Ultrastructure of Remyelination of Peripheral Nerves in Landry-Guillain-Barré Syndrome

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Summary. Ultrastructural study of peripheral nerves in a case of Landry-Guillain-Barré syndrome dying during early recovery phase reveals characteristic findings which are interpreted as being regenerative changes. Layers of compact vesicles and tubules are seen around each axon and coalescence of these vesicles appears to be followed by formation of myelin lamella. There is suggestive evidence that the vesicles may be produced by the endoplasmic reticulum of the axon.

Zusammenfassung. Die ultrastrukturelle Untersuchung der peripheren Nerven eines Falles von Landry-Guillain-Barré Syndrom, der während einer kurzen Erholungsphase starb, ergibt charakteristische Befunde, die als regenerative Veränderungen gedeutet werden. Um jedes Axon sieht man geschlossene Lagen von Vesikeln und Tubuli; das Verschmelzen dieser Vesikeln scheint von der Bildung der Myelinlamellen gefolgt zu werden. Es drängt sich die Annahme auf, daß die Vesikeln vom endoplasmischen Reticulum der Axone gebildet werden.

Key-Words: Remyelination — Landry-Guillain-Barré-Syndrome

Clinical History

The patient, a 21 year old college student, had been in good health until one morning when she awoke with tingling sensation beginning in the soles of her feet spreading to the legs and later on to the hands. This was followed two days later by progressive weakness and loss of position sense of legs, later of hands and loss of coordination. There were prodromal symptoms of a mild cold with sore throat three days prior to onset of neurological symptoms.

On admission she had mild weakness of abduction in the eyes bilaterally, marked weakness and sensory loss of trunk and all extremities and generalized decrease of deep tendon reflexes. C.S.F. showed a pressure of 150 with clear fluid; total protein 46, sugar 70 and chlorides 127.

Peripheral blood picture and blood chemistry were normal. Her hospital course was marked by rapid progression of motor and sensory disturbances leading to complete paralysis, cranial nerve palsy and respiratory paralysis. She underwent tracheotomy on the 4th hospital day and was placed in a positive pressure Bird respirator. On the 12th hospital day (17 days after onset of neurological symptoms) she suddenly bled profusely from around the tracheostomy opening and was dead in a few minutes.

Autopsy performed 15 hours after death revealed massive fresh hemorrhage into tracheobronchial tree due to erosion of right innominate artery by peritracheal abscess related to the tracheostomy implement. The brain and spinal cord were grossly normal. Small segments of spinal nerve roots and sciatic nerves were placed in 1% osmic acid buffered with collidine and embedded in Epon. Ultrathin sections were stained with uranyl lead and lead citrate. Samples of brains and peripheral nerves from nine other patients with postmortem intervals up to 17 hours were processed for electron microscopy in order to rule out postmortem artefacts.
Ultrastructural findings were unique and were not encountered in any of the controls. In the spinal nerve roots, the myelin sheaths had a characteristic stratified appearance and were composed of layers of compact vesicles of 800–1000 Å in width alternating with layers of parallel lamella (Fig. 1). The number of layers varied from 2–10, and the innermost layer adjacent to the axon consisted invariably of vesicles. The vesicles were bound by single membrane of approximately 140 Å thick. Occasionally, parallel elongated tubules and large cystic areas containing a flocculent material were also seen amongst the vesicles. The myelin lamella appeared to be formed by limiting membranes of disintegrating vesicles. The myelin segments were initially thick, wavy, discontinuous; cross striations of approximately 100 Å apart were often seen in these segments and in the limiting membranes of the vesicles. The deposition of myelin lamella appeared to progress from the axon outwards, and multiple mesaxons were often seen at the periphery. The axons contained abundant mitochondria and unusually prominent smooth endoplasmic reticulum (SER) in the form of branching tubules. Within these tubules there were scattered small vesicles identical in size and shape to those seen in the myelin sheath. In some areas, the axonal SER appeared to be continuous with the tubules and vesicles located in the myelin sheath. In still other areas, evidence suggesting branching of the axon was seen (Fig. 2).