Postinfectious Encephalomyelitis (ADEM)

Definition

- **Epidemiology**
  
  *Frequency: 1–2:100,000*  
  *Peak age: 3–5 years*  
  The disorder can manifest itself at any age.

- **Etiology, pathophysiology, pathogenesis**

  Histopathology resembles multiple sclerosis, with acute demyelinating inflammation of the brain and spinal cord  
  In contrast to MS, the course is monophasic  
  Acute disseminated encephalomyelitis is often difficult to differentiate from the initial episode of MS  
  The cause is unknown but may represent a hypersensitivity reaction such as can occur 1–2 weeks after infection, inoculation, or chemotherapy.

Imaging Signs

- **Modality of choice**

  MRI.

- **CT findings**

  Multiple, subcortical, often round hypodensities that enhance with contrast.

- **MRI findings**

  Multiple, subcortical, often round focal lesions with high signal intensity on T2-weighted images  
  All lesions are in the same stage, meaning that they enhance uniformly  
  Often there is a ring-shaped pattern of enhancement in the acute stage of inflammation  
  Enhancement decreases as the inflammation subsides  
  Occasionally bull’s eye signs will be visible on T2-weighted images, lesions showing significant central hyperintensity (cystic necrosis secondary to demyelination) surrounded by moderate perifocal hyperintensity (edema).

Clinical Aspects

- **Typical presentation**

  Similar to multiple sclerosis, although the onset of symptoms is typically abrupt and monophasic  
  Often accompanied by fever, meningism, mental status changes, and convulsions, which are nearly invariably absent in MS.

- **Treatment options**

  Glucocorticoids  
  Plasmapheresis and cyclophosphamide may be indicated.

- **Course and prognosis**

  Mortality of the postinfectious form is about 10–40%  
  Neurologic deficits often persist.

- **What does the clinician want to know?**

  Differentiate from tumor or infarction.
Fig. 2.5a–d  Postinfectious encephalomyelitis or acute disseminated encephalomyelitis (ADEM). Axial T2-weighted MR images (a, c) and axial T1-weighted MR images after contrast administration (b, d). Multiple subcortical hyperintensities on T2-weighted MR images that are ring-enhancing on T1-weighted MR images. Identical enhancement in all lesions (b, d). The patient had undergone surgery to remove an oligodendroglioma and received chemotherapy several weeks previously. Right frontal postoperative tissue defect.
**Differential Diagnosis**

- **Cerebral abscess**
  - ADC usually reduced in cystic portion

- **Cerebral ischemia**
  - Pattern of distribution corresponds to area supplied by one or more vessels
  - In acute stage is ADC invariably reduced

- **Parasitic disorders (such as toxoplasmosis)**
  - Often immunocompromised persons
  - CSF findings

- **Multiple sclerosis**
  - Predilection for periventricular white matter

- **Metastases and higher grade multifocal glial tumors**
  - Solid portion: relative regional cerebral blood volume (rrCBV) on perfusion MR images at least twice as high as in normal white matter

**Tips and Pitfalls**

Misinterpreting the disorder as brain tumor or metastasis.

**Selected References**


Definition

▫ Epidemiology
  Prevalence: 1–8%.

▫ Etiology, pathophysiology, pathogenesis
  An aneurysm is dilatation of a blood vessel. Saccular aneurysms are the most common form.
  Risk factors: Family history of intracranial aneurysms • Autosomal dominant cystic kidney disease • Fibrous dysplasia • Coarctation of the aorta • Smoking.
  Pathogenesis: Degeneration and weakening of the internal elastic lamina and the collagen fibers of the arterial wall • Hemodynamic aspects.
  Localization: Bifurcations or origins of the following vessels in order of frequency: anterior cerebral, middle cerebral, internal carotid, basilar, and vertebral arteries.

Imaging Signs

▫ Modality of choice
  DSA.

▫ CT findings
  Round to oval extra-axial hyperdensity at one of the typical locations • An aneurysm visible on plain CT is usually larger than 5 mm • Arterial wall calcifications • Significant enhancement on CT after contrast administration and on CT angiography • A partially thrombosed vessel is recognizable as a filling defect or gap in the contrast medium • Sensitivity of CT angiography for aneurysms larger than 5 mm is about 94%; for aneurysms smaller than 5 mm it is only about 60%.

▫ MRI findings
  The MR signal is complex because it depends on the MRI sequence and the rate and direction of blood flow • There may be partial or complete thrombosis • Signal loss due to blood flow (flow void) is most apparent on proton density-weighted and T2-weighted images • The signal intensity of a suspected thrombus depends on its age • Sensitivity of MR angiography for aneurysms larger than 5 mm is about 86%; for aneurysms smaller than 5 mm it is only about 35%.

▫ DSA findings
  Aneurysm is visualized as a dilatation of the underlying vessel • The precise anatomy is demonstrated (proportional relation of aneurysmal neck to sac, relationship to vessels arising from the aneurysm) • DSA is also required to demonstrate smaller aneurysms (measuring less than 5 mm) in the presence of subarachnoid hemorrhage • DSA aids in planning treatment and in determining whether the aneurysm can be treated by occlusion of the underlying vessel (collateral circulation).
Fig. 3.3a–c  Saccular aneurysm of the trifurcation of the right middle cerebral artery. Axial T2-weighted MR image (a), coronal DSA after injection of the right internal carotid artery (b) and 3D rotational angiogram (c). Oval signal void originating at the main trunk of right middle cerebral artery (a, arrows). Vascular dilatation measuring approximately 15 mm (b, c).
Clinical Aspects

- **Typical presentation**
  Usually asymptomatic • Oculomotor and trochlear nerve palsy with impaired vision • Headache • Thromboembolic disease with ischemic stroke from partially or completely thrombosed aneurysms • The most severe complication is rupture with subarachnoid hemorrhage.

  Estimated cumulative rupture risk over 5 years:
  - Aneurysm less than 7 mm: 0% (internal carotid, anterior cerebral, middle cerebral arteries) or 2.5%, respectively (vertebral and basilar arteries).
  - Aneurysm measuring 7–12 mm: 2.6% (internal carotid, anterior cerebral, middle cerebral arteries) or 14.5%, respectively (vertebral and basilar arteries).
  - Aneurysm measuring 13–24 mm: 14.5% (internal carotid, anterior cerebral, middle cerebral arteries) or 18.4%, respectively (vertebral and basilar arteries).
  - Aneurysm larger than 24 mm: 40% (internal carotid, anterior cerebral, middle cerebral arteries) or 50%, respectively (vertebral and basilar arteries).

- **Treatment options**
  Coiling • Where coiling is not feasible, clipping.

- **Course and prognosis**
  Subarachnoid hemorrhage is fatal in 30% of cases, in 30% it leads to disability, and in 30% there are no neurologic deficits.
Ruptured aneurysm: Risk of severe disability or death after coiling is about 24% • Risk of severe disability or death after clipping is about 31%.

Unruptured aneurysm: Morbidity after coiling is about 5% • Morbidity after clipping is about 10%.

What does the clinician want to know?  
Size • Localization • Number and anatomy of aneurysms.

Differential Diagnosis

Vascular sling – Several different views with demonstration of the sling

Infundibular vascular origin – Symmetrical origin of arising vessel

Extra-axial brain tumor (can be confused with thrombosed aneurysm) – Thrombus usually does not enhance

Venous aneurysm in arteriovenous malformation (AVM) – AVM demonstrated on DSA

Tips and Pitfalls

Thrombosed aneurysms on TOF MRA (without contrast enhancement) appear hyperintense like flowing blood.

Selected References


Definition

- **Etiology, pathophysiology, pathogenesis**
  
  Primary neurulation disturbance (defective closure of the neural tube) in the fourth to fifth weeks of pregnancy.  
  
  **Chiari I malformation**: Slight incongruity between the posterior cranial fossa (slightly too small) and the cerebellum (normal size), resulting in low-lying cerebellar tonsils.  
  
  Associated malformations: hydrocephalus, syringomyelia, skeletal anomalies (basilar invagination, Klippel–Feil syndrome, atlantoaxial fusion).  
  
  **Arnold–Chiari malformation (Chiari II malformation)**: Complex anomaly with skull, dural, brain, spinal, and spinal cord manifestations.  
  
  Associated malformations: almost invariably lumbar myelomeningocele, syringomyelia (50–90%), diastematomyelia, anomalies of the corpus callosum, heterotopia.  
  
  **Chiari III malformation**: Arnold–Chiari malformation with deep occipital or high cervical encephalocele with cerebellar herniation.

Imaging Signs

- **Modality of choice**
  
  MRI.

**Chiari I Malformation**

- **CT findings**
  
  Abnormally high quantity of brain tissue in the foramen magnum.  
  There may be ventricular enlargement.  
  Narrowed peripheral CSF spaces above the surface of the cerebellum.

- **MRI findings**
  
  Triangular tonsils.  
  Narrowed peripheral CSF spaces above the surface of the cerebellum.  
  Low-lying cerebellar tonsils, more than 5 mm below the level of the foramen magnum (opisthion–basion line).  
  Syringomyelia in 20–40% of all patients.  
  Reduced CSF flow in the foramen magnum.

**Arnold–Chiari Malformation (Chiari II Malformation)**

- **CT findings**
  
  Calvarial defects.  
  Concave clivus.

- **MRI findings**
  
  Extreme elongation of the cerebellar tonsils, which can extend to the level of the C4 vertebra.  
  Elongated fourth ventricle.  
  Beak-shaped tectum.  
  Z-shaped kink at the junction of the medulla oblongata and cervical spinal cord.  
  Interdigitation of the gyri of the cerebral hemispheres.  
  Cerebellum is pressed against the brainstem and bulges superiorly past the tentorial notch.  
  Hydrocephalus.  
  Large interthalamic adhesion.  
  Fenestrated flax cerebri.  
  Low-lying transverse sinus and confluence of sinuses.  
  Narrowed posterior cranial fossa, concave clivus.
Clinical Aspects

- **Typical presentation/Course and prognosis**
  
  **Chiari I malformation:** Fifty percent of all cases are asymptomatic. Brainstem compression produces cranial nerve dysfunction. Somnolence. Neck pain.

  Symptomatic syringomyelia can imitate the clinical picture of multiple sclerosis.

  **Arnold–Chiari malformation (Chiari II malformation):** Myelomeningocele. Mac- 
  

- **Treatment options**
  
  Treatment of myelomeningocele or hydrocephalus where present. Decompression osteotomy of the foramen magnum.

- **What does the clinician want to know?**
  
  Follow-up with hydrocephalus. Demonstrate associated malformations such as myelomeningocele.

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**Fig. 11.1** Schematic diagram of the pathology in Arnold–Chiari malformation (1 = elongated fourth ventricle; 2 = pronounced elongation of the cerebellar tonsils displaced into the spinal canal; 3 = spinal cord “spurs”; 4 = spinal cord kink; 5 = cerebellum pressed against the brainstem; 6 = low-lying confluence of the sinuses; 7 = concave clivus; 8 = beak-shaped tectum; 9 = large interthalamic adhesion; 10 = partial agenesis of the corpus callosum).
**Fig. 11.2** Chiari I malformation. Sagittal T1-weighted MR image. Low-lying cerebellar tonsils, approximately 10 mm lower than the foramen magnum, are the only abnormal findings.

**Fig. 11.3 a, b** Arnold–Chiari (Chiari II) malformation. Sagittal T2-weighted MR image (a) and axial T1-weighted MR image (b). Beak-shaped tectum, hydrocephalus, and a narrowed posterior cranial fossa (a). Meshing of the gyri (b; arrows).
Differential Diagnosis

Acquired low-lying cerebellar tonsils in the presence of basilar impression or elevated intracranial pressure

- Basilar impression
- Elevated intracranial pressure

Hydrocephalus from other causes

- Demonstrate the cause, best done with MRI

Selected References

