8. Postnatal Lesions of Gray Matter

As stated at the begin of Chap. 7, there is only a relative difference between perinatal and postnatal onset of the lesions described here and in Chap. 7. Several of the "perinatal" lesions described in the previous chapter can also develop in early infancy, e.g. status marmoratus or ulegyria. Conversely, cardiac arrest encephalopathy, described in this chapter, was often reported as a typical form of perinatal damage. None the less, on review one will find that the lesions described in this chapter are more likely to be of postnatal origin and that they rarely or never arise before birth.

Cardiac Arrest Encephalopathy. The cerebral lesions of cardiac arrest encephalopathy were described independently in perinatal, adult and experimental neuropathology. The significance of this pattern emerged from clinico-pathological correlation for the acute stages of the disease.

Neuronal necrosis confined to certain nuclear groups of the brain stem ("hypotensive brain stem necrosis") was reported in four patients by Gilles (1963); two were infants 17 and 18 months old, the others 9 and 22 years. All had a history of cardiac arrest, surviving in an unresponsive, decerebrate state for various lengths of time. The brain stem showed symmetric loss of neurons and glial scarring in the motor cranial nerve nuclei, including oculomotor, trochlear, trigeminal, abducens, facial and hypoglossal nerve nuclei, as well as bilateral symmetric necrosis in the tegmentum, the superior and inferior colliculi, cuneate and gracilis nuclei and other nuclei. Damage was maximal in the central part of the nucleus, sparing the cells at its periphery. Three of these patients also had anoxic encephalopathy with diffuse loss of neurons in cerebral cortex and basal ganglia, but the fourth had only brain stem lesions. Similar necrotizing lesions of the brain stem were also described in neonates (Schneider et al. 1975; Leech and Alvord 1977). Their relationship to cardiac arrest was stressed by Dambksa et al. (1976). Janzer and Friede (1980) reviewed the clinical records of eight infants and seven adults with this pattern of brain stem damage and found an episode of cardiac arrest in each. They concluded that the pattern of "hypotensive brain stem necrosis" is pathognomonic for cardiac arrest and should be called "cardiac arrest encephalopathy". All patients remained in coma up to their death. Clinical follow-ups suggest a poor outcome for children who remain comatose after resuscitation from cardiac arrest (Seshia et al. 1979). The episode of cardiac arrest is usually postnatal or later in life, but brain stem necrosis can also be seen in stillborns (Taylor and Roessmann 1984). The pattern of brain stem necrosis had been described earlier as a sequel of experimental perinatal asphyxia in rhesus monkeys (Ranck and Windle 1959; Windle 1963). The relationship of these experimental lesions to cardiac arrest was recognized later (Myers 1973; Myers and Yamaguchi 1977).
The acute lesions of cardiac arrest encephalopathy are usually not discernible to the naked eye; an indistinct grayish discoloration of the brain stem may be seen. Lesions are almost never hemorrhagic. With the microscope, neurons with eosinophilic degeneration have a characteristic distribution in the center of the various nuclei. Neurons situated at the periphery of the nuclei tend to remain spared. Large central necrotic areas in the posterior colliculi (Fig. 8.1) serve as a hallmark. Similar, less conspicuous lesions affect the reticular formation, many of the cranial nerve nuclei, gracilis and cuneate nuclei or the nuclei of the brain stem and the ventral gray matter of the spinal cord.

This pattern of brain stem damage is sometimes found alone, but it is seen more often in the context of widespread and severe anoxic damage in the hemispheres. Only the brain stem involvement is diagnostic of cardiac arrest. The syndrome may also account for some cases of the Möbius syndrome (see Chap. 30). Exceptionally severe, destructive lesions of the brain stem were associated with cerebellar hypoplasia in the two neonates reported by Gessaga et al. (1986).

**Fig. 8.1.** Hypotensive brain stem necrosis: bilateral necrosis of superior colliculi, Cresyl violet, x 6.5

**Spinal Cord Necrosis.** Necrosis of the ventral horns of the spinal cord is a typical component of cardiac arrest encephalopathy. There are a few reports on selective cord necrosis in neonates (Adams and Cameron 1965) or of congenital cervical spinal atrophy of undetermined origin (Darwish et al. 1981). Hyperextension during delivery may cause spinal cord infarction, but this is difficult to verify if the lesion developed in utero, as in the infant reported by Young et al. (1983). Tetanus neonatorum is another exceptional cause of cord necrosis (Gadoth et al. 1981).

**Retrolental Fibroplasia.** Retrolental fibroplasia due to the toxic effect of high oxygen concentration on the immature retina often combines with mental retarda-