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% \bullet Neurologic deficits often persist.

▶ What does the clinician want to know?

Differentiate from tumor or infarction.

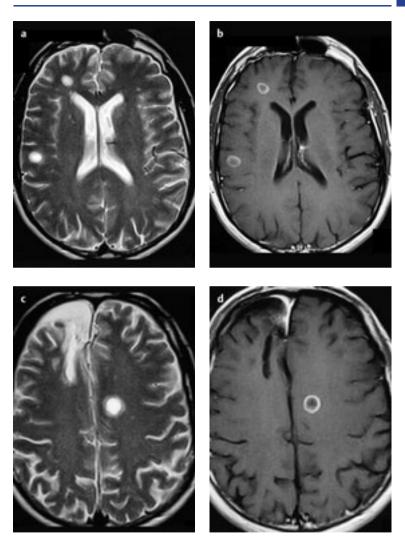


Fig. 2.5 a-d Postinfectious encephalomyelitis or acute disseminated encephalomyelitis (ADEM). Axial T2-weighted MR images (**a, c**) and axial T1-weighted MR images after contrast administration (**b, d**). Multiple subcortical hyperintensities on T2-weighted MR images that are ring-enhancing on T1-weighted 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

- ADC usually reduced in cystic portion Cerebral abscess 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

Hartmann M et al. Funktionelle MR-Verfahren in der Diagnostik intraaxialer Hirntumoren: Perfusions- und Diffusionsbildgebung. Rofo Fortschr Geb Rontgenstr Neuen Bildgeb Verfahr 2002; 174 (8): 955-964

Niedermayer I et al. Neuropathologische und neuroradiologische Aspekte akuter disseminierter Enzephalomyelitiden (ADEM), Radiologe 2000; 40 (11): 1030-1035

Schwarz S et al. Akute disseminierte Enzephalomyelitis (ADEM). Nervenarzt 2001; 72 (4): 241-254