27]. Similar to NSF, gadolinium deposition is dose-dependent and increases with number of doses. Also, the hyperintensity of brain nuclei was predominantly seen with linear agents though some degree of gadolinium deposition in brain was seen with all agents on pathology. The chemical form deposited and the mechanism of deposition is still unknown. Since the deposition is predominantly seen with linear agents that have less stability, de-chelation is proposed as one of the predisposing factors for gadolinium deposition in tissues.

Gadolinium deposition has also been noted in other tissues including skin and bones. However, these depositions including in the brain, have not shown any clinical effects and are largely asymptomatic [27].

Premedication Dosing Guidelines

Corticosteroid

Dosage

Prednisone 0.5–0.7 mg/kg PO

(Up to 50 mg)

Timing

13, 7, and 1 hr prior to contrast injection

AND Antihistmine

Diphenhydramine 1.25 mg/kg PO

(Up to 50 mg)

Timing

1 hr prior to contrast injection

OR

Methylprednisolone 1 mg/kg PO or IV at 12 h and 2 h before contrast injection (maximum: 32 mg/dose)

AND

Antihistamine

Diphenhydramine 1–2 mg/kg IV or PO (maximum dosage 50 mg/dose, 300 mg/day), I hour before contrast injection

Head and Neck Pathologies (Table 2.1)

In pediatric population, congenital and inflammatory pathologies constitute 80% of the head and neck pathologies. The ten subsets of pathologies are discussed below.

Table 2.1 Ten subsets of pediatric head and neck pathologies

	Pathologies	Examples
1	Normal variations	Mineralization patterns of bones at different ages
2	Emergencies	Ludwig's angina, Lemierre syndrome
3	Epithelial inclusion cysts	Dermoid, epidermoid, teratoma
4	Cephaloceles	Frontoethmoidal encephalocele, nasal glial heterotopia
5	Vascular anomalies	Hemangioma, venolymphatic malformation and venous malformations
6	Branchial cleft cysts	Type I, Type II, Type III, and Type IV cysts
7	Trauma	Dissection, pseudoaneurysms
8	Benign neoplasms	Juvenile nasopharyngeal angiofibroma, pilomatrixoma, nerve sheath tumors
9	Malignant neoplasms	Rhabdomyosarcoma, lymphoma, leukemia, Retinoblastoma, papillary thyroid cancer, & metastases
10	"Touch me not" lesions	Fibromatosis colli, ectopic thymus

Normal Variations

It is important to understand the appearance of the developing anterior skull base to avoid interpretive errors in this complex region. Studies demonstrated that the ethmoidal labyrinth and turbinates (derived from the primitive nasal capsule) serve as the nidus for ossification of the anterior skull base [28]. Ossification continues from this area toward the midline over the first few months of life, accounting for the midline gaps normally seen on coronal CT scans. As the child matures, ossification of the anterior skull base proceeds in a fairly constant manner, with fusion of the cribriform plate and lateral ethmoid bone masses beginning as early as 2 months of age (Fig. 2.2). Similarly, nonossified portion of C2 vertebra may appear as focal hypodensity in infants often causing errors in interpretation. Arrested pneumatization of sphenoid sinus is another normal variant, which mimics a lesion (Fig. 2.3). These findings are eventually stable in the follow-up imaging.

Non-Traumatic Emergencies

Computed tomography (CT) is the first-line imaging modality in the acute setting and subsequently, magnetic resonance (MR) imaging for further assessment. Awareness of these conditions is important not only to provide an accurate diagnosis but also to assess the extent of disease, evaluate for potential complications, and recommend definitive subspecialty evaluation. These can occur in the oral cavity (Ludwig angina), oropharynx (peritonsillar abscess), retropharynx (retropharyngeal abscess), hypopharynx (epiglottitis), salivary gland (sialadenitis, parotiditis), spine (discitis, septic facet arthritis), vascular space (Lemierre syndrome), orbits and sinuses (cellulitis, dacryocystitis, and invasive sinusitis) [29].

Ludwig angina is a serious life-threatening infection of the floor of the mouth that rapidly spreads bilaterally to the soft tissues of the oral cavity [29]. Ludwig angina is a type of cellulitis and not a focal abscess. It is caused by an infection

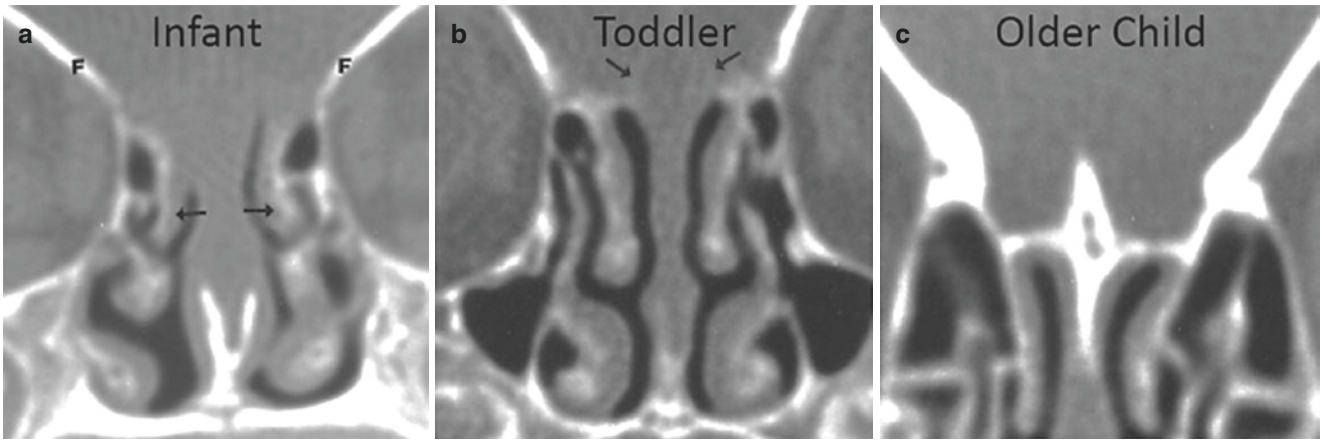

Fig. 2.2 CT of the anterior skull base showing progress of mineralization in infants, toddlers, and older children (**a**, **b**, and **c**). (**a**) In an infant, there is no ossification of the anterior skull base between the orbital plates of the frontal bones (F), although the turbinates are ossified (arrows). (**b**) In a toddler, ossification begins in the roof of the ethmoidal labyrinth laterally and spreads toward the midline (arrows). (**c**) In an older child, the entire anterior skull base is well ossified

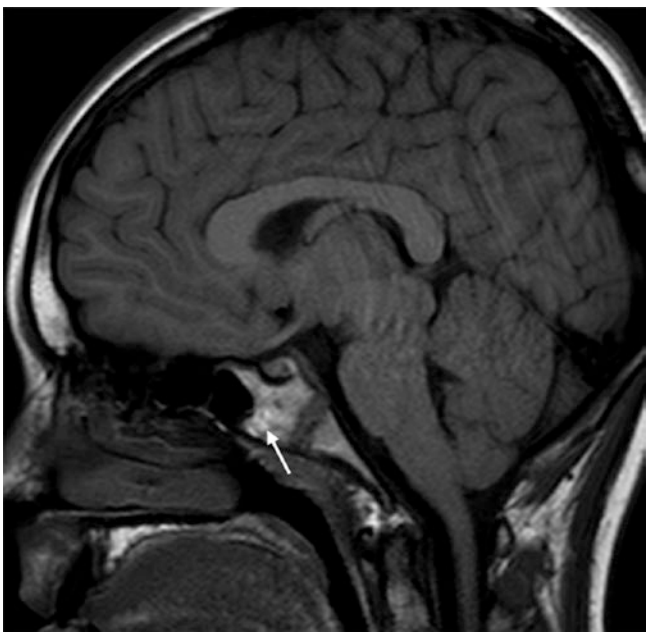

Fig. 2.3 Arrested pneumatization of sphenoid sinus. Sagittal T1 W image showing a focal hyperintense area within the left sphenoid sinus (arrow) that is due to arrested pneumatization of sphenoid sinus

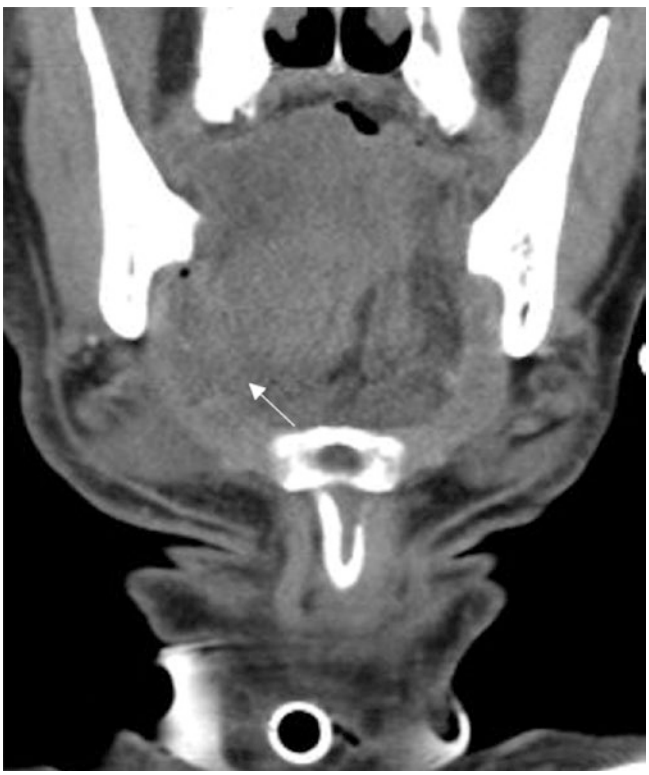

Fig. 2.4 Ludwig's angina. Coronal CT neck showing edema in the right submandibular space and base of tongue with adjacent subcutaneous fat stranding

of the third mandibular molar tooth or pericoronitis (an infection of the gums surrounding the partially erupted lower third molar tooth), both of which are caused by Streptococcus organisms. As the soft-tissue swelling extends in Ludwig's angina, it displaces the tongue into the pharyngeal airway and causes difficulty in breathing (Fig. 2.4). Treatment includes airway management and antibiotics. Imaging is performed to evaluate airway patency and determine if gas-forming organisms, assess presence of underlying dental infection, or a drainable abscess.

Lemierre syndrome is a rare and potentially life-threatening complication of acute respiratory tract infection. It can occur in healthy adolescents and young adults. In Lemierre syndrome, there is presence of septic thrombophlebitis of the internal jugular vein and disseminated abscesses, as well as septic pulmonary emboli. The causative organism

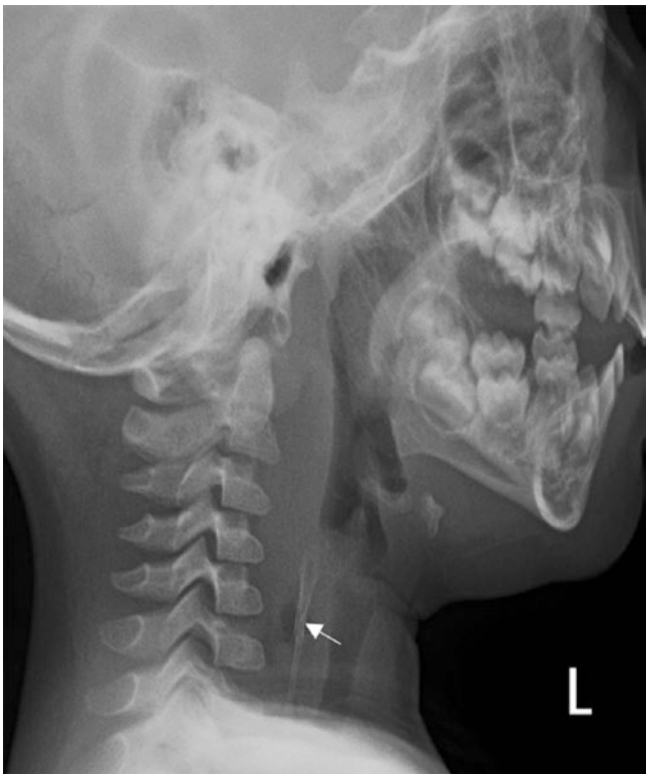

Fig. 2.5 Retropharyngeal abscess secondary to foreign body ingestion. Lateral cervical spine radiograph demonstrating prevertebral soft tissue swelling and dense linear foreign body which is a fish bone (arrow)

is *Fusobacterium necrophorum*, an anaerobe present in normal oropharyngeal flora [29].

An ingested foreign body could be an important cause of retropharyngeal abscess in children (Fig. 2.5).

Epithelial Inclusion Cysts

Epithelial cysts are benign lesions which are histologically characterized by cystic spaces lined by simple squamous epithelium (epidermoid cyst) (Figs. 2.6 and 2.7), which contain skin adnexa ("true" dermoid cyst) (Figs. 2.8 and 2.9) or tissues of all three germ layers, like muscle, teeth, bone, cartilage, etc. (teratoid cyst) (Fig. 2.10). They commonly occur at areas of embryonic fusion and constitute 1.6–6.9% of all cysts in the head–neck region [30]. They may also result from abnormal invagination of surface ectoderm along the embryologic sites of dermal fusion that form the eyes, ears, and face. Computed tomography (CT) scan would demonstrate a unilocular cyst with homogeneous, hypo-attenuating (0–18 HU) fluid or fat material that has multiple hypo-attenuating fat density nodules giving a "sack of marbles" appearance; this imaging appearance is pathognomonic for a dermoid cyst. Magnetic resonance imaging (MRI) shows fluid signal due to high protein content, and areas of fat component will show low signal on fat suppressed images. MRI helps in visualization

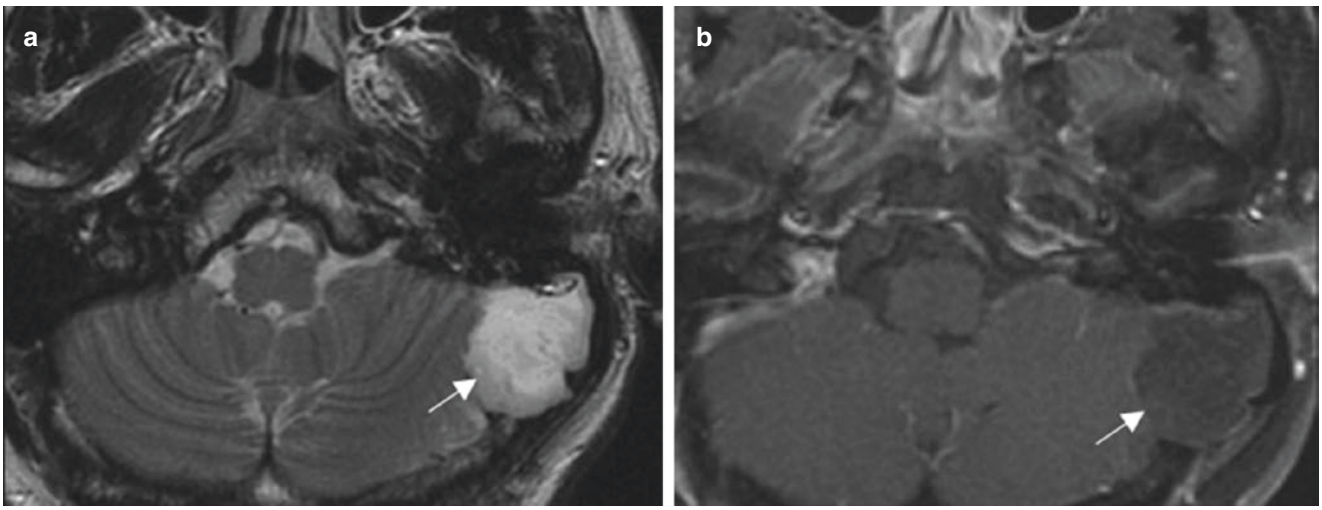

Fig. 2.6 Epidermoid. T2 (**a**), postcontrast T1 W (**b**), FLAIR (**c**), and diffusion weighted (**d**) MRI images showing a nonenhancing cystic lesion (arrow) located in the left mastoid that shows restriction on diffusion image (**d**)

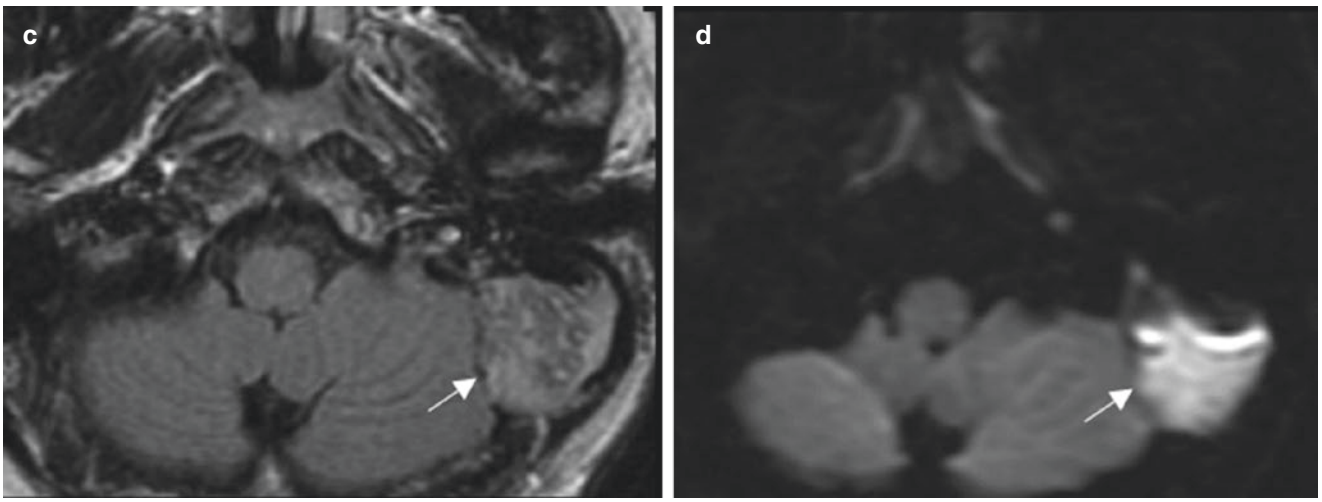

Fig. 2.6 (continued)

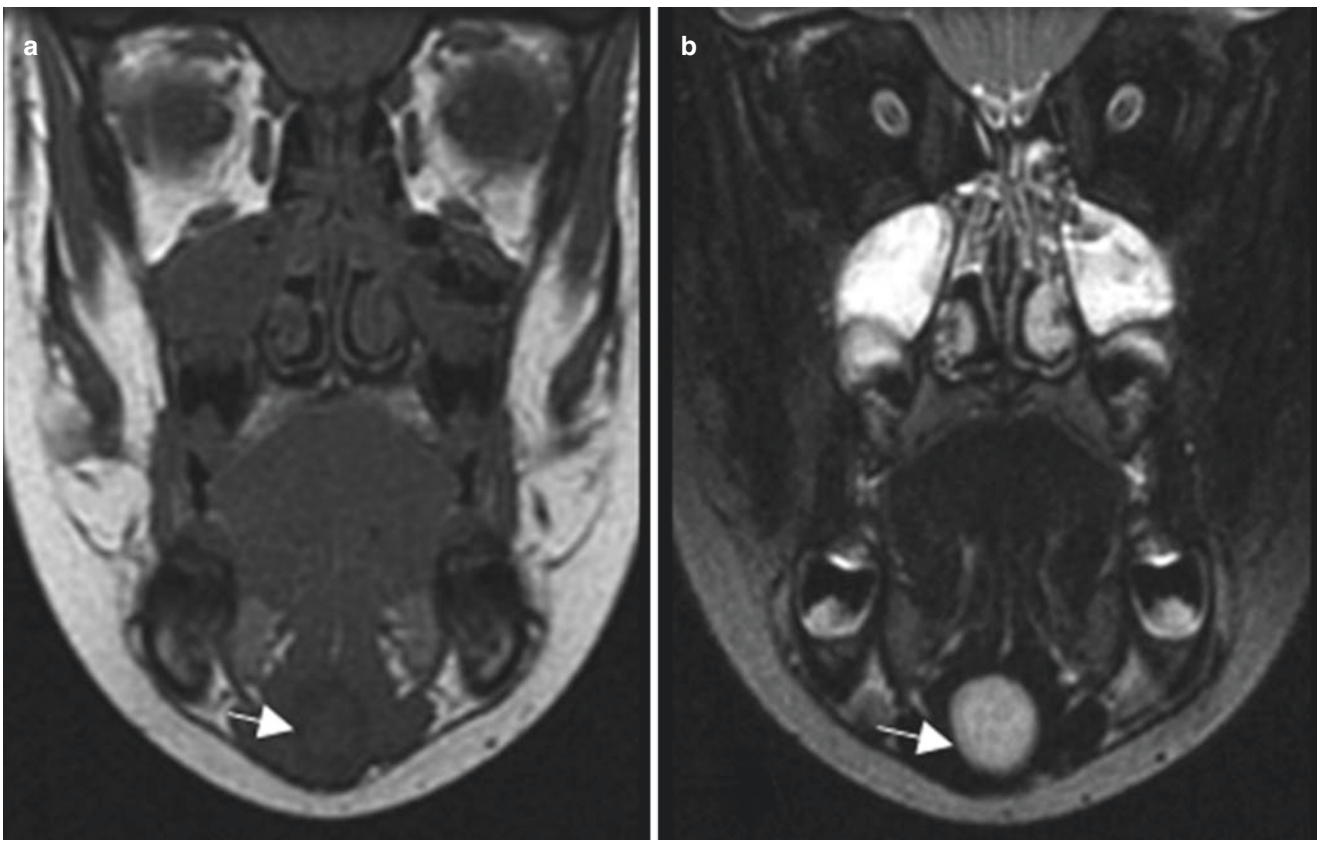

Fig. 2.7 Epidermoid. Axial T1, T2 W MR images (**a** and **b**) showing a cystic lesion in the midline of the floor of the oral cavity (arrow)

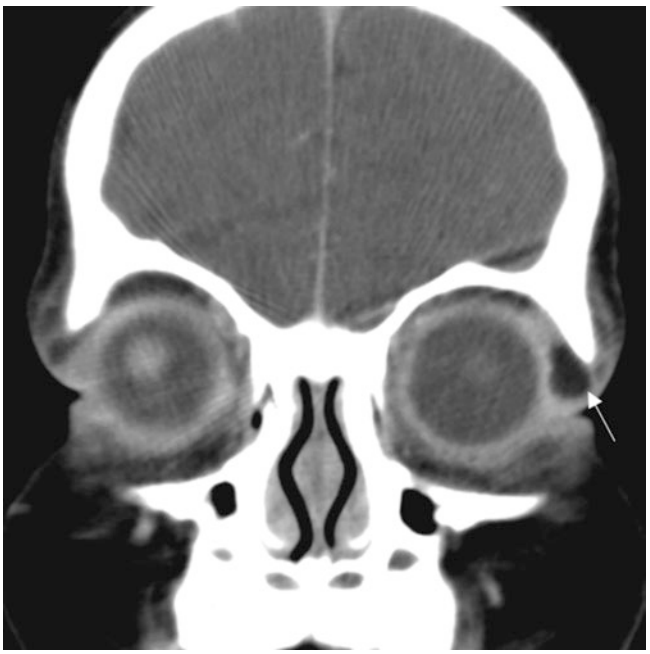

Fig. 2.8 Dermoid. Coronal CT showing hypodense lesion of fat attenuation involving left fronto zygomatic suture (arrow)

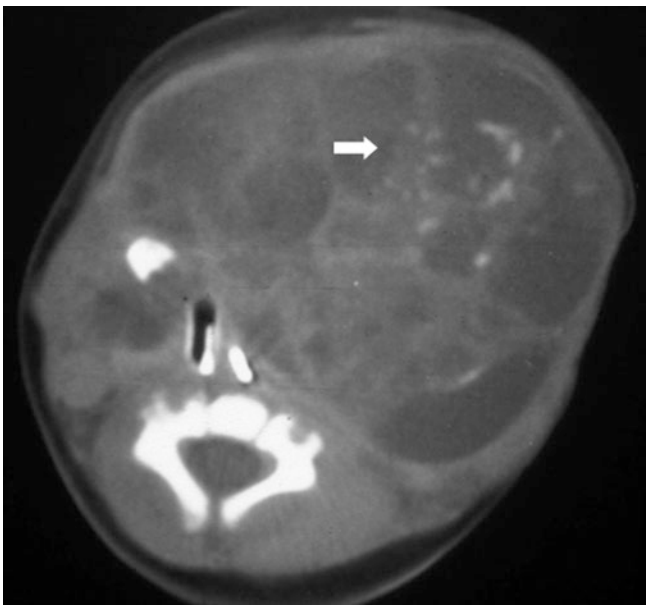

Fig. 2.10 Teratoma in the neck. CT neck showing a large lesion with foci of fat and calcification (arrow)

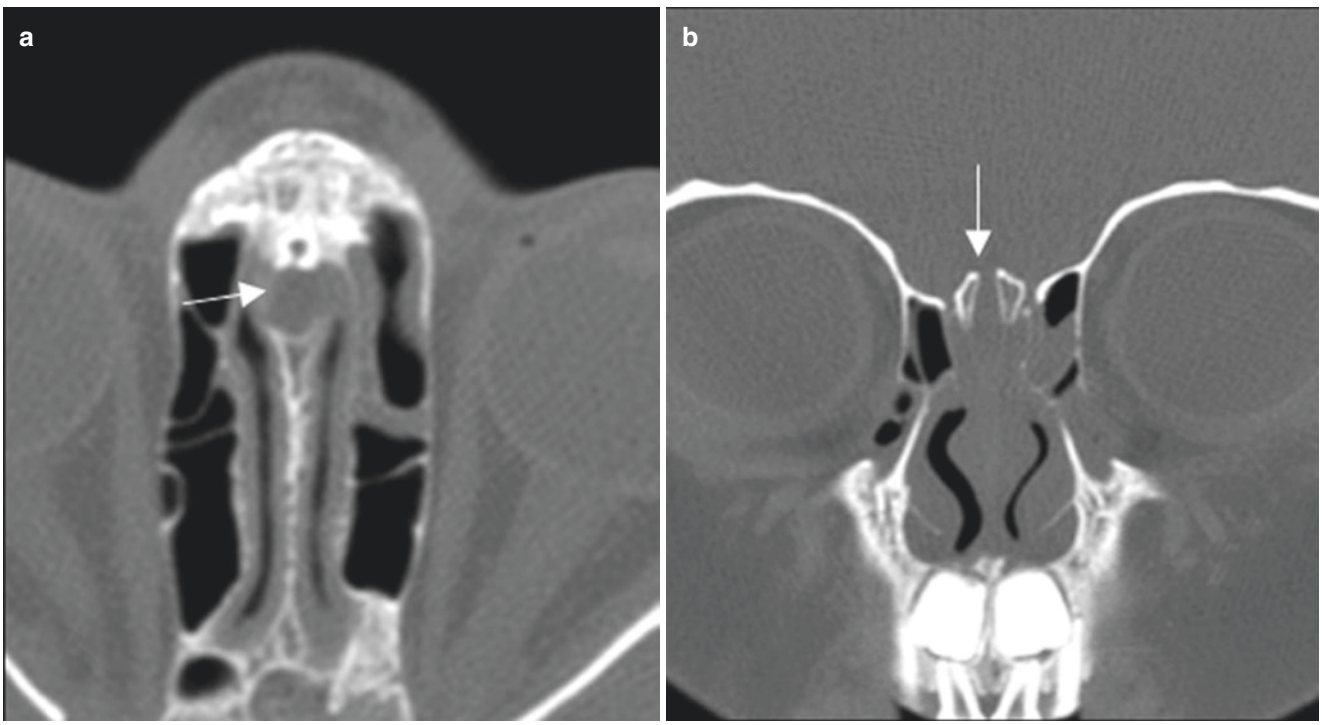

Fig. 2.9 Nasal dermoid. Axial and coronal CT images (**a** and **b**) at the level of anterior cranial fossa showing widened foramen caecum (horizontal arrow) with bifid crista galli (vertical arrow)

of the exact location and extent of cystic lesions in the floor of the mouth and is useful for determining their relationship to the surrounding muscles. These cysts carry relatively higher risk of recurrence as compared to lipomas and certain other benign lesions that mimic these cysts.

Cephaloceles

Intracranial tissue may herniate through a defect in the cranium that leads to an encephalocele. They occur in one of every 4000 live births and are most often occipital in location

(75%). Lesions are frontoethmoidal in 15% of cases and basal in 10%. There are often significant associated intracranial anomalies. Occipital encephaloceles may be associated with Chiari or Dandy Walker malformations and callosal or migrational anomalies [31].

Frontoethmoidal lesions are not typically associated with these types of anomalies. Frontoethmoidal encephaloceles are also known as sincipital encephaloceles and are further divided into nasofrontal, nasoethmoidal, and naso-orbital types. These are more common in South and Southeast Asian populations [31]. They track along the nasal bridge between the nasofrontal sutures into the glabella (nasofrontal region) (Fig. 2.11), under the nasal bones and above the nasal septum (nasoethmoidal region), or along the medial side of the orbit at the level of the frontal process of the maxilla and the ethmoid-lacrimal bone junction (naso-orbital region). Frontoethmoidal encephaloceles (Fig. 2.12) present as a clinically visible mass along the nose. The intracranial root of most frontoethmoidal encephaloceles is located at the foramen cecum, which is a small ostium located at the bottom of a small depression anterior to the crista galli and is formed by the closure of the frontal and ethmoid bones.

Basal encephaloceles are internal and do not manifest externally, although they may present as a lump or bump in the oropharynx or nasopharynx. Basal encephaloceles include transethmoidal, sphenoethmoidal, transsphenoidal, and frontosphenoidal types [31]. Trans sphenoidal and transethmoidal encephaloceles are the common varieties. In the former, there is a defect in the floor of the sella and into the nasal cavity, and in the latter, they present through a midline or cribriform plate defect into the nasal cavity. Transsphenoidal encephaloceles may be associated with a cleft palate and could also project into the oral cavity. Affected children may present with nasal obstruction. Surgery is the treatment for encephaloceles, and MR imaging is the imaging modality of choice for identifying the contents of an encephalocele prior to surgery. High-resolution CT may also be used to display the bone anatomy, but the intracranial connection is best depicted with MR imaging. Occipital encephaloceles commonly involve the cerebellar or cerebral hemispheres with involvement of the dural venous sinuses. MR venography is used to demonstrate venous involvement in these lesions.

Nasal glial heterotopia (nasal glioma) occur near the root of the nose (where the cranial portion of the nose joins the forehead). These are composed of dysplastic glial tissue and are basically congenital nonneoplastic lesions best categorized as heterotopia (Fig. 2.13). Nasal gliomas are intranasal in 30% of cases, extra nasal in 60%, and mixed in 10% [31].

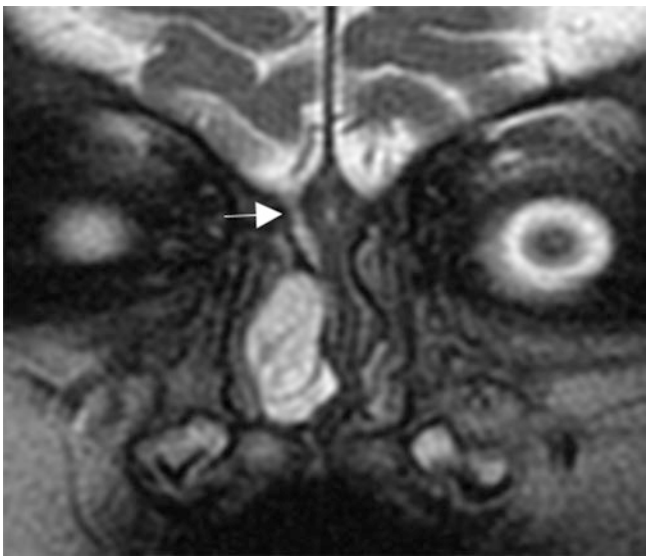

Fig. 2.12 Nasal glial heterotopia (nasal glioma). Coronal T2W MRI showing a linear hyperintense stalk connecting the right nasal mass to the intracranaial cavity (arrow)

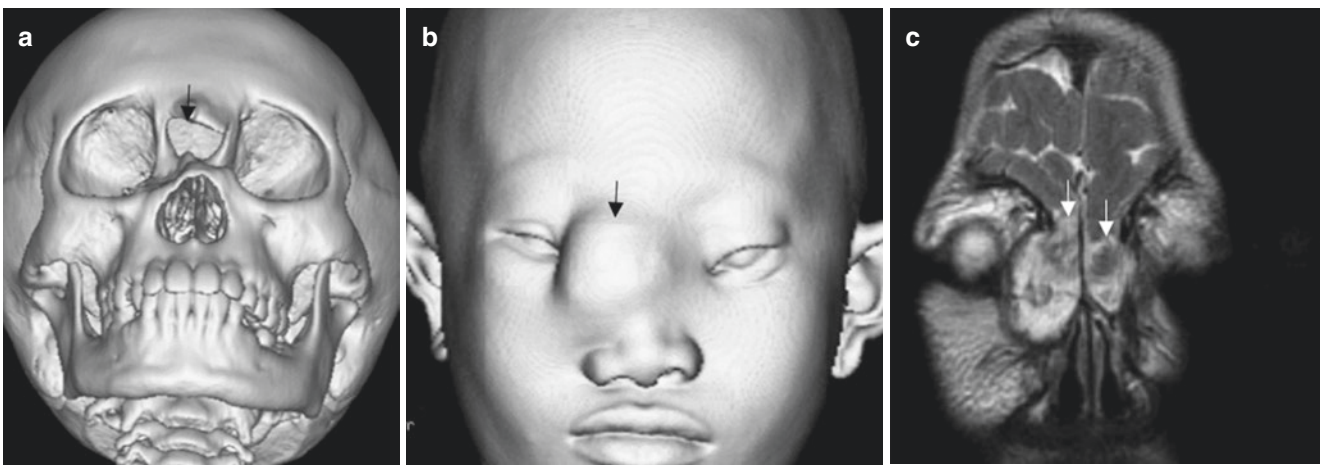

Fig. 2.11 Midline frontoethmoidal cephalocele. 3D CT reconstructions (**a**, **b**) showing a midline defect (arrow) and a nasal mass (arrow). Coronal T2 W MRI (**c**) showing brain and CSF herniating through the cribriform plate (arrows)

They are treated by surgical resection. MRI is the imaging of choice and they appear isointense relative to normal brain.

Vascular Anomalies

Vascular anomalies of the head and neck region constitute approximately 60% of vascular anomalies diagnosed in children and affect approximately 1 in 22 children [32]. These lesions broadly fall into two categoriesvascular tumors and vascular malformationsand both have physical and psychological implications for both the patient and their family, particularly if visibly disfiguring.

Imaging with ultrasound (US) and/or magnetic resonance imaging (MRI) offers exquisite soft tissue detail with delineation of blood vessel architecture and flow patterns. For a diverse group of lesions presenting in complex anatomical locations, a multidisciplinary approach is essential with representation from specialists in pediatrics, dermatology, plastic surgery, orthopedic, and craniofacial surgery, otorhinolaryngology, oncology, neurosurgery, interventional radiology, and neuro-radiology, as well as supportive input from psychology, physiotherapy, and occupational therapy when needed. Treatment strategies for vascular malformations include conservative management, drug treatment, minimally invasive interventions by interventional radiology, laser therapy and open surgery. A combination of these therapies is often required [32].

Hemangiomas and vascular malformations are endothelial disorders divided into two pathological groups: vascular tumors (including hemangiomas) and vascular malformations as mentioned below. They were initially categorized in this way by Mulliken and Glowacki in 1982, based on the natural history, histology, and cellular activity of these lesions [33]. Currently, the ISSVA classification is used [34].

Vascular tumors are composed of rapidly proliferating cells and incomplete blood vessels. Infantile hemangioma (IH) (Fig. 2.14) constitutes to the large majority of the lesions in the vascular tumors category. There is spontaneous involution noted in these lesions (Fig. 2.15).

Vascular malformations on the other hand are present at birth and grow proportionately with the child. They are composed of dysplastic arterial, venous, and/or lymphatic vessels. Unlike IH, spontaneous involution does not occur. They are further categorized according to the predominant vessel type and are further classified as "high-flow" and "low-flow" lesions, as mentioned below. Lesions that demonstrate arteriovenous shunting such as arteriovenous malformations (AVMs) and arteriovenous fistulae (AVF) are described as high flow, whereas venous malformations (VMs) (Fig. 2.16), lymphatic malformations (LMs), or combined lympho-venous/veno-lymphatic malformations (VLMs) (Fig. 2.17), together with CMs are labeled as low flow lesions.

ISSVA classification [34] for vascular anomalies

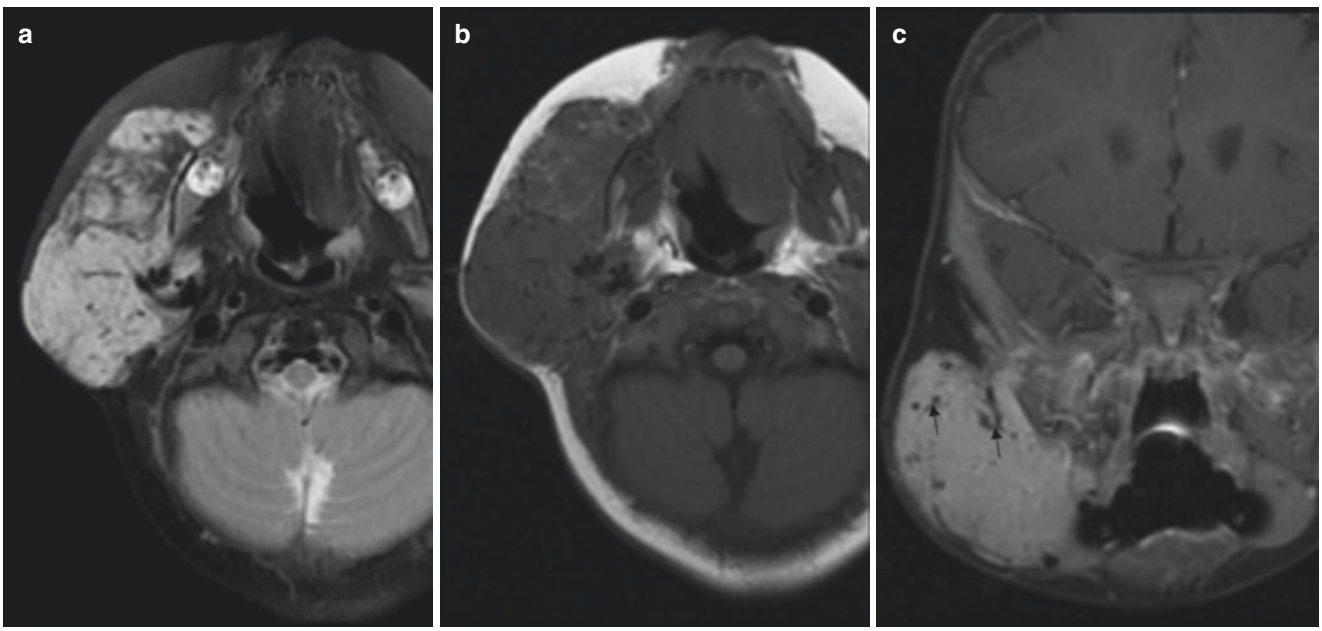

Fig. 2.13 Infantile hemangioma. Axial T2, T1, and postcontrast T1 W images (**a**, **b**, and **c**) showing an intensely enhancing mass in the right parotid space with flow voids (arrows)

Vascular tumors	Vascular malformations			
Benign • Infantile hemangioma/Hemangioma of infancy • Congenital hemangioma • Rapidly involuting (RICH) • Noninvoluting (NICH) • Partially involuting (PICH) • Tufted angioma • Spindle-cell hemangioma • Epithelioid hemangioma • Pyogenic granuloma (also known as lobular capillary hemangioma)	Simple	Combined	Of major named vessels	Associated with other anomalies
Locally aggressive • Kaposiform hemangioendothelioma • Retiform hemangioendothelioma • Papillary intralymphatic angioendothelioma (PILA), Dabska tumor • Composite hemangioendothelioma • Pseudomyogenic Hemangioendothelioma • Polymorphous hemangioendothelioma • Hemangioendothelioma not otherwise specified • Kaposi sarcoma	• Capillary malformations • Lymphatic malformations • Venous malformations • Arteriovenous malformations • Arteriovenous fistula	Capillary-venous malformation, capillary lymphatic malformation, capillary-arteriovenous malformation, lymphatic-venous malformation, capillary-lymphatic-venous malformation, capillary-lymphatic-arteriovenous malformation, capillary-venous-arteriovenous malformation	Affect: Lymphatics, veins, arteries Anomalies of: • Origin • Course • Number • Length • Diameter (aplasia, hypoplasia, stenosis, ectasia/aneurysm) • Valves • Communication (AVF) • Persistence (of embryonal vessel)	Klippel-Trenaunay syndrome, Parkes Weber syndrome, Servelle–Martorell syndrome, Sturge–Weber syndrome, Maffucci syndrome, CLOVES syndrome, Proteus syndrome, CLAPO syndrome
Malignant • Angiosarcoma • Epithelioid hemangioendothelioma				

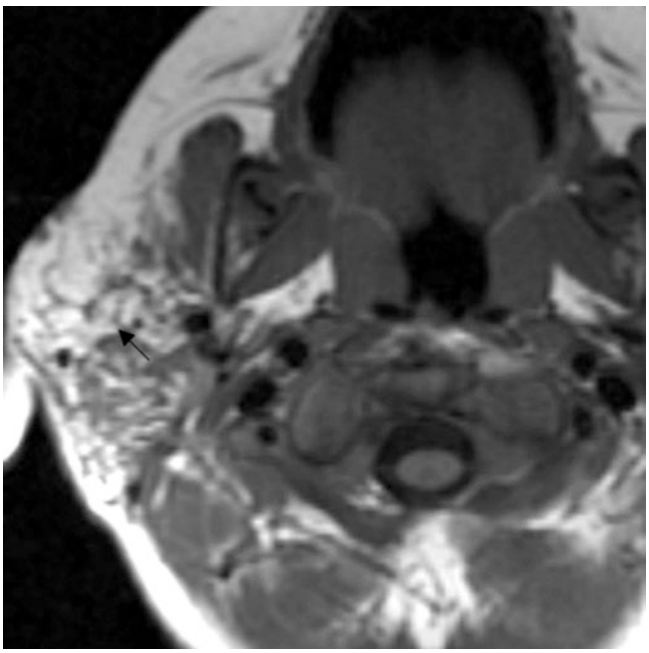

Fig. 2.14 Infantile hemangioma. Axial T1 W MRI showing fatty replacement of stroma within the lesion (arrow) located in right parotid space

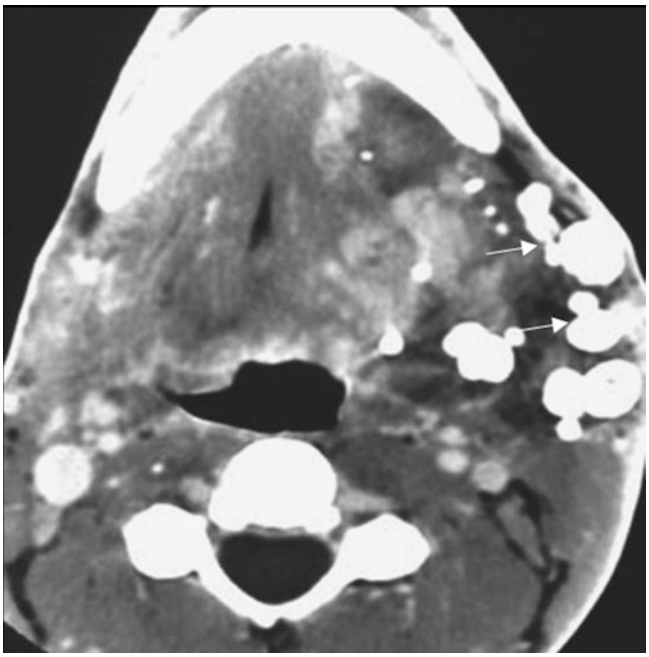

Fig. 2.15 Cervico facial venous malformation. Contrast-enhanced CT showing an enhancing mass on the left side of the neck with phleboliths (arrows) within the lesion

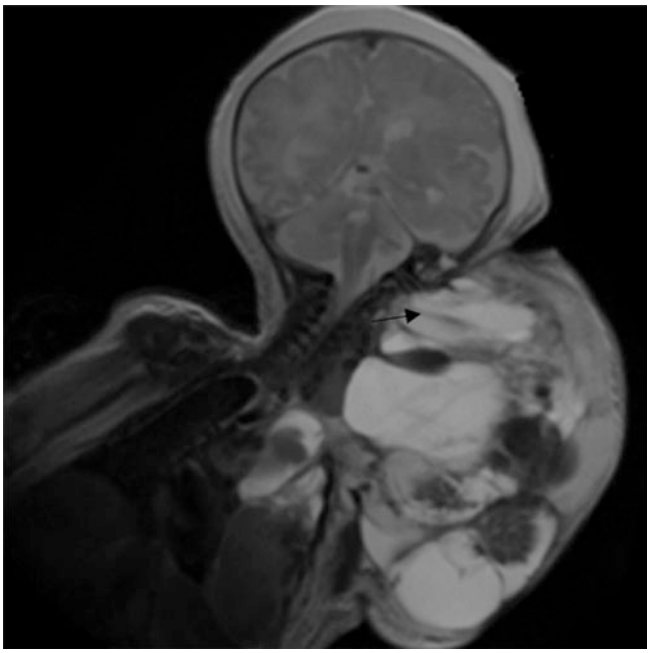

Fig. 2.16 Craniofacial venolymphatic malformation. Coronal T2W MRI showing a large lesion with fluid-levels (arrow) involving the left side of the face and neck in a neonate

Branchial Cleft Cysts

Branchial anomalies are the commonest embryological anomalies of the head and neck in pediatric population. They can present in the form of sinuses, fistulae, and cysts. The clinical history and symptoms help in raising a suspicion of branchial cleft cysts. The commonly used imaging modalities are computed tomography, magnetic resonance imaging, ultrasonography, and fine-needle aspiration [35]. The mainstay of management is usually by surgical excision. They are subdivided depending on their embryological origin

First Branchial Cleft Cysts [35]

Their occurrence is uncommon and comprise only 7% of all recorded cases of branchial cysts. On examination, it appears as a round or oval cystic mass, which can be located either within, superficial, or deep to the parotid gland or along the external auditory canal. It is a possible differential for cystic lesions observed in the parotid or peri-parotid regions.

These cysts are further classified as:

- *Type 1*: These are located close to the external auditory meatus. In a large number of cases, they are present infe-

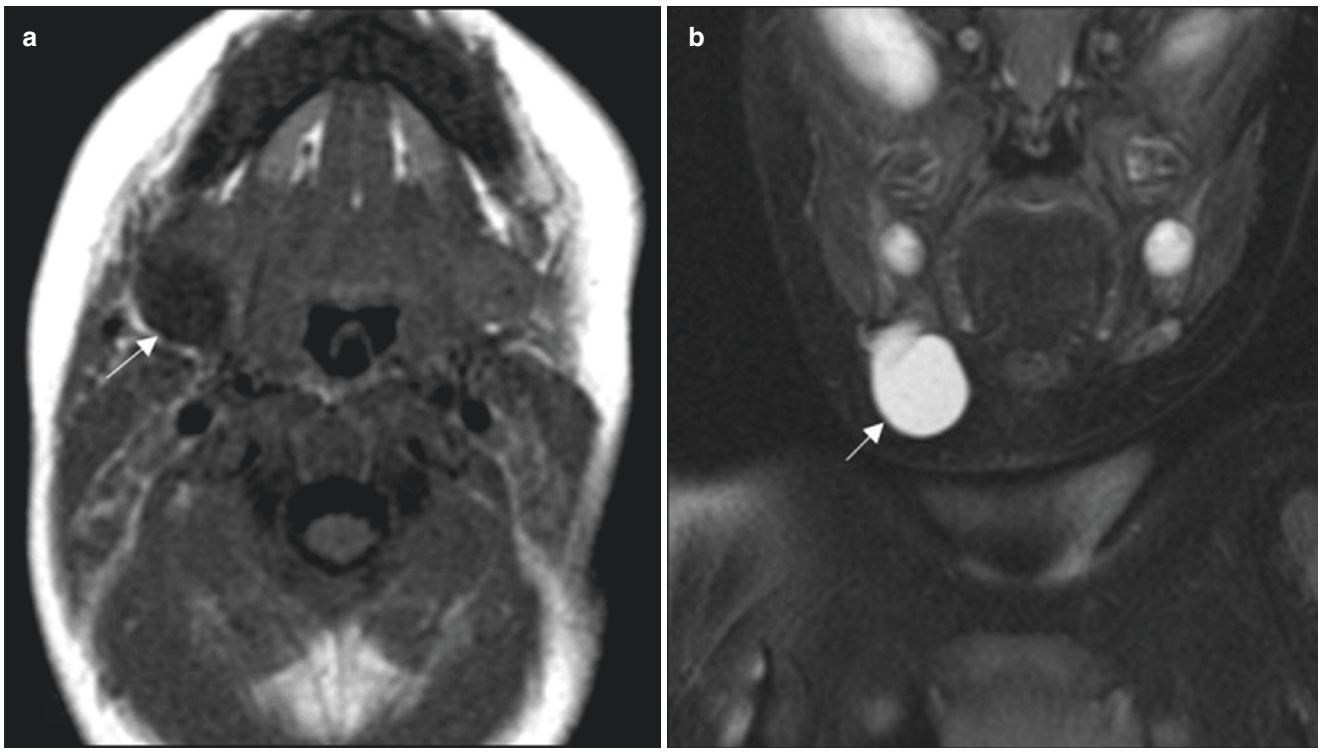

Fig. 2.17 Type 2 branchial cleft cyst. Axial T1, coronal T2 W MR images (**a** and **b**) showing a cystic lesion (arrow) deep to the sternocleidomastoid and lateral to the carotid space

rior and posterior to the tragus. However, they can also be present in the parotid gland or at the angle of the mandible.
- *Type 2*: These correspond to the submandibular gland and are found in the anterior triangle of the neck. They open into the external auditory canal.

Treatment is by surgery and surgical method is unique for every case of this anomaly in keeping with the integrity and patency of the tract for complete excision.

Second Branchial Cleft Cysts [35]

This subtype of branchial cleft cysts makes up approximately 95% of all branchial anomalies.

These are further classified into four subtypes, as follows:

- *Type 1*: Situated anterior to the sternocleidomastoid, just deep to the platysma.
- *Type 2*: The most common variant of the four subtypes, found deep to the sternocleidomastoid, lateral to the carotid space (Fig. 2.18).
- *Type 3*: These extend medially between the bifurcation of internal and external carotid arteries up to the lateral pharyngeal wall.
- *Type 4*: Located in the pharyngeal mucosal space, medial to the carotid sheath.

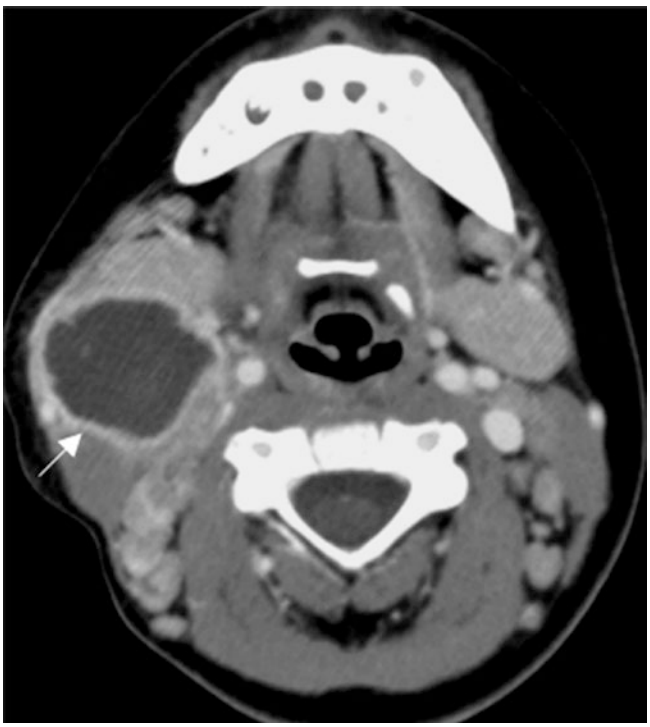

Fig. 2.18 Infected Type 2 branchial cleft cyst. Postcontrast CT shows rim enhancing cystic lesion (arrow) on the right side, lateral to carotid space

In the case of fistula or sinus, the internal opening may be found in the region of the palatine tonsillar fossa. The external opening here is along the intersection of the middle and distal portions of the anterior border of the sternocleidomastoid.

There is a possibility of secondary infection (Fig. 2.19); hence, the preferred management strategy is surgical excision. It is normally delayed until the age of two to three years. In cases of isolated type 4 second branchial cleft cyst, an intraoral approach can be considered.

Third Branchial Cleft Cysts [35]

Third branchial cleft cysts are rare in occurrence. However, they have been found to be the second most common congenital lesions of the posterior cervical area after thyroglossal cysts. They are often located deep to the sternocleidomastoid. They are found to be more commonly situated on the left side. Management is by surgical excision. Visualization of the pyriform sinus is necessary before surgery, and the preferred approach is along the sternocleidomastoid muscle.

Fourth Branchial Cleft Cysts [33]

Fourth branchial cleft cysts are very rarely prevalent and comprise 1–4% of all branchial cleft anomalies. These are common on the left side and are situated in the thyroid gland and mediastinum. These occur in early childhood, frequently after a recurrent abscess or a preceding thyroiditis (which can be superimposed, acute, and suppurative). Due to their rare occurrence, there are no precise established procedures outlined for their management. The definitive treatment is surgical excision combined with partial thyroidectomy.

Trauma

Carotid artery dissection (CAD) and vertebral artery dissection (VAD) are uncommon but in children, traumatic dissections that appear to be more common than nontraumatic dissections. The incidence of traumatic dissection for carotid and vertebral artery is 0.08–0.4% of the entire trauma population [36]. These are underdiagnosed because of the lack of early warning symptoms, and traumatic pseudoaneurysms may be misdiagnosed as saccular aneurysms, or vasospasm following subarachnoid hemorrhage.

CAD can lead to thrombosis and occlusion of the vessel [36] and is one of the major causes of stroke in children. The most common mechanism in traumatic CADs is direct blows to the neck/head or hyperextension. Motor vehicle accidents, sporting events, fights, and falls can cause arterial dissections. Although angiogram continues to be the gold standard for diagnosis, CTA is equally popular, as their specificity and sensitivity is approaching that of cerebral angiograms (Fig. 2.20). Because of its invasive nature and the radiation

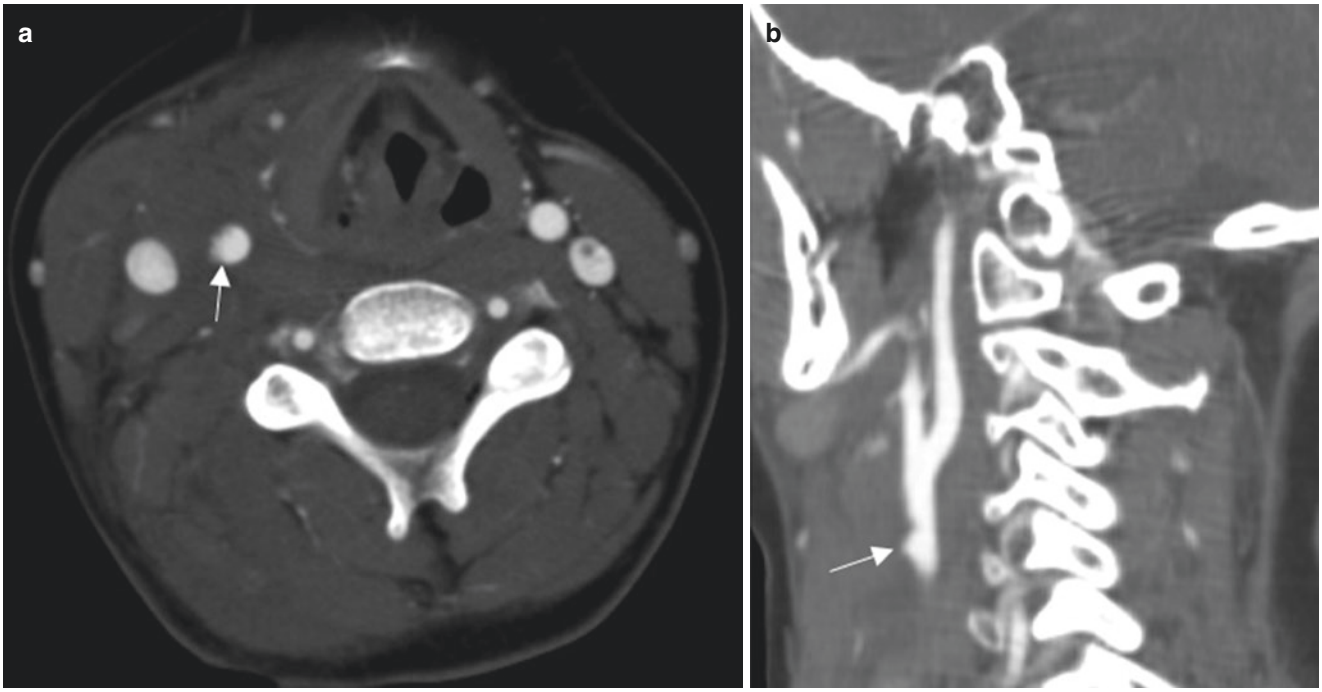

Fig. 2.19 Pseudoaneurysm of right ICA secondary to trauma in a 4-year-old boy who was stabbed with a pencil in the right side of the neck. Axial and sagittal CT angiography (**a** and **b**) showing mild irregularity of the right ICA (arrow) with a small pseudoaneurysm in the same location on sagittal CTA image (arrow)

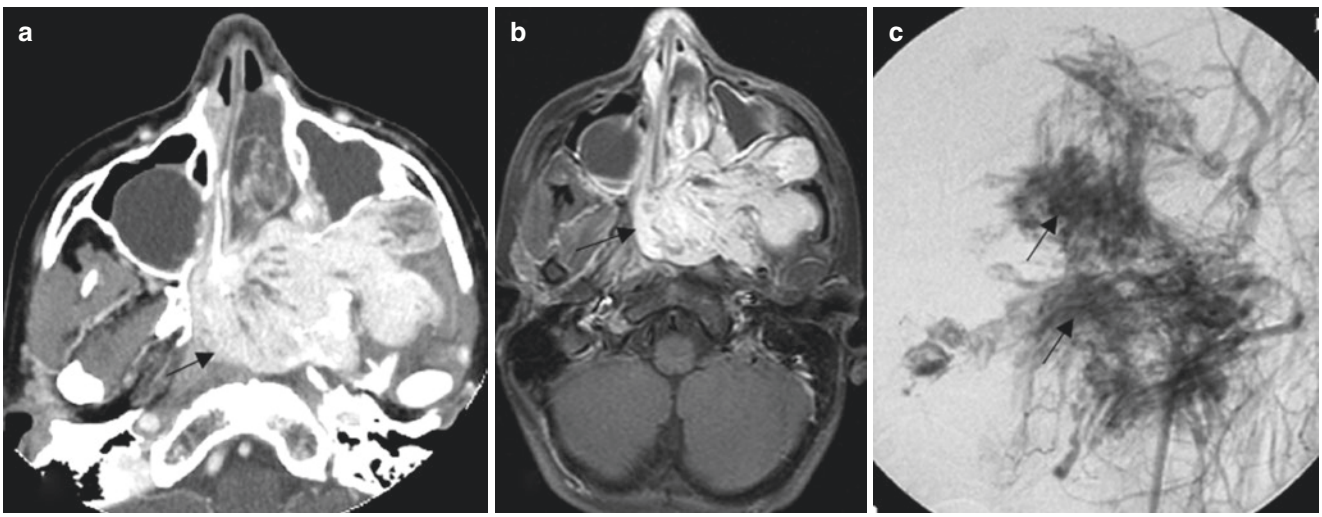

Fig. 2.20 Juvenile nasopharyngeal angiofibroma (JNA). Axial contrast-enhanced CT (**a**), axial contrast-enhanced MRI (**b**), and catheter angiography (**c**) show an avidly enhancing lesion (arrow) in the left pterygopalatine fossa that extends to posterolateral wall of the nasal cavity with an intense tumor blush (arrows) on angiography

concerns, angiography and CTA are commonly avoided in children, and instead, MRA is often performed [36].

Treatment is directed to limit the propagation of thrombus formation and reducing embolization and occlusion, while the dissection is being endothelialized over time [36]. There is still debate on which treatment option should be employed. Some authors suggest surgical therapy [37] and endovascular treatment [38], whereas others have recommended conservative options such as close observation in asymptomatic patients.

Benign Neoplasms

Figure 2.21

Malignant Neoplasms

Lymphoma (approximately 50% of cases) and rhabdomyosarcoma (approximately 20% of cases) constitute for the majority of malignant pediatric head and neck tumors [39]. Thyroid, nasopharyngeal, and salivary gland carcinomas are the most frequently noted pediatric head and neck carcinomas [39].

Lymphoma is the most common head and neck malignancy in children. Hodgkin lymphoma (HL) is primarily seen in early adolescence and is more common than non-Hodgkin lymphoma (NHL), which occurs throughout childhood. HL presents as nodal disease with a firm, nontender unilateral neck mass, or less commonly bilateral neck masses, with disease involving contiguous lymph nodes. Associated mediastinal involvement may be seen in approximately 40% of HL patients, and 80% of patients with cervical HL have disease outside of the head and neck [39].

NHL, on the other hand, presents as painless unilateral adenopathy. Approximately 30% of cases present with extra nodal disease in the head and neck and about 70% of patients have disease outside of the head and neck (65). Extra nodal NHL disease could involve the lymphoid tissue of the Waldeyer ring, or the Sino nasal, thyroid, or orbital regions. Histological subtypes of NHL in children include Burkitt lymphoma, lymphoblastic lymphoma, diffuse large B-cell lymphoma, and anaplastic large cell lymphoma.

Increased incidence of NHL is noted in children with hereditary immunodeficiencies. In immunosuppressed children, the development of lymphoproliferative disorders including lymphoma is believed to be multifactorial, related to both immunosuppressive therapy and ongoing antigenic stimulation. Infectious agents that are associated with NHL include: human immunodeficiency virus, EBV, human T-cell lymphotropic virus-1, human herpesvirus 8, Helicobacter pylori, and Chlamydia psittaci [39].

On CT, the involved nodal tissue does not enhance as avidly as infectious lymphadenitis, and stranding of the surrounding fat is usually absent. On MRI, lymphomatous involvement tends to produce enlargement of lymphoid tissue that is homogeneous and of lower signal intensity than reactive adenopathy. There is again variable enhancement that is less marked than reactive adenopathy. Whole-body imaging can be performed using nuclear medicine imaging with F-18 FDG PET for diagnosis, staging, and follow-up of disease [37].

Rhabdomyosarcoma (RMS) is the most common soft-tissue sarcoma and the second most frequent head and neck malignancy after lymphoma. Approximately 36% of cases of RMS occur in the head and neck [39]. The disease has a bimodal distribution with one peak occurring during the first decade of life and the second peak during adolescence. The common anatomic locations for RMS are the masticator space and orbit (Fig. 2.22). RMS is typically an aggressive neoplasm that erodes bone. Imaging of RMS shows a soft-tissue tumor, commonly with lytic bone destruction or occasionally bony remodeling. The tumor is usually heterogeneous, at times necrotic, and has relatively well-circumscribed borders. On contrast-enhanced CT or MRI, there is variable tumor enhancement noted. The signal intensity of tumors on T2-weighted images is variable but usually relatively iso to hypointense compared with brain reflecting the cellular nature of the tumor. When it involves the orbit it may resemble a hemangioma on imaging. However, restricted diffusion on MRI due to hypercellularity favors a

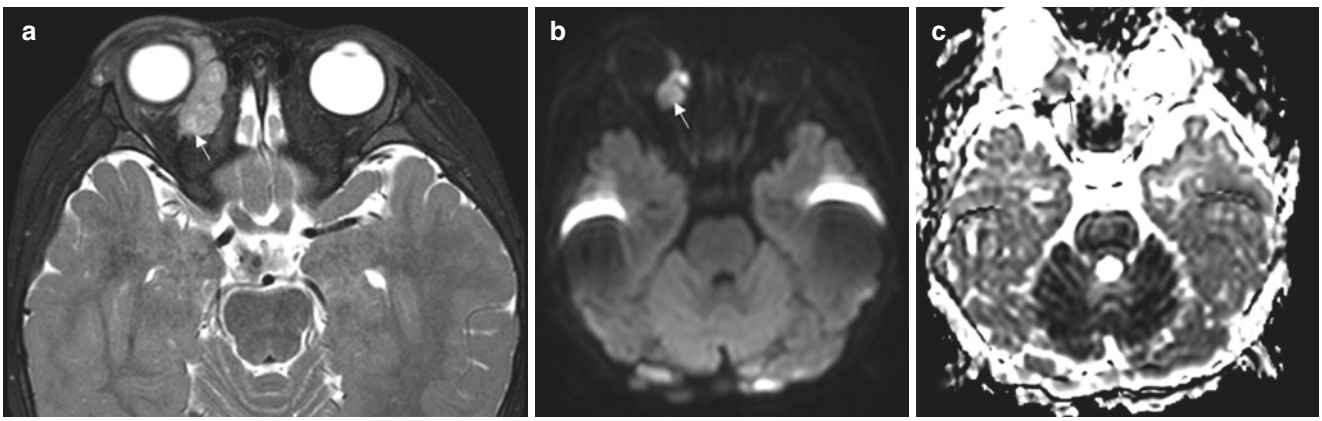

Fig. 2.21 Rhabdomyosarcoma. Axial T2 W MRI, diffusion (DW) and pparent diffusion coefficient (ADC) MR images (**a**, **b**, and **c**) showing a mass in the superomedial location of right orbit (arrow). There is restricted diffusion within the lesion on diffusion (arrow) and ADC images (arrow)

malignant lesion. Hemangiomas are benign lesions and do not show restricted diffusion on diffusion weighted MRI. Coronal contrast-enhanced fat-suppressed T1-weighted images are useful for detecting a parameningeal tumor. An assessment of the cervical lymph node chain is performed to detect metastatic adenopathy. Treatment of RMS constitutes surgery, radiation, and chemotherapy.

The most frequently occurring carcinomas of the head and neck are thyroid carcinoma and nasopharyngeal carcinoma (NPC) [39]. Thyroid carcinoma manifests as a thyroid mass with or without cervical adenopathy. The most common histological subtype is papillary carcinoma.

NPC occurs in adolescents with presenting symptoms of nasopharyngeal mass, cervical lymphadenopathy, unilateral otitis media, rhinorrhea, and nasal obstruction. On imaging NPC is visualized as a nasopharyngeal mass with cervical lymphadenopathy and aggressive characteristics including bony destruction of the paranasal sinuses and central skull base and intracranial extension.

Nuclear protein of the testis (NUT) midline carcinoma is a rare, aggressive, and fatal carcinoma that most commonly occurs in the midline of the body, which includes the head, neck, and mediastinum. It is a rare subtype of squamous cell carcinoma and is characterized by undifferentiated morphological features immunoreactive to NUT and defined by NUT rearrangement. There is a unique chromosomal rearrangement involving the NUT gene on chromosome 15 [39]. This cytogenetic abnormality is a harbinger of a poor prognosis and generally death occurs in months due to metastatic disease in spite of aggressive treatment. In the head and neck location, these tumors involve the Sino nasal region, the epiglottis or larynx. The low signal intensity on T2-weighted MR images is consistent with a cellular neoplasm; however, imaging findings are otherwise indistinguishable from other high-grade neoplasms such as lymphoma or sarcoma that are also associated with aggressive bone destruction and metastatic adenopathy.

Carcinoma involving the salivary glands is commonly mucoepidermoid in nature [39]. These tumors can be difficult to distinguish based on imaging characteristics from other parotid tumors, the most common of which is pleomorphic adenoma. The signal and enhancement characteristics of mucoepidermoid carcinoma are variable like the histological grade.

Retinoblastoma (RB) is the most common ocular malignancy in children [39]. The peak incidence occurs in first 3 years of life. Children with bilateral RB have a significant predisposition to the development of other tumors like osteogenic sarcoma, both related and unrelated to prior irradiation. Ultrasound and CT findings of RB includes an intraocular calcified mass, sometimes with associated retinal detachment. Usually the globe is normal in size or enlarged, which helps distinguish RB from other calcified lesions such as prior infection or retinopathy of prematurity. MRI is reserved for cases of bilateral retinoblastoma for evaluating for suspected extraocular extension and for the development of synchronous or metachronous tumors in the hypothalamic and pineal regions.

Children with unilateral RB undergo surgical enucleation. Bilateral RB is treated with a combination of chemotherapy and focal radiotherapy to the globe.

Metastatic disease involving the pediatric head and neck commonly involves the bony skeleton with variable involvement of cervical lymph nodes. During the first decade of life, especially in children less than 2 years, neuroblastoma is the most common [39]. Leukemic disease also causes metastases, usually in older patients, and sometimes indistinguishable in imaging appearance. Solitary and multiple facial and calvarial lesions also occur as a manifestation of metastatic disease caused by a wide variety of other tumor types, often sarcomatous, usually in older children [39]. CT demonstrates lytic, permeative bony destruction, spiculated periosteal reaction, and enhancing soft-tissue masses. On MRI, lesions are of relatively low signal on T2-weighted images with moderate to intense enhancement. Neuroblastoma causes diffuse expansion of the diploic space because of marrow involvement [39].

"Do Not Touch" Lesions

These are the lesions which mimic tumors or other pathologies on imaging but require no intervention. These include fibromatosis colli and ectopic thymus.

Fibromatosis colli is a muscular lesion which is found in newborn children (about 3 weeks after birth). It presents as swelling of sternocleidomastoid muscle, usually associated with torticollis [40]. It is assumed to be caused

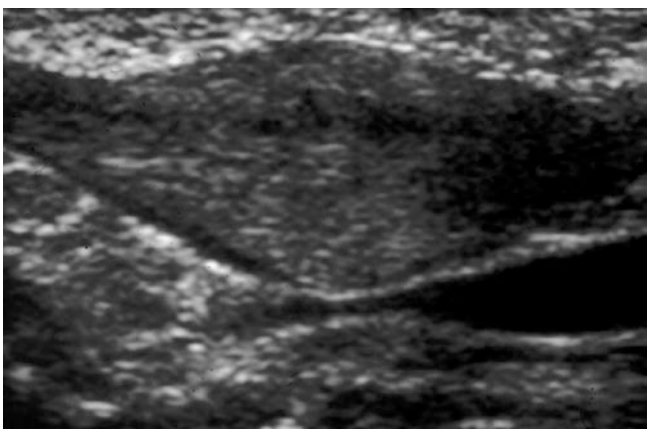

Fig. 2.22 Fibromatosis colli. Longitudinal ultrasound of neck showing an enlarged sternocleidomastoid muscle with heterogeneous echotexture

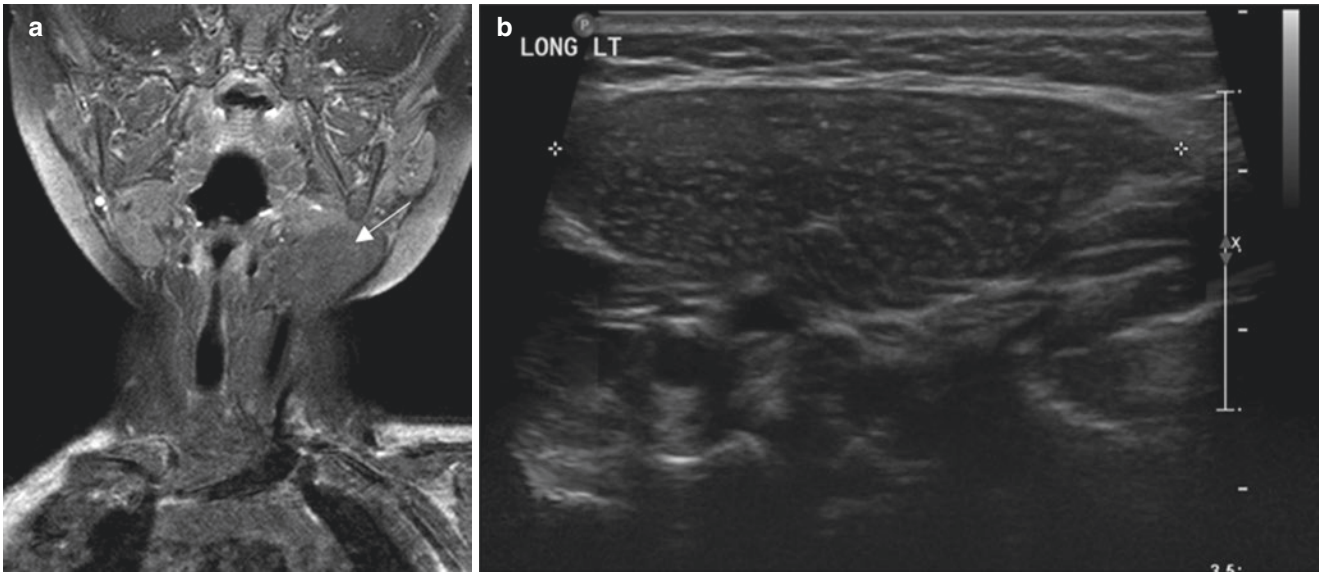

Fig. 2.23 Ectopic thymus. Coronal contrast-enhanced T1-weighted magnetic resonance (MR) image and ultrasound of the neck (**a** and **b**) showing a mild enhancing parapharyngeal mass on the left side (arrow), which is isointense relative to the mediastinal thymus on MRI. This was also confirmed on ultrasound (**b**) as this mass has same echogenicity as that of mediastinal thymus

by muscle trauma during birth. Ultrasound is the imaging modality of choice and shows swollen, fusiform sternocleidomastoid muscle in the mid portion (Fig. 2.23a), with alteration of the normal structure [40]. It may appear hypo or hyperechoic with one or both heads of the muscle being involved. The swelling usually regresses in a few months.

There are various locations and extensions of the thymus described by several studies [41]. Familiarity with these variations is essential in distinguishing thymic tissue from other pathologic masses to minimize unnecessary intervention. The cervical component of the thymus, which is occasionally detected with US, shows distinct hyperechoic foci that resembles a starry sky (Fig. 2.23b). The thymus is very pliable and hence does not cause compression or displacement of the adjacent structures [41]. This finding can be a particularly important part of a real-time sonographic examination because cardiac pulsations and respiratory motions may affect the shape of the thymus. In contrast, solid tumors or diffuse infiltrative processes are less malleable and are more rigid.

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