Pediatric radiology in oto-rhino-laryngology

Abstract

Head and neck diseases in children and adolescents present special diagnostic and differential diagnostic challenges to ENT surgeons as well as to radiologists. Both disciplines have to adapt the latest radiological and interventional technologies to the needs of the pediatric patient in order to enable a minimally invasive but successful diagnostic procedure.

High quality sonography by an experienced examiner is often the only imaging technique that is necessary in children and adolescents. Radiographs are rarely indicated in pediatric head and neck diseases. MRI, compared to computed tomography, has the advantage of absent radiation exposure. Additionally, due to current advances in high resolution techniques to delineate very small details or in visualization of different tissue characteristics, it has become an integral part of pre- and postoperative imaging.

However, children should not be denied an adequate diagnostic procedure even if it includes sedation, intervention, or exposure to radiation. The responsible use of the diagnostic options under consideration of the therapeutic consequences is essential. It is most likely to be successful in a close interdisciplinary cooperation of pediatric ENT specialists and radiologists as well as pediatric anesthesiologists in selected cases.

Although benign diseases predominate in children and adolescents, the possibility of malignancy has to be considered in cases of atypical clinical and radiological findings. In many of these young patients, the outcome and the probability of survival are directly associated with the initial diagnostic and therapeutic strategies, which should therefore be in accordance with the current guidelines of pediatric oncology therapy studies.

Our collection of clinical cases consists of representative examples of useful diagnostic approaches in common and age specific diagnoses as well as in rare diseases and malformations. It shows the significance of a special knowledge in embryology and normal postnatal development for the differentiation of normal variants from pathological findings. Only in considering the results of imaging studies in their clinical context, it is possible to succeed in detecting a syndrome behind a single malformation or adequately caring for patients with a chronic disease such as cystic fibrosis.

Keywords: ENT, pediatric, radiology, sonography, MRI

1 Introduction

Head and neck diseases in children are a special diagnostic and differential diagnostic challenge. The differentiation of age-dependent normal variations from pathological findings, the immature and small anatomy, as well as the particular spectrum of diseases require special experience and expertise of the ENT specialist and the radiologist.

This pediatric radiological expertise includes special knowledge of the embryology and the postnatal development as well as experiences with the newest imaging techniques. It is fundamental to a good interdisciplinary

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communication and a child-oriented, less stressful and effective diagnostic approach.

Methods without exposure to radiation such as highresolution ultrasound and MRI have priority to radiography and computed tomography. However, children should not be precluded from adequate diagnostics even if it is associated with sedation, intervention, or exposure to radiation. The responsible management of the diagnostic possibilities under consideration of the clinical consequences is crucial and only feasible in a good interdisciplinary cooperation.

Although benign diseases prevail in childhood and adolescence, atypical clinical and radiological findings must





Figure 1: Age-dependent size of the maxillary sinuses. Transverse section in a 6-month-old infant (A), a 3-year-old child (b; with mucosal swelling), a 10-year-old child (C), and a 16-year-old adolescent (D).

lead to the suspicion of malignant neoplasms. In many cases, the prognosis of those young patients crucially depends on the initial diagnostic and therapeutic approach. A misinterpreted radiological hint to a malignant tumor and a consequently missed opportunity of a primary R0 resection may lead to a fateful impact on the patient's probability of survival. As the treatment of children suffering from malignomas often fundamentally differs from that of adult patients, and the survival rates of children are higher if they are treated in the context of pediatric therapy studies, any suspicion of malignancy should lead to a transfer to a pediatric oncology centre. Due to such a consequent management, in Germany more than 95% of all children suffering from soft tissue sarcomas are treated according to the rapeutic guidelines in the context of acknowledged study protocols. It currently remains unclear if these better survival rates may be explained by that fact or if a different tumour biology also plays a role [1].

Often the young patient's limited abilities to cooperate are an enormous challenge for parents and physicians. The close cooperation with experienced pediatric anesthesiologists ensures an adequate and differentiated MRI examination. In such cases, anesthesia must be controlled from outside of the examination room with long ventilation pathways and only based on monitoring data during the whole procedure. With optimal coordination, an indicated surgical intervention may subsequently be performed during the same anesthesia.

1.1 Embryology

Knowledge about the development of the derivatives of the branchial pouches and the descensus of the thyroid gland is relevant not only for the diagnosis of obvious anomalies but also for the evaluation of symptoms caused by occult anomalies. This includes, for example, the identification of different cervical cysts or the correlation of several malformations to a certain syndrome.

During the 4th to 6th week of pregnancy, the branchial arches, pouches and grooves of the pharyngeal apparatus develop to become the middle ear, mastoid, and tuba auditiva (1st and 2nd branchial pouches), the palatine tonsil (2nd branchial pouch), the thymus (3rd branchial pouch), and the parathyroid epithelium (3rd and 4th branchial pouches). The thyroid gland develops from the thyroglossal duct that originates from a diverticulum on the floor of the primitive pharynx. Already in the 4th week of pregnancy the auricular placodes develop on both sides of the hindbrain (myelencephalon, the caudal part of the rhombencephalon). They develop via the auricular dimples and vesicles into a primitive membranous labyrinth which becomes the inner ear between the 8th and 20th week of pregnancy [2].

1.2 Postnatal development

In this context especially the development of the paranasal sinuses and the mastoid have to be considered. The age-dependent degree of pneumatisation and the frequent asymptomatic mucosal swellings in childhood should not lead to unnecessary radiography or to the false diagnosis of sinusitis [3].

The maxillary sinuses already develop in the fetus of the 2^{nd} trimenon. At the time of birth it is very small and is situated medially to the orbit. Up to the age of 9 years, it has nearly reached the level of the hard palate (Figure 1). The development of the ethmoid cells starts about the time of birth in an anterior to posterior direction. At the age of about 6 years, the posterior cells are fully de-



Figure 2: Regular findings of a developing sphenoid sinus of a 5-year-old child: focal inhomogeneous hyperintense signal because of high water content, STIR sequence coronal (A) and transverse sections (B; long arrows). Increased signal by fat, T1 sagittal sections (C; short arrows).

veloped, the complete pneumatisation, however, is only achieved with puberty. The development of the sphenoid sinus variably starts at the age of 2 years. During the development of the sphenoid sinus, the physiological transformation of the medullary cavity of the sphenoid bone into pneumatised cells leads to significant alterations of the signal intensities on MRI which may easily be misinterpreted as tumors (Figure 2). The development of the frontal sinus may also start at the age of 2 years, but most often it begins later. The mastoid cells already develop in the fetus. At about 6 years, pneumatisation is completed.

2 Imaging modalities

2.1 Radiography (conventional X-ray)

For ENT specific diseases in children, radiographs are indicated only in exceptional cases. It must be considered that the high percentage of hematopoietic bone marrow of the skull (27% in newborns, 16% in 5-year-old children, 8% in adolescents) should not be unnecessarily exposed to radiation [4], [5].

Because of the above-mentioned variable postnatal development of the paranasal sinuses and the frequent, asymptomatic mucosal swellings in children, radiographs are not regarded as useful for diagnosing a sinusitis (case report 11). For the depiction of the complex structures of the paranasal sinuses, but also of the temporal bone, cross-sectional techniques are much more appropriate. In our department, radiographs of the temporal bone region are only performed for documentation of the position of implantable hearing systems (cochlear implant, implantable hearing aids).

For identification or documentation of a radiopaque foreign body in the airways or the gastro-intestinal tract, a very low contrast imaging is mostly sufficient so that, depending on the size of the object, extremely dose-reduced digital imaging may be applied (case report 6). High resolution video fluoroscopy for the diagnosis of dysphagia is, with the necessary effort, possible even in infants and toddlers, however, also because of the radiation exposure it is reserved for specialized centres with appropriate therapeutic possibilities.

Conventional angiography is preferably applied when a simultaneous intervention is planned (case report 8). Merely diagnostic angiography of high quality can also be performed by MRI.

In pediatric otolaryngology, the possibilities of intraoperative imaging become more and more important, which in case of CT and MRI are associated with high technical and financial efforts. Alternatives for the demonstration of bony structures are digital volume tomography (DVT) or digital C-arm systems with 3D functions [6], however, the experience with pediatric patients is currently rather low. Despite the reduced radiation exposure compared to CT scanning, an especially careful benefit-risk analysis is therefore necessary.

2.2 Ultrasound

High-quality ultrasound is highly meaningful in many processes of the ENT region in children and adolescents. Its main advantages are the rapid and uncomplicated availability, the possibility of assessing the behavior of pathological processes under focal pressure, and the good visualization of vessels by Doppler sonography. It is often the only necessary imaging technique. Experienced examiners may - in knowledge of the MRI findings - employ sonography for reliable follow-up examinations even of complex findings. With linear transducers of 10-15 MHz and high-resolution near-field sonography a higher local resolution may be achieved than with MRI. The limitations of ultrasound of the head and neck region are mostly due to air in the airways (e.g. while assessing the retropharyngeal space) and acoustic shadowing of bony structures such as the mandible and the mastoid (e.g. skull base).



2.3 Magnetic resonance imaging (MRI)

MRI is an extremely versatile tool to demonstrate normal and pathological tissue properties. Beside a multitude of sequence techniques also high-resolution (Voxel <1mm) 3D imaging with subsequent multiplanar reconstruction of the head and neck region are possible. Recent technical developments allow - with 3-Tesla devices - an isotropic spatial resolution of 0.32 mm with high quality so that even the neuro-epithelium of the macula utriculi can be delineated in the vestibulum of the inner ear [7]. For an adequate choice of MR sequences and sections an exact interdisciplinary communication about the clinical questions that have to be answered is essential. This close cooperation helps to avoid over- as well as underdiagnosis. An examination that takes too much time inevitably leads to increased movement artefacts especially in young children. A too short examination with "wrong" sequences may prevent a rapid diagnosis and thus lead to unnecessary subsequent examinations.

Child-oriented conditions are a general prerequisite for a successful MR examination. With the parents present and audio or video tapes available, even younger children (4–8 years) are able to keep still for a sufficient period of time. In our department, toddlers and infants are examined under sedation with Propofol, and, if necessary, airway management with laryngeal masks, and appropriate monitoring by a pediatric anesthesiologist.

The indications for contrast-enhanced examinations also depend on the clinical questions. Thus the identification of inflammatory processes, especially after an abscess, or the delineation of tumors nearly always require the application of contrast agents, while merely anatomic evaluations may mostly be performed without injections. Also for MR angiography, the choice of the technique and the decision for or against contrast application depend on the question to be examined. In this context an open interdisciplinary exchange about the newest examination and surgery techniques seems to be indispensable in order to fulfill the mutual requirements. According to our experience, a current example is the visualization of cholesteatomas by diffusion-weighted MR sequences [8]. This relatively easy examination technique requires a special selection and the exact spatial alignment of different MR sequences. Only intensive interdisciplinary exchange and close correlation of imaging findings and surgical aspects allow a rapidly rising learning curve.

2.4 Computed tomography (CT)

For CT the same considerations on dose reduction are valid as for radiography, however, with the main difference of a much higher radiation exposure. The average CT scan of the skull with an effective dose of 2.3 mSv corresponds to about 35 X-rays of the skull or 115 X-rays of the thorax, and it is equivalent to a natural radiation exposure of about one year (http://www.imagegently.org/). Further, in comparison to adults, children have a much higher radiosensitivity beside a long average life expectancy in

which malignomas may develop [5]. Before performing CT scans, the diagnostic alternatives should therefore be carefully considered.

The domain of CT is the visualization of bony and aircontaining structures and their anomalies. It has significance especially for trauma diagnostics and the depiction of the middle ear and the external meatus. The inner ear and the inner meatus may only be indirectly assessed while three dimensional MRI allows their direct visualization [9] (case report 15). All processes with soft tissue involvement such as tumors or sinugenic abscesses may be better examined by MRI and only rarely require additional CT imaging of the bones (case reports 12, 14a). It has to be mentioned that also in tertiary centres adequate MR imaging is not always available, e.g. on weekends or at inconvenient times. The examination of young uncooperative patients may be an additional challenge. The ideal constellation would include the imaging and a possible surgical intervention under the same anesthesia. However, this is only possible in institutions that are logistically and medically focused on children as for example the Olgahospital of the Klinikum Stuttgart, Germany.

3 Case reports

In the following chapter some important particularities of radiology in children with ENT specific diseases will be explained on the basis of case reports. Each of our mutual patients hereby represents a group of typical pediatric ENT patients. For further information, we refer to specific literature and textbooks [2], [10], [11], [12].

For MRI examination of the neurocranium T1 and T2 weighted sequences are commonly applied which thus also depict the incidental findings. Water has a low signal intensity in T1 weighted images, and a hyperintense signal in T2. Fat appears with high signal intensity in both sequences.

For examination of the viscerocranium, before contrast application STIR sequences are applied that suppress the fat signal (low signal intensity) and show water containing structures with high signal intensity. After contrast application, fat saturated T1 sequences are applied (contrast agent with high signal intensity, water and fat with low signal intensity).

3.1 Neck

Oncologic diseases of the neck are rare in children. In contrast to adults, their vast majority is of mesenchymal origin (lymphomas, sarcomas etc.). In practice, benign cystic malformations (median and lateral) and inflammatory processes of the lymphatic system (lymphadenitis colli) are most frequent.

In most pathological processes of the neck in children, sonography of high quality is the first and only imaging method. However, radiologists should always be aware that the anatomic space of the neck begins cranially at





Figure 3: (A) Case report 1. Ultrasound. Linear transducer with 12 MHz. Sagittal section in the midline of the neck. Median cervical cyst (crosses), moderately echogenic content, adjacent to the lingual bone which causes a dorsal acoustic shadow (arrows). (B) Case report 1a. Ultrasound. Linear transducer with 12 MHz. Transverse section through the right sternocleidomastoid muscle (short arrows). A small lateral cervical cyst is found in typical position at the anterior edge.

the skull base and continues caudally to the upper thoracic aperture. In case of inflammatory or tumoral processes, those possible propagation pathways have to be considered. By means of MRI, which in this context is clearly superior to ultrasound, the skull base including the neuro-foramina must be depicted in cases of location in the cranial third, and the upper thoracic aperture must be displayed in cases of involvement of the caudal third. CT scans play only a subordinate role in the imaging of soft tissue.

3.1.1 Case report 1

Diagnosis. Median cervical cyst (Figure 3A).

General information. Residuum of the embryonic thyroglossal duct. Most frequently (80%) it is located in the midline and has a relation to the hyoid bone. The content of the cyst is often moderately echogenic due to the protein content and can also be septate [11]. Ideally the imaging is performed in a non-acute interval. Many cysts become symptomatic because of their increasing volume in the context of general diseases. In single cases, the infected cyst may appear as cervical abscess.

Case description. Increasing painless swelling in the midline at the transition of the floor of the mouth to the neck.

Which information and specific clinical question does the radiologist need to know? Previous inflammations and discharge of cystic content must be mentioned because both events may permanently change the appearance of cysts. The temporal course of the manifestation and/or clinical growth parameters are important.

Differential diagnosis and limitations of imaging. The most frequent differential diagnosis are lymph nodes that are incidentally located in the midline. A cyst can develop in the whole course of the thyroglossal duct from the base of the tongue (foramen caecum) to the thyroid gland. In cases of a mass in the base of the tongue, the orthotopic thyroid should be documented in order to avoid confusion of a dystopic gland with a cyst. It is possible that a lingual thyroid represents the only hormonally active tissue, and

that its removal under the suspicion of a median cervical cyst leads to hypothyreosis.

Ultrasound provides only limited access to cysts in the base of tongue so that depiction by MRI is preferable. In all imaging modalities the delineation of a retrohyoid fistula is unreliable and cannot replace the surgeon's decision to explore the base of tongue.

3.1.2 Case report 1a

The diagnostic procedure for lateral cervical cysts is similar (Figure 3B). Also in these cases it is not possible to radiologically verify the total extension of a fistula tract to the pharynx.

3.1.3 Case report 2

Diagnosis. Synovial sarcoma of the larynx (Figure 4).



Figure 4: Case report 2. MRI in transverse section. Left: STIR. Hyperintense, partly inhomogeneous formation at the left side of the larynx. Right: T1 SE with fat suppression after contrast agent application. Enhancing nodules (arrows) in the "cyst". Therefore, this is not a simple cyst but strongly suspicious of a cystic (malignant?) tumor.

General information. Diagnosis and treatment of soft tissue sarcomas are performed in pediatric oncology centres according to the guidelines of specialized study groups: EpSSG (European Paediatric Soft-Tissue Sarcoma Study Group: http://epssg.cineca.org), CWS (Cooperative Weichteilsarkom Studiengruppe http://cws.olgahospitalstuttgart.de, http://www.kinderkrebsinfo.de), COG STS (Children's Oncology Group Soft Tissue Sarcoma Committee, USA, http://www.cancer.gov/cancertopics/types/ childrhabdomyosarcoma). The clinical findings of a sarcoma may be rather unspecific and mimic less harmful differential diagnoses. The experienced pediatric radiologist can and should be the decisive "advisor" for the ENT specialist to find the correct diagnosis.

A central requirement for the histological assessment of a suspected sarcoma is to avoid the spreading of tumor cells into the surrounding tissue. Thus imaging is eminently important for the planning of the surgical approach. **Case description.** The 9-year-old child presented with unspecific but increasing complaints (foreign body sensation when swallowing, mild hoarseness). Because of a smooth protrusion of the pharynx with displacement of the epiglottis, which became obvious during an examination in a private practice, the emergency presentation in our hospital was indicated. Endoscopy could confirm the findings, however, primarily a congenital laryngeal cyst was suspected in the sense of an encapsulated laryngocele.

Which information and specific clinical question does the radiologist need to know? The exact description of the clinical and endoscopic findings, and the principal symptom.

Differential diagnoses and limitations of imaging. Benign cysts are frequent in otolaryngology. Their lumen may be filled with echogenic material (see case report 1). A focal especially nodular wall thickening should always lead to the suspicion of a malignant disease, even in children. An appropriately careful surgical management is essential (with RO resection whenever possible). In case of suspected sarcoma, the experienced pediatric radiologist should emphasize the particular requirements of the histological confirmation and the potential risks of iatrogenic spreading of tumour cells.

The anterior and lateral wall structures of the larynx and the trachea may be well assessed by means of ultrasound, with only few limits caused by the aerated lumen. The exact dimensions of soft tissue lesions may best be displayed with high resolution MRI (slice thickness of max. 5 mm, pixel size of <1 mm) with fat saturation before and after contrast agent application.

3.1.4 Case report 3

Diagnosis. Parapharyngeal abscess (Figure 5).

General information. Cervical abscesses are most frequently associated with pharyngeal inflammations (tonsillitis etc.) or they occur in the context of necrotizing lymphadenitis. The differentiation of possible causes is a particular diagnostic challenge in children. It should always be taken into consideration that malformations may be the primary origin of such findings. The typical clinical findings with protrusion of the tonsillar region and the palate are not always present in deeply located abscess cavities. In search for a fluid retention, ultrasound or MRI are very helpful. Especially persistent swallowing disorders (refusal to drink in infants and toddlers) should lead to an exact examination of the parapharyngeal and retropharyngeal space. A restricted mouth opening indicates the involvement of the masticatory muscles, e.g. retrotonsillar or dental abscesses. Paratonsillar abscesses may already develop within the first year of life. They may occur bilaterally and lead to a life threatening event (in case of a failing intubation).



Figure 5: Case report 3. Transverse section at the level of the pharynx. Above: STIR. Hyperintense formation on the left side of the pharynx (short arrows). Below: T1 with fat suppression after contrast agent application. Ring-shaped enhancement with central focus of low signal intensity, leading, in the context of the clinical findings, to the diagnosis of a parapharyngeal abscess.

Case description. At the time of first presentation, the 17-year-old girl reported about increasing unilateral sore throat and swallowing complaints. The patient had been intermittently treated with immunosuppressants for several years because of juvenile arthritis. She was off treatment at the time of presentation. Her family physician had introduced an oral antibiotic therapy which did not improve the complaints. Emergency hospital admission was therefore indicated. The clinical examination revealed no significant protrusion of the palate. The tonsils appeared relatively symmetrical and only slightly reddened. As ultrasound led to the suspicion of a retrotonsillar





Figure 6: Case report 4. Typical findings of an embryonal rhabdomyosarcoma. Coronal section. On the left: STIR: Extensive tumor in the right fossa pterygopalatina reaching the skull base and the oval foramen (long arrow). On the right: The reconstruction of a T1 3D sequence with fat suppression and contrast agent application shows the decisive findings of the intracranial meningeal involvement (short arrows).

abscess, tonsillectomy was performed and the abscess cavity behind the left tonsil was drained.

Four weeks later, the swallowing disorders re-appeared. MRI showed a parapharyngeal abscess which was surgically drained. In the course of the following three years, 4 further abscess events occurred which were 2 times transcervically and 2 times endoscopically drained.

Which information and specific clinical question does the radiologist need to know? Duration of anamnesis, previous therapeutic approaches, the presence of swallowing complaints (refusal to drink in infants and toddlers) or restricted mouth opening.

Differential diagnosis and limitations of imaging. In pediatric patients, sonography of a lymphadenitis colli should include not only the superficial but also the deep parapharyngeal lymph nodes. An experienced examiner can often identify the parapharyngeal or retropharyngeal extent of an inflammation or abscess cavity by ultrasound. The extension to the skull base, however, cannot be covered by sonography. In cases of doubt, MRI is always indicated or alternatively – if MRI is not available – a CT scan with contrast agent application.

3.1.5 Case report 4

Diagnosis. Embryonal rhabdomyosarcoma (eRMS) of the right fossa pterygoidea with intracranial extension via the foramina of the skull base (Figure 6).

General information. The location of an eRMS at the skull base is typical. The description of the exact extent (a parameningeal location with intracranial extension has a poorer prognosis) is essential for therapy planning and prognosis. Diagnosis and treatment of soft tissue sarcomas are established according to the guidance of acknow-ledged study groups (see case report 2). Due to the optimized multimodal therapy in the frame of these guidelines, the prognosis of the disease could be enormously improved during the last 30 years.

Case description. A 6-year-old boy presented with a slowly progressive indolent swelling of the right parotid region. Endoscopic examination revealed a protrusion of the right pharyngeal wall. The radiological suspicion of a sarcoma was histologically confirmed by endoscopic biopsy through the pharyngeal wall.

Which information and specific clinical question does the radiologist need to know? Type and duration of the symptoms and the endoscopic findings.

Differential diagnosis and limitations of imaging. For therapy planning and assessment of the prognosis, highresolution MRI (under sedation if needed), with exact depiction of the whole local extension and involvement of locoregional lymph nodes according to the study guidelines is necessary. Diagnostics and therapy are ideally performed in a pediatric oncology centre according to the appropriate therapy study.

3.1.6 Case report 5

Diagnosis. Infantile hemangioma of the parotid gland (Figure 7).

General information. Hemangiomas are the most frequent soft tissue tumors in infants. They often occur in the parotid gland, but may be found everywhere in the face, head, and neck. Meanwhile local therapy is only required in rare cases because systemic therapy with propranolol can induce involution in a high percentage of the cases. Case description. A 4-month-old preterm born infant was presented with increasing swelling of the right parotid gland. Ultrasound showed the typical mass of multiple, well delineated hypoechoic parts of the salivary gland. Doppler sonography revealed numerous veins and arteries with high flow rates (spectral analysis!).

Which information and specific clinical question does the radiologist need to know? Growth rate of the swelling and cutaneous or mucosal manifestations.





Figure 7: Case report 5. Ultrasound of an infant with indolent swelling of the right parotid gland. Linear transducer of 14 MHz. Above: B-mode image. Below: Color Doppler sonography. Typical findings of an infantile hemangioma.

Differential diagnosis and limitations of imaging. Diagnosis can be very reliably confirmed by ultrasound. If the age of the child, the anamnesis, and the findings are typical, no further diagnostic is necessary. Differentiation from lymphangiomas, congenital hemangiomas, or congenital vascular malformations is also possible by the patient's history and by Doppler sonography. Flow profiles and calibers of the afferent arteries (e.g. facial artery) give hints on hemodynamic changes during therapy. Further imaging (MRI with dynamic contrast-enhancement) is only required if the margins cannot be completely depicted by sonography (e.g. intraorbital extension).

3.1.7 Case report 6

Diagnosis. Foreign body (coin) in the hypopharynx/entry of the esophagus (Figure 8).

General information. Unequivocal anamnesis and symptoms require an endoscopy to look for and remove the foreign body without further diagnostics. However, sometimes the anamnesis is not clear so that conventional radiography of the thorax allows a rapid statement about the location of a metallic (radiopaque) foreign body. In cases of unclear aspiration events, such an examination is recommended also for medico-legal reasons. The differentiation between an object that is stuck in the esophagus and one that is already in the stomach, has a direct therapeutic consequence. In the latter case, endoscopic extraction can be avoided in most of the cases. Radiological imaging, however, should only be performed if it could have an impact on the further management.



Figure 8: Case report 6. Two-year-old child who had played with coins 3 days before. Now refusal to drink and hypersalivation. Last image hold (LIH) of fluoroscopy with very low radiation exposure (not measurable with standard devices). Despite the high level of image noise there is a clear documentation of a coin in projection onto the hypopharynx.

Case description. A 2-year-old child had been observed playing with several coins that he had also put into his mouth. His mother reported about refusal to drink and drooling.

Which information and specific clinical question does the radiologist need to know? The (probable) time of ingestion and the symptoms should be reported (swallowing and/or breathing complaints).

Differential diagnosis and limitations of imaging. Dosereduced conventional X-ray is sufficient in most of the cases. Modern digital radiography and fluoroscopy devices allow a significant reduction of the radiation dose, so that, depending on the kind of the foreign body, images of just acceptable quality according to the ALARA principle (as low as reasonably achievable) suffice.

3.1.8 Case report 7

Diagnosis. Lymphadenitis colli caused by atypical mycobacteria (Figure 9).

General information. In childhood, an atypical lymph node tuberculosis is not rare and presents with partly indurated multifocal lymph node swellings of the neck without





Figure 9: Case report 7. Three-year-old girl with indolent submandibular swelling on the right side. Sonography (left): Enlarged hypoechoic lymph node with loss of the normal structure and development of satellite nodules (arrows) in direction to the skin surface, suggestive of an infection with atypical mycobacteria. MRI shows a hyperintense lesion in the STIR sequence in a similar section (right).

general signs of infection. Often a cutaneous penetration of the infection with development of a fistula is observed. The focal livid discoloration of the skin covering the enlarged lymph node is characteristic. Primary therapy includes the complete surgical resection of the affected lymph nodes. Ultrasound by an experienced examiner is often the only necessary imaging procedure in this context. Important sonographic signs of an infection with atypical mycobacteria are: development of satellite-like nodules at the edge of the lymph node, fistula development to the skin as well as echogenic intrinsic reflexes with and without acoustic shadows [13].

Case description. At the time of first diagnosis, a 3-yearold girl presented with an indolent swelling of the right submandibular region that had been observed for about 8 weeks. No general signs of infection were reported.

Which information and specific clinical question does the radiologist need to know? Type and duration of symptoms, signs of infection, pain.

Differential diagnosis and limitations of imaging. The interdisciplinary synopsis of all findings allows the differentiation from bacterial lymphadenitis. A clear differentiation from "typical" tuberculosis is not possible by means of imaging. Further diagnostics such as MRI are only required if the margins of the findings cannot be completely assessed by sonography. Differential diagnosis: Actinomycosis also leads to livid skin discoloration and fistula development, however, typically does not spread via the lymphatic system [14].

3.1.9 Case report 8

Diagnosis. Ganglioneuroma in the right carotid sheath (Figure 10).

General information. Isolated benign nerve sheath tumors outside the neurocranium are generally rare in children and adolescents. Such tumors are mostly found in the neck where the vagus nerve (N X) is considered as origin. Symptoms are unspecific because the tumor grows very slowly. Swallowing complaints and hoarseness are reported. Imaging is primarily performed with ultrasound while

the exact description of the relation to the skull base requires an MRI. It is the method of choice to document the exact location of the tumor with regard to the surrounding structures [11], [12]. As the growth dynamics of the disease are usually unclear at the time of first diagnosis, short-term follow-up by sonography or MRI belongs to the standard if general symptoms are absent. In cases of large or progressive findings, the complete extirpation of the tumor via a cervical approach should be achieved. Constant or very slowly growing lesions may be observed for several years by appropriate follow-up imaging ("waitand-see").

Case description. A palpable indolent swelling on the right side of the neck was found in a 10-year-old boy. The symptoms were intermittent hoarseness and swallowing complaints. The ENT specific examination revealed a significant narrowing of the pharynx by a protrusion of the right lateral wall. The mobility of the vocal folds was normal during flexible endoscopy. The lesion was sono-graphically identified as well vascularized tissue. No cyst was found.

Which information and specific clinical question does the radiologist need to know? Duration and type of complaints, if present also general accompanying symptoms. Signs of inflammation? Clinically suspected diagnosis?

Differential diagnosis and limitations of imaging. The differential diagnoses are, among others, a lateral cervical cyst, swollen lymph nodes or an adenoma. The differentiation of a parapharyngeal salivary gland adenoma is important for the surgical removal because an intraoperative disruption of an adenoma may lead to an uncontrollable local tumor dissemination. Hormonally active glomus tumors are extremely rare in children [11].

3.1.10 Case report 9

Diagnosis. Lymphangioma of the left parotid space (Figure 11).

General information. Lymphangiomas are benign cystic soft tissue tumors. About 50% of them are congenital. All





Figure 10: Case report 8. Ten-year-old boy with a big oval mass in the right carotid sheath. Histology: Ganglioneuroma. MRI (left): STIR in coronal sections with numerous low-signal flow voids (arrows) as a sign of high blood flow. Bright contrastenhancement in the early phase (not shown). Conventional angiography (right): Tumor blush after selective canulation of the right ascending pharyngeal artery. Subsequent preoperative embolization. (Courtesy of Prof. Hans Henkes, Department of Neuroradiology, Klinikum Stuttgart, Germany).



Figure 11: Case report 9. 16-month-old girl with soft swelling at the left mandibular angle. (A). Sonography: Linear transducer of 14 MHz. Typical findings of a lymphangioma with delicate wall structure and anechoic content. (B) MRI can display the extension medially to the mandible (in the acoustic shadow at sonography, arrow).

parts of the body may be affected; most frequently they appear in the head and neck region. Their size and extent vary enormously, from only a few millimeters up to giant disfiguring tumors. An acute enlargement is mostly caused by spontaneous bleedings with typical fluid-fluid levels in cross-section images. Combined malformations with hemangiomas are possible. Spontaneous adhesion and involution are possible after bleeding. An obliterative therapy by sonographically controlled injection of Picibanil (OK-432) is promising in cases of macrocystic lymphangiomas.

Surgical extirpation should be planned very carefully in order to avoid accidental neural lesions. Incidental observations of regressive lymphangiomas as side effect of a treatment with Sildenafil in children with pulmonary hypertonia still need to be confirmed in controlled studies [15].

Case description. The parents of a 16-month-old girl reported about a soft swelling at the left mandibular angle. Which information and specific clinical question does the radiologist need to know? Duration of history, growth behavior, discoloration (e.g. bluish shining through the skin).

Differential diagnosis and limitations of imaging. The diagnosis can almost always be confirmed by ultrasound. Thin septa of the multicystic lesion with anechoic content or with fluid-fluid levels are pathognomonic. Those are only visible after bleedings and in calm patients (other-



Figure 12: Case report 10. MRI STIR transverse section (left) and sagittal section (right). Well delineable round tumor paramedian on the left side, which displaces the teeth and thus clearly originates from the maxilla. Congenital melanotic neuro-ectodermal tumor.

wise sedimentation is not possible). MRI is indicated if an extension to the skull base, in retropharyngeal direction, or into the thorax cannot be completely documented by sonography.

3.2 Nose

Many symptoms with origin in the nasal region which lead to a child's presentation to an ENT specialist may be treated without imaging. This is only required if complaints and clinical findings – also considering the age of the children – deviate from the typical symptoms.

Because of the bony and air-containing structures, the possibilities of ultrasound are limited to superficial, subcutaneous processes; however, it has its clear value, for example, for the diagnosis and monitoring of hemangiomas. An excellent overview of the nasal cavity and the paranasal sinuses as well as the epipharynx may be provided by MRI so that only exceptional cases have to be referred to computed tomography.

All radiological findings have to be considered in their clinical context. Especially the physiological and age-dependent, mostly harmless mucosal swellings in the paranasal sinuses always require a close correlation to the clinical findings, in order to avoid a wrong diagnosis of sinusitis and unnecessary therapeutic interventions. In children, even sonography and radiography may not differentiate these frequent mucosal swellings from pathological processes. On the other hand, an intracranial or intraorbital extension of an abscess must not be overlooked. Our case report 15 on a unilateral choanal atresia additionally demonstrates the importance of malformations of the ENT region for the detection of an underlying complex syndrome. A holistic view is also necessary for patients suffering from chronic primary disease, as explained by the example of cystic fibrosis (case report 13).

Malignant neoplasms in the nasal region are rare in children, however, they require our full attention regarding a careful diagnostic and therapeutic approach according to oncologic guidelines. Specialized interdisciplinary pediatric oncology boards facilitate such procedures and may be very useful also in cases of rare tumors with benign radiological appearance.

3.2.1 Case report 10

Diagnosis. Congenital melanotic neuro-ectodermal tumor (Figure 12).

General information. Rare congenital, rapidly growing tumor in typical location with progressive behavior [11], [16]. Therapy consists of local control by complete resection. In this context, imaging plays an important role for the exact delineation of its margins. The radiological findings determine the planning of the resection and the discussion of possible reconstructive measures.

Case description. Some months after birth, a smooth protrusion into the oral vestibule became obvious. The development of the child was not impaired. The encapsulated tumor was surgically removed and intraoperatively proved to originate from a dental germ.

Which information and specific clinical question does the radiologist need to know? Begin of symptoms resp. clinical appearance. Further the time course of progression is important.

Differential diagnosis and limitations of imaging. The diagnosis may radiologically be suspected, however, a histological confirmation is required. Differential diagnosis: hemangioma (mostly without involvement of the bone).

3.2.2 Case report 11

Diagnosis. Mucosal swelling of the paranasal sinuses (Figure 13). Incidental findings in a cranial MRI without clinical relevance.

General information. In children mucosal swellings of the paranasal sinuses and mastoid cells are frequent incidental findings. Even a severe mucosal swelling is not necessarily indicative of infection. Therefore in radiological reports the terms "sinusitis" or "mastoiditis" should be employed with great caution. Treatment may be introduced only on the basis of clinical symptoms and not on radiological findings alone.

An evaluation of MRI examinations of 147 children and adolescents in our institution, who had received a cranial MRI because of other reasons than sinusitis or mastoiditis, showed one or more conspicuous findings within the their paranasal sinuses or mastoid cells in 61% of the cases. A total of 48% had mucosal swellings of the paranasal sinuses, 25% or the mastoid cells. The prevalence was higher in children below the age of 10 (60% and 42%, respectively) and in children with current infection of the upper airways (71% and 35%, respectively). A correlation to headache was not found [3].



Figure 13: Case report 11. MRI T2 transverse section. Seven-year-old boy suffering from headaches without ENT specific symptoms. Incidental finding of extensive mucosal swelling of the ethmoid sinuses and a still small sphenoid sinus. Findings without pathological relevance.

Case description. A 7-year-old boy who had received a cerebral MRI because of recurrent headaches.

Which information and specific clinical question does the radiologist need to know? A clear description of symptoms is appropriate.

Differential diagnosis and limitations of imaging. MRI (and CT as second choice) with contrast is an excellent tool to reveal intraorbital and intracranial complications of sinusitis or mastoiditis (case reports 14 and 14a). However, an uncomplicated sinusitis remains a clinical and not a radiological diagnosis. Because of the frequently occurring mucosal swellings in healthy children, radiography, MRI, CT, and ultrasound have only low diagnostical specificity for the presence of sinusitis.

3.2.3 Case report 12

Diagnosis. Schwannoma of the nose (Figure 14).

General information. A tumor of the nasal cavity is extremely rare in children. The most important clinical question concerns the dignity of the mass and the resulting surgical options. Before histological confirmation, this issue may only be addressed with the help of indirect signs such as characteristics of the surface, delineation of margins etc. MRI is the method of choice for assessing the tumor extension and delineating neighboring structures. Imaging can only support diagnostic efforts to specify the final diagnosis. The exact radiological delineation of the tumor margins, however, enables exact surgical planning with high reliability. Often, a biopsy for histological confirmation is performed as a first step. However, in cases of confident delineation of a morphologically benign mass, also a complete primary resection and thus healing is possible.

Case description. Slowly progressive unilateral nasal obstruction with chronic sniffing was reported. Endoscopically, a subtotally obstructing mass with a smooth surface was found in the right nasal cavity.

Which information and specific clinical question does the radiologist need to know? Duration and type of subjective complaints, the clinically suspected diagnosis, and the endoscopic findings.

Differential diagnosis and limitations of imaging. The question of probable malignancy arises first. As the duration of complaints is often difficult to assess in children, imaging details are helpful to interpret the clinical course of the disease. For example, the deviation of the nasal septum and the deformation of the medial wall of the maxillary sinus are signs of a slow tumor growth and support the probability of a benign lesion. A smooth delineation under intact epithelium in the endoscopic and radiological imaging may be interpreted as a sign of mesenchymal origin of the mass. As in this particular case the tumor completely filled the nasal cavity, its origin could not be revealed by imaging techniques.

3.2.4 Case report 13

Diagnosis. Chronic rhinosinusitis with recurrent nasal polyposis in mucoviscidosis (cystic fibrosis = CF) (Figure 15).

General information. CF is the most frequent autosomal recessive genetic disease of Caucasians. It is caused by more than 1,000 different mutations (http:// www.genet.sickkids.on.ca) of the CFTR gene (cystic fibrosis transmembrane conductance regulator gene) located on the long arm of the chromosome 7 in the region 7q31. Due to the standardization of treatment of CF, the mean expectancy of life for this patient population has been doubled during the last 30 years. That is why clinical issues beside typical pulmonary and gastrointestinal manifestations occur more and more frequently in older patients.

In CF patients, two ENT specific symptom complexes are known: (1) nasal obstruction, anosmia, chronic sinusitis, and severe nasal polyposis as well as (2) ototoxic inner ear damage as consequence of repeated systemic application of aminoglycoside antibiotics.

CF involvement of the upper airways is mainly assessed by imaging. This is especially important for the discussion of a possible surgical intervention. In this context MRI is





Figure 14: Case report 12. 1. CT scan in coronal section without contrast agent application (left). MRI STIR in coronal (middle) and transverse sections (right). Large mass of the right nasal cavity which deforms the sinus wall (arrow) as well as the septum. Histology: Schwannoma. Mucosal swelling of the right ethmoid and maxillary sinus. 2. (A) intraoperative endoscopic finding of bright tumor tissue (#) in the right nasal cavity (UM=inferior turbinate, SP=septum). (B) endoscopic view of the tumor stalk (#) located at the nasal septum (SP) after complete resection. (MM=middle turbinate, ...=epipharynx).



Figure 15: Case report 13. 1. 14-year-old patient with CF. Coronal STIR sequence. Extensive hyperintense signal in the paranasal sinuses and the nasal cavity because of mucosal proliferation. Inferior nasal turbinate still delineable on both sides (white arrows). The walls of the maxillary sinuses are medially displaced, the middle turbinate can no longer be identified (black arrows). 2. 14-year-old female patient with CF. (A) Polyposis of the right middle nasal passage as shown by MRI. Polyps and small mucoceles are resected by means of a shaver with oscillating knives (#). (B) Free view into the middle nasal passage lateral to the middle turbinate (*) in direction of the posterior ethmoid cells after resection of the polyposis.



primarily recommended. This method has major advantages regarding discrimination. With the help of contrast agents it allows to differentiate well between retained secretion and mucosal swelling. The second significant advantage is that it avoids radiation exposure. The CT, as an imaging alternative, should be avoided especially in cases of repeated examinations, as they may be required in CF patients. The cumulative radiation exposure must not be underestimated. Recent epidemiological long-term evaluations point to the increased lifetime cancer risk of patients who underwent a CT scan in childhood [17].

Case description. The 14-year-old girl complained about increasing nasal obstruction. Endoscopic examination revealed a typical bilateral nasal polyposis and medialization of the lateral nasal walls. The tympanum on both sides was aerated and without mucus retention.

Which information and specific clinical question does the radiologist need to know? Basic knowledge of the underlying disease of CF. Duration and type of complaints and possible complications.

Differential diagnoses and limitations of imaging. The documentation of pathological alterations of the paranasal sinuses complements the endoscopic and clinical findings, but is not as decisive for the indication of surgery as in patients who do not suffer from CF.

MRI of the paranasal sinuses in CF patients helps to understand the differences in the development of sinusitis compared to non-CF patients. Nearly all CF patients show an abnormal mucosal swelling, but only a small part of them report nasal symptoms or develop polyposis. Another peculiarity of CF sinusitis is the primary location of the relevant pathological changes in the maxillary sinuses that frequently medialize the lateral nasal walls similar to a mucocele. Often the ethmoid cells are not affected, so that in CF a sinugenic rather than a rhinogenic origin (as in non-CF) of the sinusitis may be assumed.

Chronic sinusitis with nasal polyposis in children should always lead to further diagnostics considering systemic and genetic causes. Beside CF, those may be other genetic defects such as primary ciliary dyskinesia (PCD) or Kartagener syndrome.

MRI is not able to directly display the osseous lamellae. However, with some experience they can be easily recognized as "negative images". Usually sinus surgeons are not used to this type of imaging and preferably rely on CT scans. However, in order to avoid radiation exposure in children MRI is highly recommended as first line imaging. It is the surgeon's responsibility to decide if the anatomic situation of an individual case is sufficiently described by a certain imaging method. Generally CT scan, DVT, and MRI are not completely comparable, but all are appropriate for guiding a surgical intervention [18].

3.2.5 Case report 14

Diagnosis. Orbital complication of ethmoid sinusitis (Figure 16).



Figure 16: Case report 14. MRI in coronal sections. STIR (left): hyperintense signal in the right orbit (long arrow) crossing the

lamina papyracea which is blurred and hyperintense in comparison to the opposite side. Reconstruction of the T1 3D GRE with fat saturation in the same section (right): No contrast

enhancement in the centre of the abscess (short arrows), which could not be clearly delineated before contrast agent application. Increased enhancement of the right M. rectus

medialis.

General information. Orbital involvement is the most frequent complication of acute sinusitis in children. Appropriate imaging allows to estimate the severity of the inflammation and the likelihood of an abscess. Indication for emergency surgery is always based on a combination of imaging and clinical findings, there are several grading systems for the severity of orbital complications, however, clinically they are of limited practical use. Generally a definable pus retention (abscess) within the paranasal sinuses and especially within the orbit has to be considered as an emergency indication for a surgical intervention.

Case description. A 2³/₄ year old girl had suffered from fever and a reddened and swollen right eye for 6 days. Despite an oral antibiotic therapy periorbital swelling progressed.

Which information and specific clinical question does the radiologist need to know? Duration of clinical manifestation, initial signs and endoscopic findings are very useful in defining the grade of inflammation and complications.

Differential diagnoses and limitations of imaging. In cases of acute clinical symptoms and signs of inflammation, imaging has its role in differentiating a pre-septal from an intraorbital process and may therefore have important surgical implications. Although an abscess is generally accepted as an indication for surgery, small pus retentions of only a few millimeters might be treated successfully by intravenous antibiotic therapy. In case of absent clinical signs of inflammation, the most important differential diagnoses of intraorbital masses are hemangiomas or rhabdomyosarcomas.

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3.2.6 Case report 14a

The additional case report 14a of an intra-cranial sinugenic abscess (Figure 17) demonstrates the significance of contrast agent application also for CT and the superiority of MRI in the depiction of soft tissue pathologies.



Figure 17: Case report 14a. 7-year-old boy with swelling of the right upper lid for 2 weeks. CT (left): The coronal reconstruction of the orbit shows the thinned right lamina papyracea (figure courtesy of Prof. Hans Henkes, Department of Neuroradiology, Klinikum Stuttgart, Germany). MRI (right): The coronal STIR sequence shows an intracranial supraorbital abscess (arrows) that, even retrospectively, cannot be seen in the CT scan without contrast agent application.

3.3 Ear and temporal bone

The depiction of tiny structures of the temporal bone, especially the of the middle ear is a domain of high resolution CT. New techniques, however, promote the MRI as an increasingly indispensable part of preoperative diagnostics. On one hand, these methods allow for detailed high-resolution imaging of important structures of the inner ear and the inner auditory canal, on the other hand they enable the visualization of certain tissue properties such as restrictions of Brownian motion (diffusion). Special diffusion weighted sequences may be successfully employed even at the skull base and thus provide new options for the diagnostics of cholesteatomas. Absent radiation exposure renders it particularly appropriate for children. The next years will show if this technique allows a more specific planning of surgical interventions or may even help to avoid second-look surgeries in selected cases.

In showing the bony structures in detail, CT of the temporal bone appears to be complimentary to MRI, which directly shows the nerves of the inner meatus and the course of the facial nerve through the temporal bone. For a maximum benefit of this relatively time consuming highresolution MR techniques a clear clinical question to the radiologist and a good interdisciplinary communication are essential.

3.3.1 Case report 15

Diagnosis. Bilateral malformation of the inner ear (Figure 18).

General information. A combination of choanal atresia and bilateral combined hearing loss with malformation of the ear lead to the suspicion of CHARGE syndrome (coloboma of the eye, heart defects, atresia of the nasal choanae, retardation of growth and/or development, genital and/or urinary abnormalities, and ear abnormalities and deafness). The complete spectrum of symptoms may not be present in each patient suspected of this syndrome. Genetic counseling is recommended.

Malformations of the labyrinth become obvious during examinations of hearing disorders. If children are born deaf, imaging belongs to the standard of preparing a cochlear implant. In many centres MRI is considered primarily sufficient in cases of normal anatomy. However, in cases of developmental anomalies, the additional depiction of the bony structures by a high-resolution CT scan of the temporal bone is very helpful [9]. Even in older children a malformation may be the cause of a slowly progressive sensorineural hearing loss.

Case description. Shortly after birth, this child was noticed to have a nasal obstruction. A left side choanal atresia was found and opened several weeks after birth in order to insert a feeding tube. During the first year of life, hearing loss was diagnosed and initially considered as conductive. The child received hearing aids, which allowed a nearly normal linguistic development. Despite hearing aids, the hearing ability was unstable. Later on, in addition to the missing aeration of the tympanic cavity and the resulting conductive hearing problems a clear deficit of the inner ear function became obvious leading to a profound mixed hearing loss with a bilateral sensorineural auditory threshold of 40-50 dB. An adhesive process developed on both sides and could be stabilized on the right side. CT-imaging of the middle ear could visualize the suspected anomaly of the ossicles with a missing stapes. A subsequent MRI revealed an additional malformation of the inner ear on both sides.

Which information and specific clinical question does the radiologist need to know? Clinical characteristics of the hearing loss (conductive, sensorineural, congenital, acquired). Additional malformations.

Differential diagnoses and limitations of imaging. Highresolution T2 weighted 3D MR sequences allow the assessment of very small anatomic details of the inner ear and the nerves of the internal auditory canal. Associated cerebral anomalies may be evaluated during the same examination. Bony structures (e.g. ossicles), however, may not or only insufficiently be delineated by MRI. Therefore, there is an indication for high-resolution CT (with consideration of the radiation exposure) in special cases of suspected malformation (e.g. for the fitting of implantable hearing systems).





Figure 18: Case report 15. 1. Female patient with choanal atresia and combined hearing disorders (CHARGE syndrome). MRI: transverse MIP reconstruction of the CISS 3D: incomplete turns of the right cochlea compared to the left side (long arrows). On both sides only one rudimentary semicircular canal (short arrows).

2. Nerves of the inner meatus. Regular findings (left): facial nerve (short arrow), cochlear nerve (arrowhead), superior and inferior vestibular nerve (long arrows). Patient with choanal atresia and combined hearing loss (right): The vestibular nerve (long arrow) is abnormally thin and does not show a clear division into a superior and inferior part.

3.3.2 Case report 16

Diagnosis. Temporal bone fracture (Figure 19).



Figure 19: Case report 16. 14-year-old boy with fracture of the right temporal bone. Transverse sections of CT scan. The subtle, but diagnostically important findings of trapped air in the soft tissue medial and posterior to temporal bone (arrows) are well displayed in the CT scan (the actual fracture lines are not visible), however, both findings would not be reliably identified by MRI. (Courtesy of Prof. Hans Henkes, Department of Neuroradiology, Klinikum Stuttgart, Germany). **General information.** CT is the technique of choice for the identification or exclusion of skull base fracture. Also indirect hints on fractures such as trapped air in the soft tissue may be reliably detected.

Case description. A 14-year-old boy presented after head trauma from a 2 meters fall.

Which information and data specific clinical question does the radiologist need to know? Information about of the trauma mechanism. Liquorrhoea or a hemato-tympanon?

Differential diagnoses and limitations of imaging. On MRI low-signal fracture lines or trapped air cannot be differentiated from signal losses of other origin. High-signal fracture lines may be masked by mucosal swellings in the mastoid. Radiography is obsolete because of the inability to depict the complex structures of the skull base.

3.3.3 Case report 17

Diagnosis. Peripheral paresis of the facial nerve (Figure 20).

General information. In children, the percentage of neuroborreliosis with isolated facial paresis is particularly high. In cases of typical clinical findings and electro-





Figure 20: Case report 17. 18-year-old adolescent with recurrent peripheral paresis of the right facial nerve. T1 3D GRE with fat suppression after contrast agent application. The right facial nerve has a higher signal intensity and is thicker than the left one in the sagittal (short arrows) as well as the axial (long arrows) reconstruction. Therefore: Signs of neuritis, no tumor.

physiology, imaging is not needed. If the clinical findings and the course are not typical, MRI helps to exclude other causes such as tumors or abscesses of the temporal bone.

Case description. Recurrent unilateral right facial nerve palsy with positive borrelia titer.

Which information and specific clinical question does the radiologist need to know? Information about localization and grading of the facial nerve paresis. The probable cause as suggested by the patient's history: i.e. postoperatively, after trauma, or infection.

Differential diagnoses and limitations of imaging. Highresolution MRI can depict the course of the facial nerve in the cerebellopontine angle, the inner meatus, the middle ear, and the mastoid to its exit at the skull base. The direct delineation of the nerve may be impaired by signal artefacts of the temporal bone.

3.3.4 Case report 18

Diagnosis. Cholesteatoma with several recurrences (Figure 21).

General information. The surgical treatment of cholesteatoma in children is a major challenge because of its high recurrence rate. The long-term success significantly depends on the individual decision of the otosurgeon for or against a second-look surgery at the time of the first intervention. This second intervention is usually performed 12–18 months later and is regularly critically questioned by the parents, who ask for alternatives.

Because of its specific lamellar structure ("onion skin structure"), the cholesteatoma matrix has the typical characteristic to limit the free movement (Brownian motion) of water molecules. Modern MRI techniques, such as the diffusion-weighted HASTE sequences, make use of this diffusion restriction to identify cholesteatomas [8]. **Case description**. The child first presented at the age of 3 years because of persisting mucotympanon and was referred for adenotomy and grommets insertion. Despite tympanic drainage, a typical epitympanic cholesteatoma developed in the course of three years on the right side, and led to erosion of the malleus and the incus as well as the decomposition of the stapes superstructure. The cholesteatoma was removed during a tympanoplasty with ossiculoplasty. 18 months later a revision tympanoplasty was performed as a second-look procedure, revealing a large recurrent cholesteatoma which required the creation of a radical cavity. Another four years later, an additional intervention became necessary because of recurrent cholesteatoma in a retraction pocket. Again four years later, chronic secretion of the infected mastoid cavity required a surgical revision which uncovered an asymptomatic cholesteatoma residue in the course of the canal of the facial nerve. The latest intervention another year later revealed a very small cholesteatoma pearl within a cholesterol granuloma. MRI directly performed before surgery (Figure 21.1) detected the restricted diffusion within the cholesterol granuloma but did not distinguished the small cholesteatoma residue in the tympanum.

Which information and specific clinical question does the radiologist need to know? The most important information includes all anatomic particularities due to previous interventions, e.g. the presence of a mastoid cavity or an implanted middle ear prosthesis.

Differential diagnoses and limitations of imaging. Limitations of MRI result from additional fluid retentions in the mastoid and from cholesteatomas smaller than 4 mm, as in the case presented here. The assessment of the exact localization of the findings within the temporal bone requires 3D fusion of the coarse diffusion weighted images with the anatomically detailed T2-weighted images of a CISS sequence.

Preoperative MRI can give valuable information for the planning of revision tympanoplasty, however, it cannot replace it. Children who underwent cholesteatoma surgery need a consequent long time follow-up. For the first time, diffusion-weighted MRI allows a specific and non-invasive monitoring. Experiences with larger pediatric populations, however, are still missing.

3.3.5 Case report 18a

Diffusion-weighted MRI (HASTE) with the typical finding of a cholesteatoma (Figure 22).





Figure 21: Case report 18. 1. Only moderately intense signal in the diffusion-weighted HASTE sequence (left). Anatomic correlation in the 3D CISS sequence (right). Histology: cholesterol granuloma, not a cholesteatoma.

2. Cholesteatoma pearl (*) in the middle ear measuring 2 mm in diameter. The findings are below the detection limit of MRI. The salient area in the MR image intraoperatively corresponded to the cholesterol granuloma.



Figure 22: Case report 18a. 10-year-old boy after resection of a cholesteatoma of the left side. Typical bright signal of a cholesteatoma recurrence in the diffusion-weighted HASTE sequence (left). Anatomic correlation to the 3D CISS sequence (right). Intraoperative confirmation of the findings.

Abbreviations

CISS – Constructive Interference in Steady State (high resolution T2 weighted MR sequence) GRE – Gradient echo sequence

HASTE – Half-Fourier Acquisition Single-shot Turbo Spin Echo (here: diffusion weighted MR sequence) MIP – Maximum intensity projection STIR – Short tau inversion recovery (MR sequence with fat suppression)

T1 SE - T1 weighted spin echo (MR sequence)



Notes

Competing interests

The authors declare that they have no competing interests.

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