

but there may be a postherpetic neuralgia syndrome (particularly after age 60 years) characterized by persistent pain, sometimes induced by stimuli that are otherwise not painful.

Overt CNS involvement by herpes zoster is much rarer, but can be severe. Herpes zoster can cause a granulomatous arteritis. In immunosuppressed individuals, herpes zoster may cause acute encephalitis characterized by numerous sharply circumscribed demyelinating lesions that subsequently undergo necrosis.

Cytomegalovirus

CMV infection of the nervous system occurs in fetuses and immunosuppressed individuals. The outcome of infection in utero is periventricular necrosis that produces severe brain destruction followed later by microcephaly and periventricular calcification. CMV is a common opportunistic viral pathogen in individuals with AIDS, with CNS involvement also occurring in this setting.

MORPHOLOGY

In the immunosuppressed individual, CMV most commonly causes subacute encephalitis, which may be associated with CMV inclusion-bearing cells (see Fig. 8-15). The infection tends to localize in the paraventricular subependymal regions of the brain, where it results in a severe hemorrhagic necrotizing ventriculoencephalitis and a choroid pleatitis. The virus can also attack the lower spinal cord and roots, producing a painful radiculoneuritis. Any cell in the CNS (neurons, glia, ependyma, or endothelium) may be infected. Prominent enlarged cells with intranuclear and intracytoplasmic inclusions can be readily identified by conventional light microscopy and CMV infection is confirmed by immunohistochemistry.

Poliomyelitis

While paralytic poliomyelitis has been eradicated by vaccination in many parts of the world, there are still a few countries where it remains a serious problem. In nonimmunized individuals poliovirus infection causes a subclinical or mild gastroenteritis, similar to that caused by other members of the picornavirus group of enteroviruses. In a small fraction of the vulnerable population, however, the virus secondarily invades the nervous system.

MORPHOLOGY

Acute cases show mononuclear cell perivascular cuffs and neuronophagia of the **anterior horn motor neurons of the spinal cord**. The inflammatory reaction is usually confined to the anterior horns but may extend into the posterior horns, and the damage is occasionally severe enough to produce cavitation. Poliovirus RNA has been detected in anterior horn cell motor neurons. The cranial motor nuclei are sometimes involved as well. Postmortem examination in long-term survivors of symptomatic poliomyelitis shows loss of neurons and gliosis in the affected anterior horns of the spinal cord, some residual inflammation, atrophy of the anterior (motor) spinal roots, and neurogenic atrophy of denervated muscle.

Clinical Features. CNS infection manifests initially with meningeal irritation and a CSF picture consistent with aseptic meningitis. The disease may progress no further or advance to involve the spinal cord. When the disease affects the motor neurons of the spinal cord, it produces a flaccid paralysis associated with muscle wasting and hyporeflexia in the corresponding region of the body—the permanent neurologic residue of poliomyelitis. Because of the destruction of motor neurons, paresis or paralysis follows; when the diaphragm and intercostal muscles are affected, severe respiratory compromise and even death may occur. A myocarditis sometimes complicates the acute infection. *Postpolio syndrome* can develop in patients 25 to 35 years after the resolution of the initial illness. It is characterized by progressive weakness associated with decreased muscle mass and pain, and has been attributed to superimposed neuronal loss of aging with inflammatory mechanisms, but without any convincing evidence of viral re-activation.

Rabies

Rabies is severe encephalitis transmitted to humans by the bite of a rabid animal, usually a dog or various wild mammals that are natural reservoirs. Exposure to certain species of bats, even without a known bite, can also lead to rabies.

MORPHOLOGY

External examination of the brain shows intense edema and vascular congestion. Microscopically, there is widespread neuronal degeneration and an inflammatory reaction that is most severe in the brainstem. The basal ganglia, spinal cord, and dorsal root ganglia may also be involved. **Negri bodies**, the pathognomonic microscopic finding, are cytoplasmic, round to oval, eosinophilic inclusions that can be found in pyramidal neurons of the hippocampus and Purkinje cells of the cerebellum, sites usually devoid of inflammation (Fig. 28-25). Rabies virus can be detected within Negri bodies by ultrastructural and immunohistochemical methods.

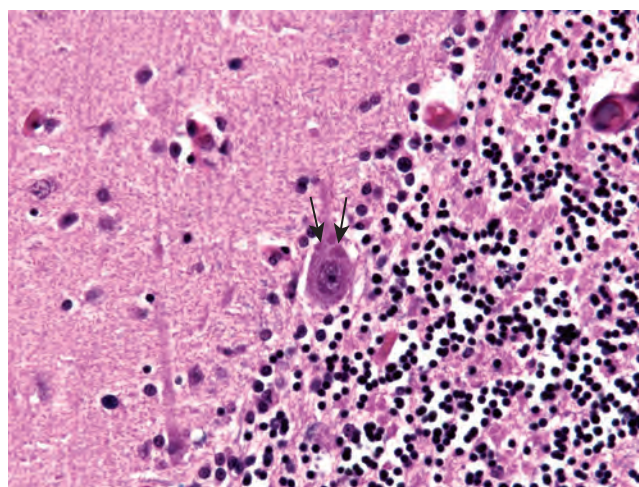


Figure 28-25 The diagnostic histologic finding in rabies is the eosinophilic Negri body, as seen here in a Purkinje cell (arrows).