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MORPHOLOGY AND HISTOCHEMISTRY OF WILSONIAN AND HEPATOGENIC GLIOPATHY IN TISSUE CULTURE

The essential feature of Wilson's disease and hepatogenic encephalopathy consists in coexsistence of generalized regressive and progressive changes of glia mostly astrocytes and in occurrence of special glial forms such as Alzheimer cells type I and II Opalski cells. The similarity, both of these gliopathies might go so far that morphological differentiation of some cases of hepatogenic encephalopathy from Wilson disease is impossible without additional biochemical studies.

The aim of present work was to establish the direct effect of serum from patients with hepatolenticular degeneration and those with hepatic coma of the cultures of glia cells from rat cerebellum. These experiments were suplemented by the studies on the direct effect of copper and ammonia ions themselves, given to the normal culture medium in the ammount corresponding respectively to this of copper in the brain tissue in cases of Wilson disease and to that of ammonium in the blood in the conditions of hepatic coma.

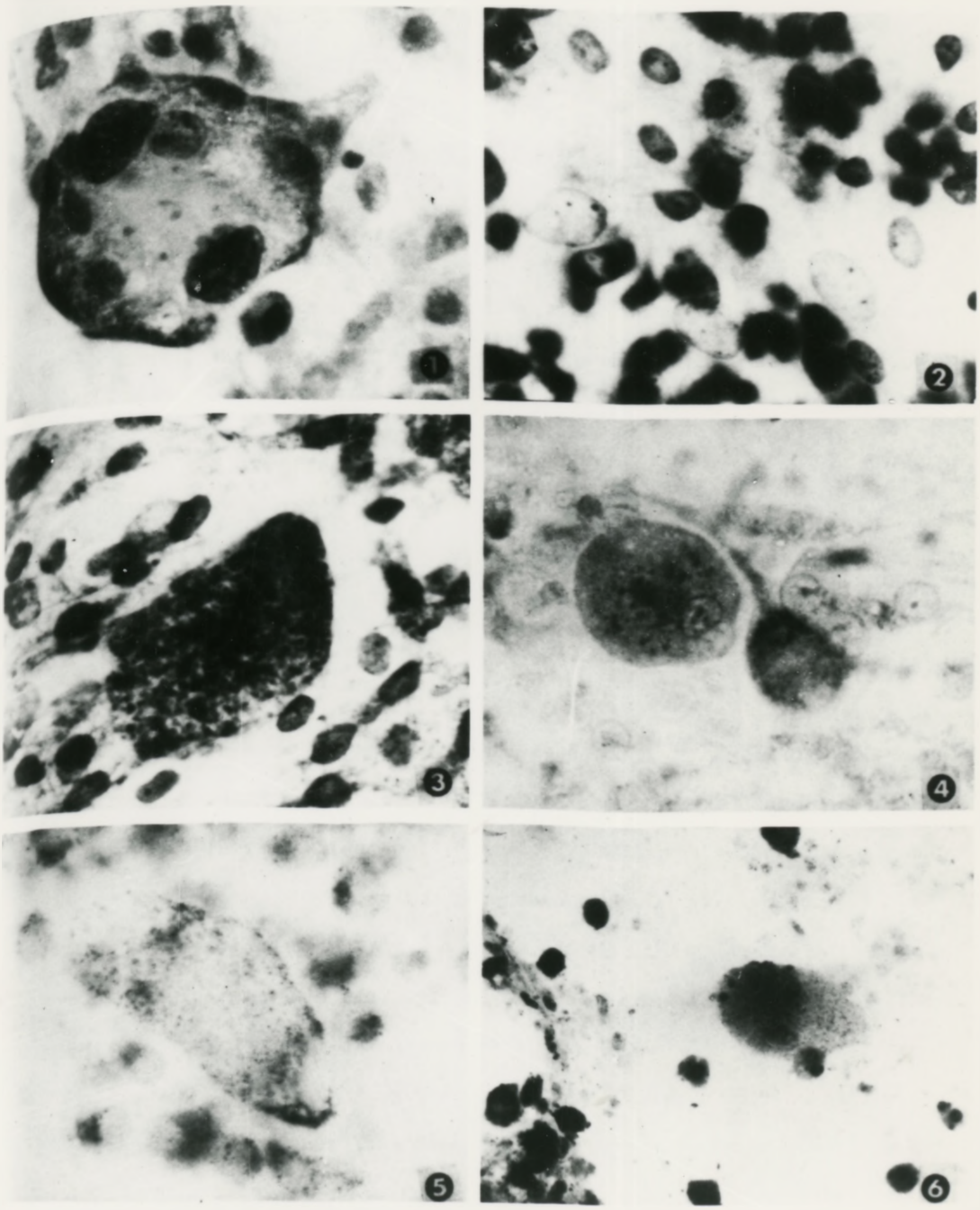
In all experimental groups characteristic morphological changes were observed; these consisted in the occurrence of Alzheimer cells type I and II and Opalski cells /Fig. 1, 2/. The above cellular types occured against the background of a normal and/or degenerating glial population. In addition some cellular elements, which could be considered as intermediate forms between normal astrocytes and Opalski cells, were observed.

Morphological changes were accompanied by histochemical abnormalities concerning mostly Opalski and intermediate cells. Accumulation of both neutral and acid mucopolysaccharides in the cytoplasm of Opalski cells, and to a lesser degree in intermediate cells was the most typical feature /Fig. 3/.

Paralelly to this reduction of the enzymatic activity of succinic and glutamic dehydrogenases was found /Fig. 4, 5/; Glucose-6-phosphate dehydrogenase and acid phosphate activities /Fig. 6/ were markedly increased.

In the authors' opinion both copper and ammonia damage some enzymes, involved in certain stages of cellular carbohydrate metabolism. Abnormal glucose metabolism of astrocytes leads to intracellular production and accumulation of mucopolysaccharides bound with the cellular proteins. The enzymatic disturbances and the storage of the above mentioned substances in the astrocyte cytoplasm is in turn, responsible for the observed morphological abnormalities.

The changes noted in tissue culture correspond both in their morphology and histochemistry to those described in the central nervous system in Wilson's disease and in hepatogenic encephalopathy.



Legends: Fig. 1. Opalski cell in culture /hepatic coma serum/.Cresyl violet. x 600. Fig. 2. Numerous Alzheimer cells type II /hepatic coma serum/.Cresyl violet. x 600. Fig. 3. Opalski cell with cytoplasmic accumulation. PAS-positive granules /normal serum with copper acetate/. x 600. Fig. 4. Opalski cell and intermediate one showing low SDH activity /serum from Wilsonian patient/. x 600. Fig. 5. Opalski cell with low GDH activity /normal serum with ammonium chloride/. x 600. Fig. 6. Acid phosphatase activity in Opalski cell / normal serum with copper acetate/. x 300.