Neurology

Christel Vockelmann, Ursula Blum, Martina Kahl-Scholz and Guido Heilsberg

Contents

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The brain controls all important functions, from motor skills and sensory perception to vital processes such as breathing, heartbeat and digestion. It is a complicated system of neurotransmitters and neuroreceptors. The spinal cord is, so to speak, the connection between the central switching station "brain" and the other parts of the body such as the neck, trunk and extremities.

14.1 Anatomical Structures

Christel Vockelmann

The anatomical structures of the neurological system include the neurocranium with its various parts, the myelon as well as the cranial nerves (central nervous system, CNS) and the peripheral ganglia and nerves (peripheral nervous system, PNS).

The imaging of the CNS plays a major role, therefore we will limit ourselves here to the brief imaging of the brain (encephalon) and spinal cord (medulla spinalis).

The **cerebrum** (telencephalon) forms the largest part of the brain and is structurally characterized by the two hemispheres, several furrows (sulci) and convolutions (gyri). Other parts are the **diencephalon** with thalamus, subthalamus, hypothalamus, pituitary gland and epiphysis as well as the **mesencephalum**, **cerebellum**, **pons** and the **medulla oblangata**, which merges into the myelon. The boundary between the latter two structures is at about the level of the **foramen magnum**. Important in the context of imaging are the basal ganglia (also called the truncal ganglia), which include the putamen and pallidum (together Nucl. lentiformis) and Nucl. caudatus. between the aforementioned nuclei is found the capsula interna, laterally to it the capsula externa, the striatum and the capsula extrema, in each of which important pathways run.

The brain is supplied by numerous blood vessels. The four **main arteries** are the left and right internal carotid arteries and the right and left vertebral arteries. The vertebral arteries form the basilar artery, which in turn feeds the cerebral arterial circle (also known as the circle of Willis) inside the skull (frequent location of aneurysms, etc.). The term carotid T for the intracranial part of the internal carotid artery with its branching into the middle cerebral artery and anterior cerebral artery is commonly used in clinical practice and is particularly important in the acute diagnosis of stroke. The intracranial vessels are divided into segments M1 to M4 for the middle cerebral artery or P1 to P4 for the posterior cerebral artery (each to the next vessel division), and A1 (to the anterior communicating ramus) and A2 for the anterior cerebral artery.

Brain and spinal cord are surrounded by cerebrospinal fluid, which is formed by the choroid plexus.

The spinal cord is about 45 cm long and extends to the 1st/2nd LWK. Like the cerebrum, it is divided into the grey and white matter, which carry different nerve fibers. The spinal nerves, which are responsible for the nervous supply of the neck, trunk and the arms and legs, branch off from the spinal cord.

14.2 Disease Patterns

Christel Vockelmann

14.2.1 Intracranial and Spinal Hemorrhages

Intracranial and intraspinal hemorrhages are described according to their localization. **Epidural hemorrhages** can be localized both intraspinally and intracranially between the cranial bone or vertebral body and the dura mater. In the skull in particular, the cause is often a calvaria fracture, which leads to a rupture of the meningeal artery and can thus progress rapidly.

Because of its space-occupying nature and potentially rapid progression, epidural hematoma is a neurologic or neurosurgical emergency.

Subdural hemorrhages (SDH = subdural hematoma) often occur post-traumatically between the dura mater and the arachnoid with any necessary relief. In older people in particular, even a trivial trauma is sufficient to lead to rupture of the bridging veins (Fig. 14.1).

Subarachnoid hemorrhage (Fig. 14.2) can occur with aneurysm rupture.

A history of falls should not deter one from looking for an aneurysm in the basal cisterns in a SAB, as the patient may have fallen due to the aneurysm rupture.

An important sign is the accumulation of blood in the basal cisterns with punctum maximum around the aneurysm. Another cause is a post-traumatic SAB, which is then not localized basally.

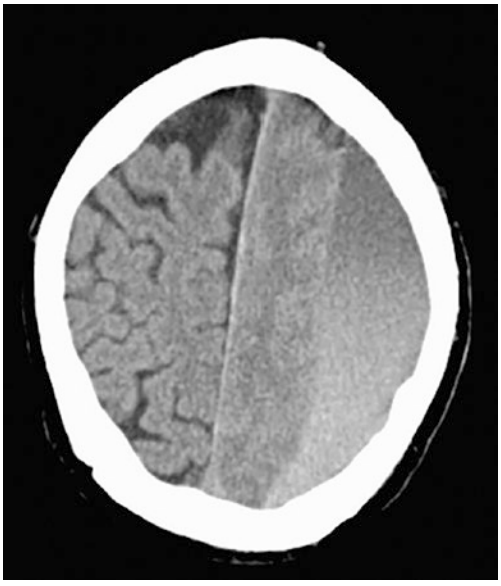

Fig. 14.1 Subdural hematoma on multiple CT scans

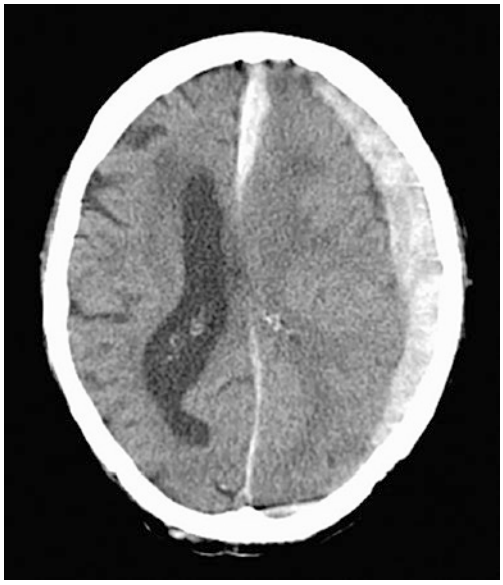

Fig. 14.2 Subarachnoid hemorrhage on CT scan

Intracerebral hemorrhage (ICB), when localized in the basal ganglia (Fig. 14.3) or pons, is usually hypertensive in origin; when localized elsewhere, reasons for ICB must be sought. Possible causes are:

- Arteriovenous malformations,
- Cavernomas,
- Intracerebral tumors or metastases,
- Sinus vein thrombosis (bleeding in the neighborhood of the thrombosed sinus).

Table 14.1 shows the main distinguishing features.

Clinic

Epidural and subdural hematomas become clinically obvious mainly because of increasing headache; a history of trauma and possibly medication with anticoagulants or antiplatelet agents suggest hemorrhage. SAB is characterized by a thunderclap headache of unknown severity. Typically, aneurysm ruptures affect younger people who report physical exertion before symptom onset. ICB results in neurological deficits similar to ischemic stroke, matching the affected portion of the neurocranium.

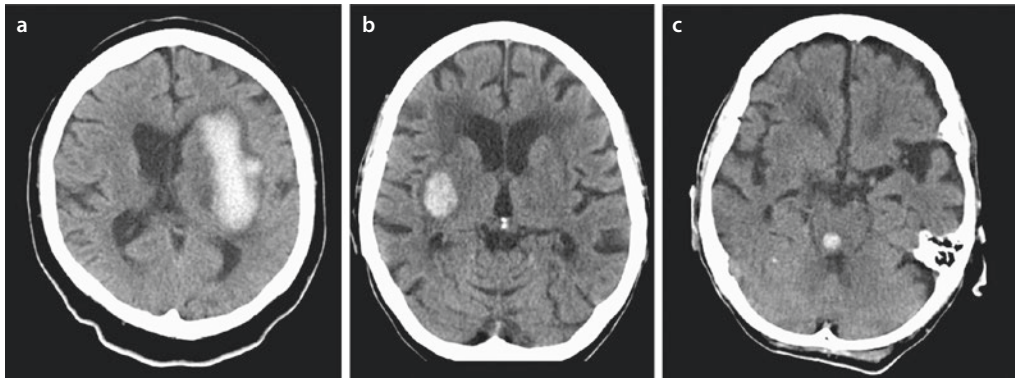

Fig. 14.3 Intracerebral hemorrhage on CT. **a** Basal ganglia on left. **b** Basal ganglia on right. **c** Pons

Table 14.1 Differentiation of hematoma localization

		Epidural Hematoma	Subdural Hematoma	Subarachnoid Hemorrhage
Anamnesis		Acute trauma	Often insidious onset with trauma that has already occurred some time ago	Thunderclap headache after physical exertion
Patients concerned		Any age	Rather older patients	Often younger patients
Localization		Often temporoparietal	Frontoparietal, often along the falx or tentorium	Basal cisterns → aneurysm rupture, parietal/occipital → rather traumatic
Form		Biconvex, does not exceed the cranial sutures, does not respect the falx	Concave crescent-shaped, exceeds the cranial sutures	Along the gyri and sulci of the brain surface

14

Diagnostics

CT

The method of first choice is the cranial CT, with which an acute hemorrhage can be sensitively detected or excluded. In case of SAB in the basal ganglia, CT angiography should be performed immediately to detect an aneurysm. With increasing duration of SAB, vascular spasms occur, which make aneurysm detection difficult or impossible.

MRI

MRI is necessary in the further work-up of atypical ICB with then blood-sensitive sequences and angiographic procedures. It is important to detect **CSF congestion**, e.g., due to dilatation of the temporal horns by entrapment or hemorrhage infiltration into the ventricular system. In these cases, neurosurgical relief must be performed.

Spinal hemorrhages are usually poorly recognizable on CT; in this case, MRI is necessary at an early stage with appropriate sequence selection (hemorrhage-sensitive sequences, T1s fat-saturated).

Intracranial hemorrhages change their characteristics on imaging over the course of days and weeks (Table 14.2). Because of the changes with T1-weighted signal enhancement on MRI, an MRI should be performed within a maximum of three days for atypical intracerebral hemorrhages to detect contrast enhancement.

Table 14.2 Temporal changes in imaging

	CT	MRI Compared to White Matter		
		T1w	T2w	T2*w
Peracute (0–24 h)	Hyperdens (cA. 50 HU)	Isointens	Slightly hyperintense	Slightly hypointense
Acute (1–3 days)	Hyperdens (cA. 50 HU)	Slightly hypointense	Greatly hypointense	Hypointens
Early subacute (3–7 days)	Slowly deflating	Strongly hyperintensive	Strongly hyperintensive	Hypointens
Late subacute (7–14 days)	Increasingly isodens	Strongly hyperintensive	Strongly hyperintensive	Hypointens
Chronic (>14 days)	Hypodense to liquorisodense. If applicable calcifications	Central isointense, slightly hypointense rim	Central slightly hyperintense, margins strongly hypointense	Greatly hypointense

14.2.2 Ischemic Diseases

A lack of blood supply to the brain is the most frequent cause of **stroke**, accounting for about 70%. The incidence is 130/1,00,000 inhabitants. After myocardial infarction and tumor disease, stroke is the third most frequent cause of death. The cause is often arteriosclerotic vascular disease of the extracranial vessels supplying the brain, followed by embolic occlusions. In younger patients, inflammatory changes of the vessels or sinus vein thrombosis may lead to stroke.

Ischemic myelon infarction is rare overall. A typical cause may be aortic dissection. Myelon infarction is feared as a complication after surgical or interventional procedures on the aorta.

Clinic

A classic symptom is brachiofacial hemiparesis of the contralateral half of the body. Dizziness and visual disturbances or cranial nerve failures may also be symptoms of an acute stroke. Prodromes are, for example, amaurosis fugax or TIA symptoms, i.e. a transient ischemic attack.

Diagnostics

CT

An acute stroke is, comparable to a heart attack, an absolute emergency that requires immediate imaging. In this case, the native cranial CT is the first elementary component due to its speed and high availability. If there is no hemorrhage (hyperdens with density values around cA. 50 HU) and no demarcated infarct (hypodense, missing gray-white differentiation, edema), an intravenous lysis therapy is started directly in the appropriate clinic. Time is brain! Every minute saved improves the patient's prognosis.

Perfusion Imaging

The native CT is usually supplemented by perfusion imaging (section computed tomography) with which large infarct areas that can no longer be saved even by immediate therapy and areas that are threatened but can still be saved can be detected, even if no infarct is yet demarcated in the native image.

CT Angiography

The third pillar is CT angiography to find intra- or extracranial occlusions. Acute thrombotic occlusion of the middle cerebral artery or internal carotid artery is recana-

lized by interventional techniques similar to those used in myocardial infarction (Sect. 14.4.1).

MRI

Diagnosis of ischemic diseases of the neurocranium and myelon is performed in MRI. The classic constellation here is **signal enhancement in diffusion weighting** with **signal depression in ADC**, which is already present in the peracute stage. With increasing time, **edema with signal enhancement in T2w sequences** then develops (Fig. 14.4).

This edema has its peak approximately between the 3rd to 5th day. In the further course, the necrosis zone is organized with glioses and cystic formations. This process can be well traced on imaging with a regressing diffusion disorder and increasing glioses (CT: hypodense to the parenchyma; MRI: hyperintense in the FLAIR, T1 hypointense) and cystic formations (CT and MRI liquorisodense and -isointense, respectively, Fig. 14.4).

The localization and extent of an infarct allow conclusions to be drawn about its genesis. Thus, territorial (embolic) infarcts are assigned to the supply area of the intracranial vessels. Border zone infarcts are localized in the transition zones between the supply areas and are hemodynamically caused. Lacunar infarcts are of microangiopathic origin (Fig. 14.5).

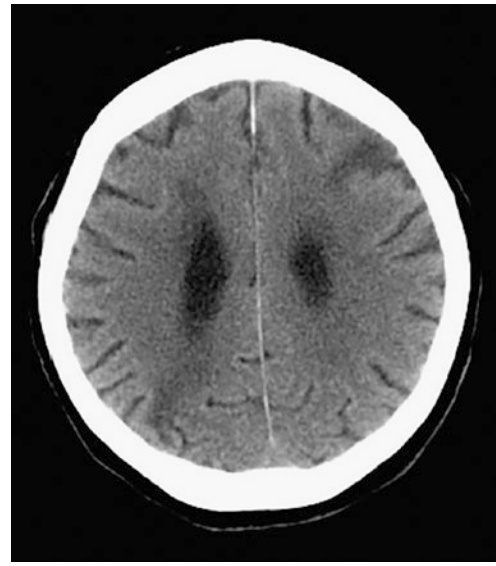

Fig. 14.5 Anterior border zone infarct on the left and posterior border zone infarct on the right in the native CT scan

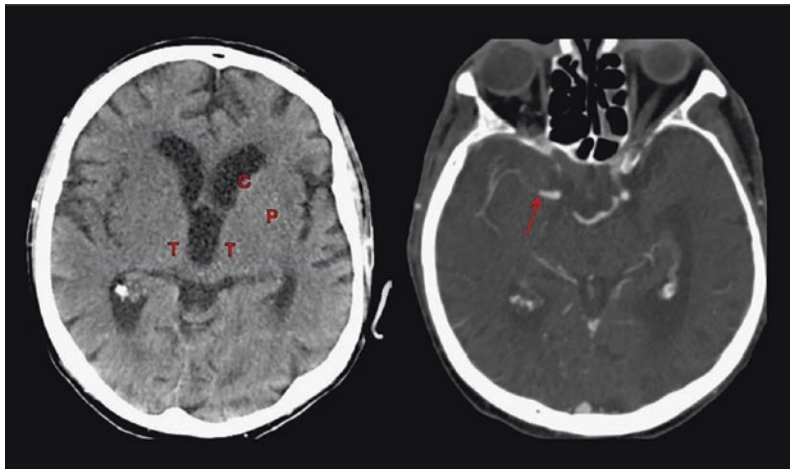

Fig. 14.4 CT-native and CT-A with blunted basal ganglia (caput nucleus caudatus and putamen/pallidum on the left with occlusion of the middle cerebral artery in the M1 segment—arrow)

14.2.3 Intracerebral Tumors

The classification of brain tumors is based on the WHO classification (Table 14.3). This reflects the degree of malignancy of the tumors.

In addition, there are other tumors, some of which have typical localizations and age peaks. In the end, the clear image-morphological assignment is often not successful.

In adults, **metastases** occur more frequently than brain tumors (Fig. 14.7). Here, in addition to intracerebral metastasis, **meningeal carcinomatosis** or **intramedullary metastasis** is increasingly common. Characteristic imaging shows melanoma metastases and frequently also metastases of renal cell carcinoma, both of which can be delineated **hyperdense** on **CT** and **native T1-weighted hyperintense** on **MRI.**

Table 14.3 Classification of brain tumors according to WHO

Grade	Description	Example
I	Benign tumors that can potentially be cured by surgical removal	Craniopharyngeoma, pilocytic astrocytoma or neurinomas and schwannomas
II	Infiltrative tumors, histologically benign, frequently recurrent	Oligodendroglioma, diffuse growing astrocytoma
III	Malignant tumors with reduction of survival time	Anaplastic astrocytoma, plexus carcinoma
IV	Very malignant tumors with a significant reduction in survival time	Glioblastoma (Fig. 14.6), medulloblastoma

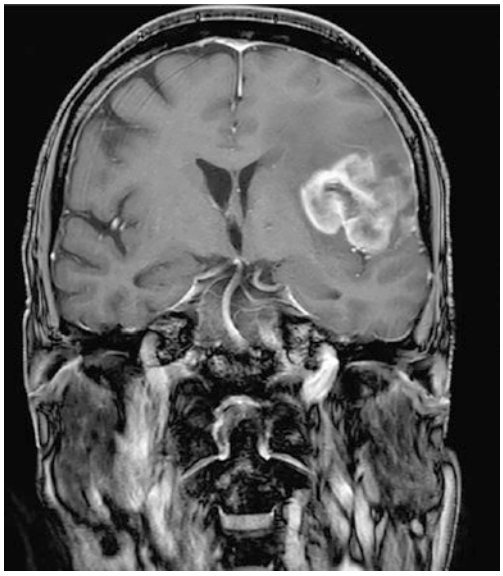

Fig. 14.6 Glioblastoma (coronary T1, contrast enhanced)

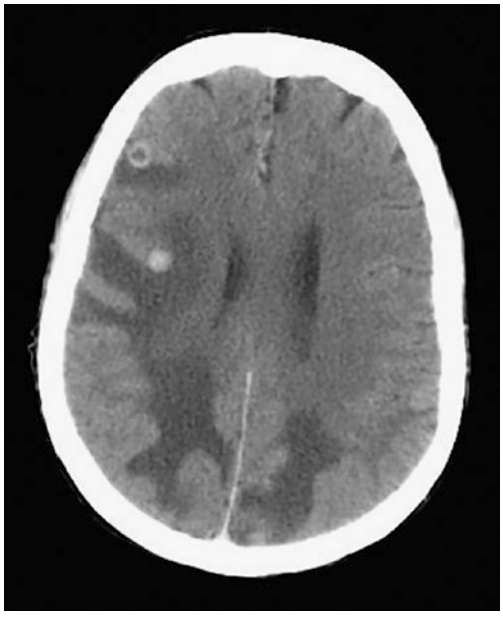

Fig. 14.7 Cerebral metastases bds. with annular enhancement and finger-shaped edema in contrast-enhanced CT

Clinic

The symptomatology of cerebral masses depends on the localization of the finding.

For example, a mass in the frontal brain may be accompanied by a change in the patient's personality. Other symptoms are stroke-like symptoms or a seizure.

Cerebral metastasis can also be the first symptomatic manifestation of a tumor, and the first look should then be at the lung as the most common organ of origin.

In contrast, in the absence of a tumor history, a primary tumor is more likely to be assumed in the case of a myelon mass; if myelon metastases occur, the tumor is usually already known.

Diagnostics

CT

Often the first diagnosis is a cranial CT scan due to stroke-like symptoms or a seizure. Here, a **hypodense "finger-shaped" edema** can be detected, which mostly respects the cortex. The space-occupying character can be delimited by a **constriction of the cerebrospinal fluid spaces.**

MRI

Further imaging then requires the administration of a contrast agent, although this can also be performed as part of a complementary MRI diagnosis. The **finger-shaped edema** can also be delineated on MRI. To allow contrast passage through the blood-brain barrier, imaging should be performed no earlier than 5 min after contrast administration. On the basis of the localization, the age of the patient, any calcifications in the CT and the contrast medium accumulation, a tentative diagnosis of the type of mass can be made. If necessary, this can be reinforced by MR spectroscopy, but ultimately a definite statement about the type of tumor is not always possible. Cerebral metastasis is indicated by the presence of several contrast-enhancing lesions.

An important differential diagnosis (Table 14.4) to intracerebral tumor is **abscess**, which is classically characterized by marked **hyperintense signaling** in the diffusion.

Signal enhancement of a space involvement in the diffusion weighting is indicative of an intracerebral abscess!

Table 14.4 DD of selected brain tumors

	Frequent localization	Frequency of all primary brain tumors	Age and gender distribution	Tumor grading	Imaging
Astrocytoma	Supratentorial	9%	30–60 LJ M > w	II + III	With increasing de-differentiation, increasing KM enhancement, surrounding finger-shaped edema
Glioblastoma multiforme (=astrocytoma grade IV)	Cerebral hemispheres, bars (butterfly glioblastoma)	20%	50TH–70TH LJ M > w	IV	Severe KM enhancement, garland-shaped, extensive necrosis and hemorrhage
Oligodendroglioma	Frontal brain, basal ganglia	4%	40TH–60TH LJ M > w	II + III	In 2/3 of the cases, extensive scaly calcifications, little surrounding edema, KM uptake depending on the grading (II or III)
Ependymoma	Proximity to the ventricular system	<0.5%	Children and adolescents, 30–40 LJ	Variable	Inhomogeneous KM enhancement, no perifocal edema; caution: drip metastases
Primary CNS lymphoma	–	Rarely	For immunocompetence 50–60th year, for immunoincompetence earlier		On CT often hyperdense due to cell richness, on MRI isodense in T1w and T2w with homogeneous KM enhancement
Pilocytic astrocytoma	Infratentorial	0.3%	1st-2nd cent of life; m > w		Large cystic tumor portion with vigorous KM-absorbing node
Medulloblastoma	Infratentorial	Rarely	1st decade of life	IV	Inhomogeneous tumor of the cerebellum, hemorrhages, frequent infiltration of the ventricular system, drip metastases
Colloid cyst	Third ventricle, foramen monroi…	<2%	Mostly between 20 and 40 LJ, m > w	Benign	CT: Hyperdense, smooth bordered in 3rd ventricle; variable on MR, T1w usually hyperintense, usually no enhancement

14.2.4 Cerebrospinal Fluid Circulation Disorder

Common to all cerebral space-occupying lesions is the risk of cerebrospinal fluid circulation disturbance, i.e. congestion of the ventricular system. This can be caused supra as well as infratentorially by an entrapment of the parenchyma at the tentorium, falx or foramen magnum. Obstruction of interventricular foramen or aqueduct in hemorrhage also results in CSF circulatory obstruction.

Clinic

The classic symptom is headache. Often the examination of the fundus of the eye reveals a congestion papilla.

Diagnostics

CT/MRI

The most important imaging procedure is the CT. Further clarification takes place in the MRI. In both procedures, the **widening of the cerebrospinal fluid spaces** and, if necessary, the cause for this can be detected.

An early sign of a cerebrospinal fluid circulation disorder is a widening (>3 mm) of the temporal horns.

14.2.5 Intracranial Extraaxial Tumors

In addition to cerebral tumors (synonym: intraaxial tumor), meningioma is a frequent finding in neurocranial imaging. This usually benign tumor originates from the meninges and displaces the adjacent brain parenchyma.

Other extraaxial rare tumors are the epidermoid, schwannomas especially of the vestibulocochlear nerve ("acoustic neuroma") or tumors of the pituitary gland.

Clinic

Mostly incidental finding. Symptoms usually occur only with very large meningiomas. Acoustic neuroma is characterized by tinnitus, dizziness and a disturbance in sound perception.

Diagnostics

CT

Classical is besides the convex contour to the cranial dome with a protrusion to the dura, the so-called **dural tail**, a very strong, early and homogeneous contrast enhancement. The blood supply of a meningioma is via branches of the external carotid artery, and the contrast enhancement does not have to cross the blood-brain barrier. In addition, meningiomas are frequently calcified.

Meningiomas can occur anywhere on the meninges, although spinal meningiomas are very rare (Fig. 14.8).

A special case of meningioma is malignant meningioma, which exerts pressure on the adjacent brain parenchyma and becomes symptomatic accordingly.

Acoustic neuromas show a strong contrast enhancement of the partly very small tumors. Larger findings may lead to a widening of the internal acoustic meatus with an "ice cream cone"-like configuration of the tumor.

In the case of space occupying lesions in the cerebellopontine angle, three diagnoses must be considered: meningioma, acoustic neuroma (=wannoma) and epidermoid tumor. The differential diagnosis is made in MRI with KM sequences and diffusion weighting (the epidermoid shows a strong diffusion restriction with signal enhancement).

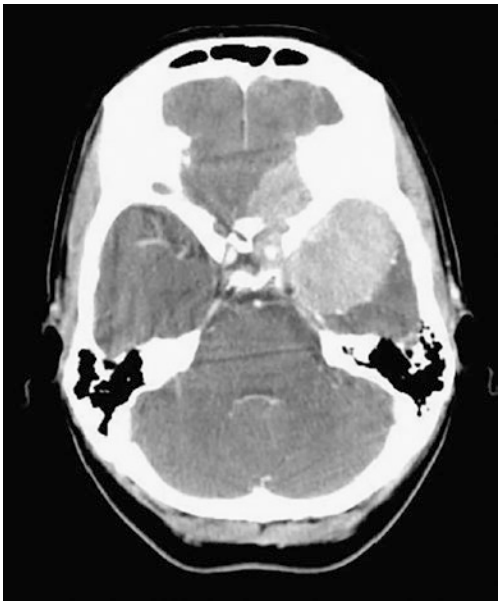

Fig. 14.8 Cuneiform wing meningioma on the left with strong contrast enhancement on CT

14.2.6 Cystic Intracranial Lesions

As with tumors, it must first be decided whether the finding is intrecerebral or intracranial but not originating in the brain. The most common lesion is the **arachnoid cyst**, a cyst often in the middle cranial fossa that displaces the brain but is not space-occupying. More rarely, cysts are intracerebral, such as a **neuroepithelial cyst**. In this case, the presence of a cystic mass or a cerebral infection must always be considered in the differential diagnosis. **Virchow-Robin spaces** are protrusions of the subarachnoid space around vessels, which appear as intracranial cysts. The most frequent localization is the basal ganglia, where the cysts develop along the lenticulostriatal branches of the middle cerebral artery. Differential diagnosis is a lacunar infarction in the basal ganglia.

▪ Clinic

Most often it is an asymptomatic incidental finding. Large arachnoid cysts may occasionally cause intracranial pressure symptoms.

▪ Diagnostics

CT/MRI

CT and MRI with liquorisodense and -isointense imaging, respectively. The arachnoid cyst displaces the adjacent brain parenchyma without signal alterations. No contrast enhancement.

Neuroepithelial cysts are often located supratentorially adjacent to the lateral ventricles.

14.2.7 Chronic Inflammatory CNS Processes: Multiple Sclerosis

The most common chronic inflammatory CNS disease is multiple sclerosis (MS). Often optic neuritis is the first symptom with no pathological findings on imaging. MS is a demyelinating disease in which MS plaque forms with destruction of myelin due to etiologically unclear inflammation. The disease mostly affects younger patients between 20 and 40 years of age. In most cases, the disease progresses in relapses. Glucocorticoids are used as therapy during the relapse.

▪ Clinic

In about 1/4 of the cases, optic neuritis ("The patient sees nothing, the doctor sees nothing!") is the initial symptom. Sensory disturbances, weakness of the extremities or even a loss of sensitivity in the supply area of the trigeminal nerve are further symptoms.

▪▪ Diagnostics

CT

In addition to imaging, CSF puncture is obligatory in the suspected diagnosis of MS. The CT shows **hypodense areas of** the white matter only in advanced disease, which are not distinguishable from a pronounced microangiopathic damage of the brain.

MRI

MRI of the neuroaxis shows typical changes, but imaging alone cannot prove MS. Typical findings are **highly oval T2w**

hyperintense demyelinating foci periventricularly, which then show a **cockscomb-like appearance in** the sagittal image. Other typical findings include **juxtacortically located lesions** and distribution **supra- and infratentorially** (this includes a spinal manifestation). The lesions are **T1w frequently hypointense** (so-called "black holes") (Fig. 14.9). Temporal dissemination (corresponding to the relapsing course of the disease) can be evidenced by KM-receiving lesions (typically not completely annular but horseshoe-shaped) or new lesions at least one month after symptomatology (Table 14.5).

An important differential diagnosis in a clinic similar to MS is acute disseminated encephalomyelitis (ADEM), which causes similar lesions in the CNS, but here all lesions have the same stage.

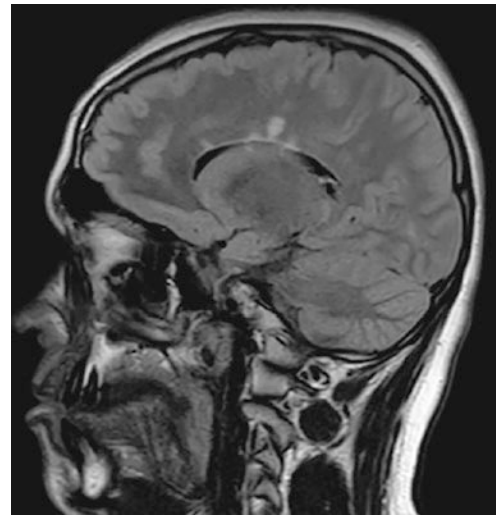

Fig. 14.9 Sagittal flair with hyperintense demyelinating foci around the corpus callosum

Table 14.5 McDonald criteria for spatial dissemination (at least three criteria for radiological diagnosis)

At least nine 2w hyperintense lesions	At least one infratentorial lesion (also located in the myelon)	At least one juxtacortical lesion	At least three periventricular lesions and evidence of temporal dissemination
Dissemination over time			
KM-absorbing focus at least three months after initial symptoms		Evidence of a new lesion T2w, at least 30 days after symptomatology	

14.2.8 Acute Inflammatory CNS Processes

This includes the following processes:

- **ADEM**: Rare disease with symptoms typical of MS, but monophasic.
- **Encephalitis**: Infection of the brain with various pathogens. Occurs frequently as meningoencephalitis derived from inflammatory processes, e.g. of the sinuses or mastoids. Typical manifestation in herpes encephalitis (type I, herpes simplex) in the temporal lobes and the limbic system. Important differential diagnosis for this is paraneoplastic limbic encephalitis with a similar pattern of distribution. This occurs most frequently in bronchial carcinoma as the primary tumor.
- **Abscess**: circumscribed melting inflammatory process, often derived from sinusitis or due to hematogenous dissemination. Common in immunocompromised patients.

Ultimately, almost all pathogens can also affect the CNS. These include, for example, **neurocysticercosis** due to infestation with the pork tapeworm, **toxoplasmosis** and **tuberculosis**. It is important to assess all clinical and imaging findings in order to arrive at the correct diagnosis.

Clinic

The classic symptom of meningitis is neck stiffness combined with headache. Other symptoms depend on the severity and localization of the disease. Immunocompromised patients in particular are more frequently affected by encephalitis.

Diagnostics

MRI (Fig. 14.10**)**

In the case of encephalitis or meningoencephalitis, the cerebrospinal fluid (CSF) puncture is a very important diagnostic tool for detecting cells and possibly also a germ in the CSF. Imaging often shows normal findings on MRI in encephalitis. If changes are detectable, it is often **edema with signal elevations T2w** and possibly **KM enhancement**. If there are corresponding changes in the limbic system, herpes simplex encephalitis or limbic encephalitis must be considered. Parasitic infections often show **calcifications** in later stages. A cerebral abscess shows a **marginal KM enhancement** with **central fluid**. For the differential diagnostic differentiation from a tumor the diffusion is important, which shows a strong diffusion restriction with a strongly hyperintense signal.

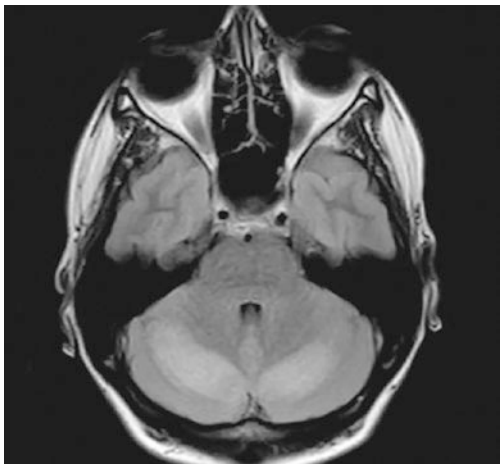

Fig. 14.10 Cerebellitis with hyperintense, relatively symmetrical signal elevations in the flair in the cerebellum bds

14.2.9 Epilepsy

Epilepsy is characterized by the repeated occurrence of seizures. If these seizures are primarily generalized and affect the whole brain, there are often genetic causes. In the case of focally initiated seizures, there is often local damage to the brain. Therefore, it is important to know the possible localization of the damage during imaging.

Tumors, infarcts or bleeding can be the cause of epilepsy. More difficult to detect are anlage disorders. The most common are **heterotopia**, i.e. scattered grey matter that has not migrated to the cortex, and **focal dysplasia**, a developmental disorder with a blurring of the medullary-cortical boundary or disturbances in the grey matter. Another disorder in the setting of epilepsy is **hippocampal sclerosis** (also known as ammonic horn sclerosis). Etiologically unclear, the disease leads to nerve cell destruction of the hippocampus.

Clinic

Depending on the location of the damage, the picture of epilepsy is very diverse. From the classic seizure with twitching of extremities to sensory disturbances and absences, other symptoms can also occur in the course of a seizure.

Diagnostics

CT

CT is the rapidly available method for the first seizure that can rule out bleeding or a tumor as the cause.

MRI

Further imaging takes place in the MRI. Causes, such as old infarcts or tumors, are often already clear after computed tomography. In the case of malformations such as heterotopia or focal dysplasia, thin-slice sequences are required both T1w and as flair sequences in order to be able to detect the malformation here.

Hippocampal sclerosis can be well delineated in coronal images as a **reduction in volume of a hippocampus** with **enlargement of the adjacent temporal horn. In** addition, there is a **signal enhancement in T2w or better flair sequences**.

14.2.10 Phacomatoses

Phacomatoses are neurocutaneous syndromes, i.e. diseases involving the skin and nervous system. They are based on certain genetic defects, which are often inherited in an autosomal-dominant manner. The most common is neurofibromatosis type 1 Recklinghausen's disease). Typical are café-au-lait spots of the skin and neurofibromas, especially optic gliomas. In neurofibromatosis type 2, the leading feature is acousticus schwannomas. Bilateral acoustic schwannomas are sufficient to establish a diagnosis of neurofibromatosis type 2. Tuberous sclerosis (Bourneville-Pringle disease) also belongs to the phacomatoses. Classically, subependymal nodules, cortical tuberosities, hypomelanotic patches of the skin as well as angiomyolipomas of the kidney and also rhabdomyomas of the heart as well as giant cell astrocytomas are found. These are mostly located in the area of the foramina monroi and belong to WHO grade I tumors. Sturge-Weber syndrome is an angiomatosis, mainly in the supply area of the trigeminal nerve. Such angiomas are also found intracerebrally. Another phacomatosis is Von Hippel-Lindau disease. The disease is characterized by cerebral hemangioblastomas. In addition, renal cell carcinomas, but also renal cysts, pheochromocytomas and cystadenomas of the testis occur frequently in these patients.

Clinic

Neurofibromatosis type 1 and also Sturge-Weber syndrome and tuberous sclerosis are usually manifested in childhood. Neurofibromatosis type 2 and Von Hippel-Lindau disease are often diagnosed in young adulthood. The clinic is characterized by the skin changes and the corresponding affected structures of the nervous system.

Diagnostics

MRI

Neurofibromas, which include optic gliomas and acoustic schwannomas, can be **homogeneously** delineated on MRI **with a strong KM enhancement.** Large acoustic neuromas lead to a **dilatation of the internal**

acoustic meatus. The image occasionally resembles an ice cream cone in large acoustic neuromas.

14.2.11 Neurodegenerative Diseases

The brain is subject to a natural aging process with a degradation of brain substance as well as iron deposition in the basal ganglia. However, certain neurodegenerative diseases present a relatively typical pattern of findings on MRI, so that MRI is performed as standard for the clarification of neurodegenerative diseases (Fig. 14.11). Important differential diagnostic criteria are shown in Table 14.6.

Every dementia should be clarified once with a sectional image diagnosis of the neurocranium, in order to exclude e.g. tumors or a normal pressure hydrocephalus as a cause.

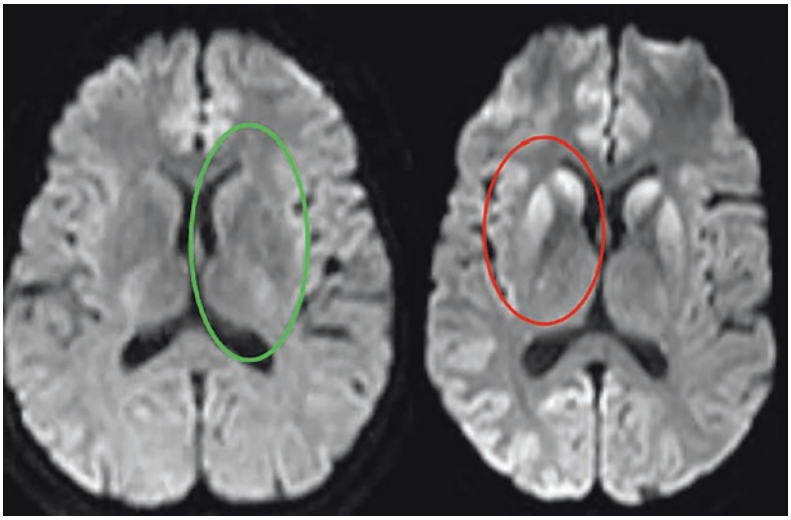

Fig. 14.11 Diffusion enhancement (right image) in the basal ganglia in Creutzfeldt-Jakob disease, for comparison normal findings on the left

Table 14.6 Differential diagnosis of different neurodegenerative diseases

	Clinic	Diagnostics
M. Alzheimer	Short-term memory impairment, no delirium, existing > six months, onset at mean 70th year	Temporally accentuated atrophy, atrophy of the hippocampus
Vascular dementia	Abrupt onset, history of stroke, other neurological deficits	Pronounced microangiopathy, infarcts with involvement of both thalami and/or temporomesial structures
Multisystem atrophy	Parkinson-like symptoms, additional e.g. orthostatic dysregulation, micturition disorder, impotence	Atrophy putamen and/or olivopontocerebellar, hot cross bun sign (hyperintense cross figure in bridge foot in T2w)
Frontotemporal lobar degeneration	Behavioral problems, change of character	Unilateral frontotemporal atrophy

(continued)

Table 14.6 (continued)

	Clinic	Diagnostics
Normal pressure hydrocephalus	Triad of gait disorder, dementia and urinary incontinence	Dilatation of the ventricular system; sample liquor puncture
M. Parkinson	Onset with unilateral rigor, tremor and hypokinesia	Mostly normal findings; possibly changes in the iron content of the substantia nigra, atrophy of the hippocampus
Creutzfeld-Jakob disease	Rapid dementia, myoclonia…	Flair and diffusion with signal enhancement of the basal ganglia and/or cortex
Progressive supranuclear paralysis	Parkinson's-like symptoms	Mesencephalic atopy (Mickey mouse character)
Huntington's chorea	Excessive movements (choreatic hyperkinesia)	Bilateral atrophy of the nucl. caudatus (coronary T1w), also putamen and globus pallidus
Wernicke's encephalopathy	Brain-organic psychosyndrome, unsteadiness of gait and stance, eye movement disorders	Atrophy or KM uptake of the corpora mamillaria

14.3 Diagnostics

14.3.1 Diagnostic Radiology

Sonography

Sonography is used in newborns as the primary diagnostic tool for assessing the neurocranium. In adults, access for the sound waves through the cranial dome is very limited. Here, sonography is used as transcranial Doppler sonography for the assessment of the intracranial vessels, often supplemented by CT or MR angiography.

Conventional X-ray Diagnostics

Conventional X-ray diagnostics of the brain skull are no longer performed. One of the last indications is the position control of a valve in certain VP shunts before or after MRI examinations.

Fluoroscopy/Angiography

Angiographic examinations are used to clarify intracerebral vascular processes such as AV malformations or aneurysms and, increasingly, to treat these interventions. Coils, i.e. small metal spirals, are used to fill aneurysms.

Computed Tomography (CT)

CT is the method of choice for almost all emergency indications for imaging of the neurocranium. CT can reliably detect or rule out hemorrhage. Also, most tumors that have become symptomatic can already be detected with a CT. Since the lenses of the eye are very sensitive to radiation, the layers in the CT of the head are angulated at the base of the skull. This ensures that the lenses of the eye are not in the direct radiation field.

In acute stroke, a so-called lysis protocol is generally used today. First, a native CT of the head is performed. After exclusion of hemorrhage, intravenous lysis therapy can be initiated if indicated. In addition, a CT perfusion and a CT angiography of the supra-aortic vessels are performed. In the case of large infarctions, the CT perfusion can be used to estimate how much infarction is imminent and how much tissue can still be

saved. This is achieved by evaluating repeated images of the brain during a contrast medium run, which allows parameters such as blood volume and blood flow in the various parts of the brain to be calculated. CT angiography then reveals stenoses or occlusions of the vessels so that, if necessary, therapy can also be initiated immediately with interventional reopening of the vessels.

Magnetic Resonance Imaging (MRI)

MRI with its high soft tissue contrast is ideally suited for clarifying the neurocranium. Here, the examination protocol, which always consists of several sequences, must be adapted to the question. When asking for fresh infarcts, a diffusion measurement must be performed. For the question of old infarctions, T2w sequences are performed, which are susceptible to susceptibility artifacts (T2* or SWI). Sagittal flair sequences, with which the configuration of the demyelinating foci can be well delineated, are helpful in the clarification of MS. Contrast medium, which is detectable in T1w sequences, is required for the clarification of tumors and metastases, but also inflammatory processes.

14.3.2 Nuclear Medicine

Ursula Blum

Brain

Blood Flow, Regional Cerebral Blood Flow (RCBF)

Regional cerebral blood flow (rCBF) can be visualized using various tracers. The questions range from focal imaging in epilepsy, psychiatric diseases, a statement about the perfusion reserve of the brain in previous TIA's to brain death diagnostics.

Different areas of the brain are of particular interest for different questions, e.g. the search for a focus in epileptic seizures or the identification of the speech or visual center before a planned operation.

There are ^{99m}Tc-ECD (ethyl cysteinate dimer) and ^{99m}Tc-HMPAO (hexamethylpropenylene aminooxime) available. Both substances are almost equivalent.

ECD is used in questions about surviving brain tissue after a cerebral infarction because it does not accumulate in brain cells that are still perfused but dead. Another domain of ECD is inflammation diagnostics. In addition, the contrast between gray and white brain matter, especially in the temporal lobe, is better than with HMPAO.

Other possibilities for the determination and visualization of rCBF lie in PET.

For this purpose, ^{15}O labelled water ($H_2{}^{15}O$) or ^{15}O-butanol can be used. However, these products have a very short half-life of 2 min, so that a cyclotron must be available in the immediate vicinity.

Since rCBF is associated with regional sugar metabolism, ^{18}F-FDG can also be used.

Normal findings depend on the age of the patient. Normally, the accumulation in the gray matter is 2–3 times higher than in the white matter.

In **epilepsy diagnostics,** the radiopharmaceutical is injected during the seizure (under EEG monitoring, if possible within 10 s), and the image is then recorded in the phase after the seizure. During the seizure, the focus is stronger than the surrounding brain tissue (hyperperfused), while in the seizure-free interval it is usually weakened (hypoperfused).

In the case of **reduced** cerebral perfusion in the context of a TIA (transient ischemic attack) or a PRIND (prolonged reversible ischemic neurological deficit), there are usually no conspicuous findings in cross-sectional imaging. In the perfusion examination, a reduction in cerebral perfusion in the affected section of the brain can already be detected.

If vasoconstriction in the brain is suspected, the examination can be performed with additional application of Diamox (1000 mg over 5 min i. v.). This reveals the so-called perfusion reserve of the brain. Diamox dilates the arterioles, and perfusion therefore increases significantly in a healthy state of the vessels. If vasoconstriction is present, the vessels are already maximally dilated, so that increased perfusion cannot occur in these regions. The examination should be performed at the earliest 24 h after a basic scintigraphy.

Dementia is characterized by reduced blood flow in different regions of the brain.

Forms of Dementia, Localization in the Cerebrum

– Alzheimer's dementia:	Bitemporal and/or biparietal
– Lewy body dementia:	Similar to Alzheimer's disease, additional visual cortex
– Fronto-temporal dementia:	Frontotemporal
– Vascular dementia:	Multiple circumscribed defects

In **brain death diagnostics** (Fig. 14.12), the patient is placed directly on the camera and injected. This is followed by the creation

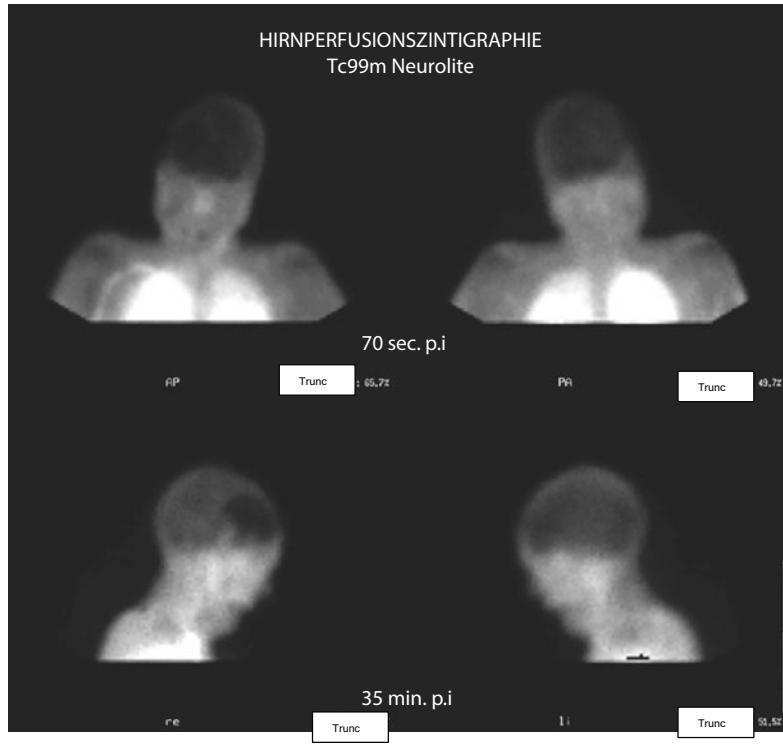

Fig. 14.12 Brain death diagnosis

of a dynamic sequence, as well as static images of the skull, thorax (including thyroid and stomach) and, if necessary, a SPECT.

In these patients, the blood-brain barrier is non-functional. There is no accumulation in the brain at all.

The marking must be checked for quality. This check should be carried out according to the kit manufacturer's specifications (e.g. thin layer chromatography).

The images of the thorax must not show any major accumulations in the thyroid gland or stomach. These images also serve as a quality control, but are not sufficient as the sole quality control.

Neurodegenerative Diseases

▪ Receptor Scintigraphy

Receptor scintigraphy is used in the diagnosis of Parkinson's disease. Especially in the early stages of the disease, clinical differentiation from other diseases may be difficult. Receptor scintigraphy can be used to distinguish idiopathic Parkinson's disease (PD) from atypical PD syndromes such as multisystem atrophy (MSA), progressive supranuclear gaze palsy (PSP), and corticobasal degeneration (CBD). This distinction is important for the treatment of the disease and the prognosis of the patient.

SPECT tracers such as ^{123}I-benzamide (IBZM) and ^{123}I-ioflupane (FP-CIT), which are commercially available in Germany, are used.

Another decision option is the determination of rCBF by means of ^{18}F-FDG-PET, which can accurately separate the diseases in one examination (Hellwig, Meyer).

PET tracers are ^{18}F-DOPA and ^{18}F-Fallypride.

The tracers differ in the binding site on the receptor system.

Brain Tumors

Brain tumors (gliomas) are characterized by an increase in amino acid transporters. These can be visualized with radioactively labeled amino acids.

Three tracers are currently used, ^{11}C-methionine (MET), 3-^{123}I-iodine-α-methyl-L-tyrosine (IMT) and ^{18}F-ethyltyrosine (FET). All tracers have not yet been approved in Germany.

Indications include biopsy planning, determination of the exact extent of the tumor, therapy monitoring and recurrence diagnosis (differentiation between necrosis and tumor).

A statement on the degree of malignancy is not possible. The differentiation between tumor and non-malignant changes or radionecrosis can be made by establishing quotients. Here, the uptake in the tumor tissue is significantly higher than in the other tissues (depending on the tracer cA. 1.5 to 2.2 times increased), in the context of therapy control, the quotient should fall by >10%.

▪ Meningiomas

Due to the somatostatin receptors, which are detectable in many meningiomas, these changes can be detected with a somatostatin receptor scintigraphy (111-In-Oxin). Here, the differential diagnosis to an acoustic neuroma is in the foreground.

CSF Space

CSF scintigraphy allows statements to be made about the distribution, circulation, and any leaks or fistulas that may be present. In addition, a statement about the function of existing shunt systems is also possible.

CSF scintigraphy is performed after sterile puncture of the CSF space, usually the lumbar region. Only very rarely is the punc-

ture performed suboccipitally. The patient should remain in bed after the puncture.

If leakage is suspected, tamponade of the nasal cavity is necessary. The tamponades must be weighed before insertion into the nose and after removal. After 24 h the tamponade is removed or changed, here the side indication is very important. The tamponades are measured in the borehole. In addition, a blood sample must be taken in each case, as some of the tracer enters the tamponades even without a fistula. Now the tamponade activity/blood activity quotient is calculated. If this quotient is >2, a fistula is present.

14.3.3 Valence

Christel Vockelmann

Table 14.7 shows the use of the respective therapeutic options depending on the problem

Table 14.7 Value of the therapeutic procedures

	Sonography	Conventional	Fluoroscopy/ Angiography	CT	MRI	Nuk	PET
Acute stroke	N	N	W	P	W	N	N
Dementia assessment	N	N	N	P	W	W	N
Brain death diagnostics	N	N	W	N*	W	P	N

N Not indicated, *P* Primary diagnosis, *W* Further diagnosis
N* Of course, every patient who is diagnosed with brain death will receive a computer tomography of the skull. However, this cannot be used to diagnose brain death

14.4 Therapy

14.4.1 Interventional Radiology

Christel Vockelmann

Angiographic Interventions

In acute occlusions of the proximal middle cerebral artery with corresponding stroke symptoms, the thrombus that led to the vessel occlusion is removed with special suction catheters and stent retrievers. For this purpose, the internal carotid artery of the affected side is probed via a femoral access route. Via a long sluice, i.e. a working channel, the occluded vessel is visited and probed with a microcatheter. The thrombus is then removed by aspiration. Alternatively or complementarily, the vessel is first released by stent implantation. In this case, the thrombus is initially only pushed to the side. After a few minutes, the inserted stent, in which the thrombus has then lodged, is removed together with the thrombus.

Symptomatic stenoses of the internal carotid artery, which are typically located close to the origin, should be treated within 14 days after the initial event such as a TIA, since the risk of a further event such as a large infarction in the area supplied by the middle cerebral artery is significantly increased. The primary procedure is surgery of the stenosis, indications for interventional therapy by means of stent angioplasty are restenosis after surgery, postradiogenic stenosis or unfavorable anatomical conditions such as a very short neck or a high division of the carotid artery. For **stent angioplasty,** a transfemoral approach with insertion of a long sheath into the common carotid artery is also performed. Often, a wire-guided filter system is then first inserted into the internal carotid artery above the stenosis to prevent possible intracranial emboli caused by detached plaque materials during the course of the intervention. The filter wire is then usually used for pre-dilatation before a stent is inserted. This is inserted as a bifurcation-bridging stent from the ACI to the ACC, with the external carotid artery being stented over (Fig. 14.13). After postdilation, the filter is then recaptured via a retrieval system and the procedure can be terminated. To avoid a vasovagal reaction due to the dilatations—comparable to an external carotid pressure—0.5 mg atropine is applied i. v. before each dilatation.

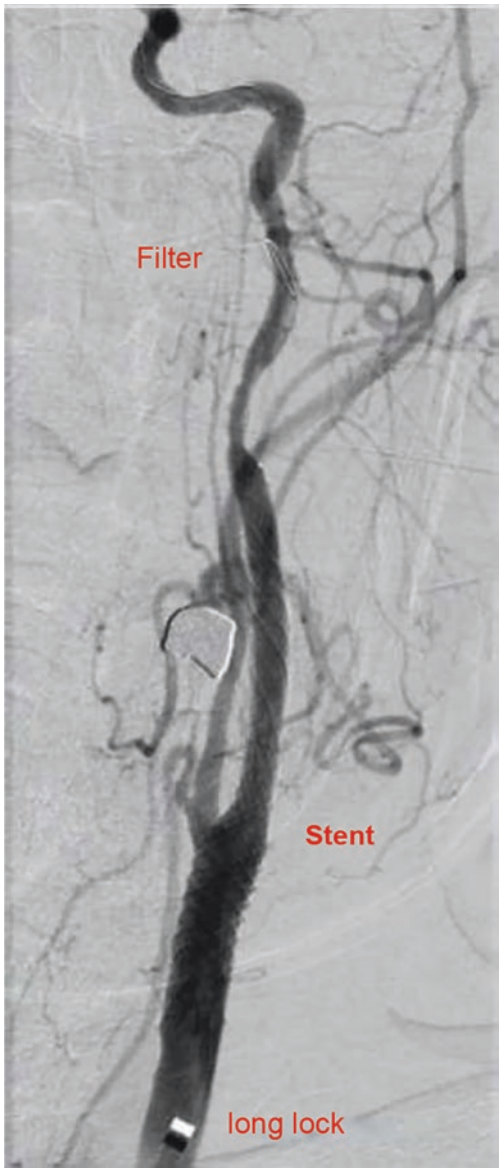

Fig. 14.13 Carotid stent

14.4.2 Radiotherapy

If a tumor can be treated surgically, it is usually removed by neurosurgery and, if there are residual findings, irradiated with or without chemotherapy.

The following are used

- **Stereotaxy** (e.g. for 1–3 brain metastases) or the
- **Proton therapy** (e.g. for chordomas, chondrosarcomas)

Side Effects

Appetite or sleep disturbances, optic nerve in a stereotaxy >15 Gy → visual impairment in about 1/3 d. F., acoustic neuroma (vestibular schwannoma) → hearing ability↓.

Complications

Radionecrosis (therefore compliance with the absolute doses for the single dose of a maximum of 10 Gy per 10 mL of brain tissue).

Gliomas

Glioblastoma

Therapy

- Definitive radiotherapy of the tumor in case of inoperability and always postoperative radiotherapy of the tumor bed. Dose in each case 60 Gy (5 × 2 Gy/week) or 40.05 Gy (5 × 2.67 Gy/week).
- Chemotherapy, e.g. with temozolomide, especially patients with an altered DNA repair enzyme MGMT benefit from this.

Astrocytoma

Definitive radiotherapy of the tumor in case of inoperability or postoperative RT of the tumor remnant from WHO grade II. Dose 54–60 Gy each (5 × 1.8–2.0 Gy/week).

Oligodendroglioma

Definitive radiotherapy of the tumor in case of inoperability or postoperative RT of the tumor remnant in WHO III, dose 54–60 Gy each (5 × 1.8–2.0 Gy/week).

Skull Base Meningiomas

- Definitive radiotherapy of the meningioma in case of inoperability or postoperative radiotherapy of the residual finding in case of incomplete tumor removal. The 5-year progression-free survival is 40–61% after subtotal surgery and 68–95% with subsequent radiotherapy.

Acoustic Neuromas (Vestibular Schwannomas)

- Radiotherapy of the tumor: The total dose in conventional technique is around 54 Gy (1.8–2 Gy/week). In the case of a small tumor, 12–15 Gy are given stereotactically.

Pituitary Adenoma

- Definitive radiotherapy of the adenoma in case of inoperability or postoperative radiotherapy of the residual findings in case of incomplete tumor removal
- Conventional radiotherapy: 45–50 Gy (5 × 1.8–2.0 Gy/week)
- Particle therapy for hormone-active pituitary adenoma
- Stereotaxy with 1 × or 3–5 × 5 Gy for prolactinoma and ACTH-producing adenomas

Chordomas, Chondrosarcomas of the Skull Base

- Conventional RT: 48–66.6 Gy
- Stereotaxy: 1 × 14-16 Gy
- Protons, heavy ions (helium, carbon): 60 CGE–83 CGE

Craniopharyngeomas

- Definitive radiotherapy of the tumor in case of inoperability or in case of residual tumor postoperative RT of the residual tumor, 54 Gy (5 × 1.8 Gy)
- Stereotaxy and particle therapy

Childhood

Cerebrospinal Fluid Space

Investigations into medulloblastoma have shown that at the time of diagnosis, 25–40% of tumor cells are floating in the CSF, and there is also a higher risk with germ cell tumors. In these cases, the entire CSF space is therefore irradiated.

Whole Brain

In leukemias, the entire brain is irradiated due to the diffuse cell distribution.

Brain Metastases

Therapy for Multiple Brain Metastases

- Multiple brain metastases: Whole brain irradiation

Therapy for 1–3 Brain Metastases

- Surgery, whole brain radiation (increasingly with hippocampal excision and stereotactic therapy of the individual metastases are used individually either solo or in combination).
- Stereotaxy for deeper metastases up to 3 cm in size, irradiated with high single dose, e.g. 1 × 15–24 Gy.

Glioblastoma

In the case of glioblastoma, definitive radiotherapy is given to the tumor if it is inoperable, and always postoperative radiotherapy to the tumor bed. The dose in each case is 60 Gy (5 × 2 Gy/week) or 40.05 Gy (5 × 2.67 Gy/week).

Brain Metastases

- Whole Brain Irradiation.

Case Study

Volker Asmacher, 59, is woken up in the morning by the alarm clock. However, he does not manage to switch off the alarm clock with his right hand. His wife notices that the right corner of his mouth is also hanging down. She immediately calls the emergency doctor. He suspects an acute stroke. Since Mr. Asmacher woke up with the symptoms, the time window, i.e. the exact onset of the symptoms, is unclear. There is a so-called wake-up stroke. Nevertheless, the emergency physician takes the patient to the nearest stroke unit as quickly as possible and also announces the patient there as an acute stroke. Here the patient is received by the neurologist on duty, Dr. Hammer. After a quick anamnesis, Mr. Asmacher is first taken to the CT. The native cranial CT suggests a hyperdense media sign on the left as an indication of a thrombotic occlusion of the cerebral artery, otherwise it is inconspicuous. As the patient is otherwise in a very good general condition, a CT perfusion and a CT angiography are performed in addition. Perfusion imaging shows an area at risk (called tissue at risk or penumbra) in the middle mediastinal flow area, but no demarcated area yet. CT angiography confirms occlusion of the left proximal middle cerebral artery in the M1 segment. Based on the good condition and CT perfusion result, Dr. Hammer discusses with the wife and patient the option of intravenous lysis with rtPA despite the unclear time window. Additionally, Dr. Hammer advises a thrombectomy to quickly reopen the vessel. Both measures are initiated after a few minutes of consideration. In the angiography to which Mr. Asmacher is taken, the occlusion of the left cerebral artery is still present. The thrombus is removed by means of a stent retriever. The symptoms improved rapidly after the operation. Subjectively, Mr. Asmacher is symptom-free again after a few days. The MRI three days later shows only a few small punctiform diffusion disturbances in the supply area of the left cerebral artery. Bleeding due to the lysis therapy did not occur.

Practice Questions

1. How to distinguish an epidural from a subdural hemorrhage?
2. Which localizations of intracerebral hemorrhage are called typical localizations and what is the general cause of these hemorrhages?
3. How can tumor-related edema be distinguished from ischemic edema on native CT?
4. What differential diagnoses should you be aware of for space-occupying lesions in the cerebellopontine angle?

Solutions ▶ Chap. 27