Electron-microscopical Investigation of the Skin in Angiokeratoma corporis diffusum

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It has been known for several years that angiokeratoma corporis diffusum should be regarded as a symptom of a disorder of lipid metabolism. The latter affects vascular smooth muscle, endothelial cells, the muscle fibres of the heart, ganglion cells etc., resulting among other things in severe damage of blood vessels, kidneys and heart. The prognosis is poor in the long run. The nature of the stored pathological lipid is not quite clear up to now. Kühnau refers to a phosphatid related to sphingomyelin. Sweeley and Klionsky believe to have demonstrated a neutral glycolipid (ceramide-thrihexoside). Electron microscopy might constitute a new approach to the problem of this "thesaurismosis".

Electron microscopic findings in the skin in two cases of angiokeratoma corporis diffusum (males 41 and 38 years of age resp.) are presented. In one of these cases only formol fixed autopsy material was available; in the other case in respect of electron microscopy adequately fixed biopsy material was used.

In the formol fixed autopsy material, more especially in endothelial cells and fibrocytes, chiefly osmiophilic granules were observed, which were indicated as type 1 and 2.

Type 1 represented round or oval inclusions consisting of more or less concentrically arranged wide osmiophilic bands (500 Å thick) alternating with light zones (83 Å thick). Type 2 represented more elongated osmiophilic bodies, in which a transverse striation (zebra pattern) could be discerned. The black bands varied considerably in width (being of the order of 700 Å), and the same was true of the white bands between them.

In the adequately fixed biopsy material of the second patient osmiophilic granules of a lamellar type (type 3) were mainly seen. The latter consisted of lamellar systems with tightly concentrically arranged alternating dense and light lines. The thickness of the osmiophilic lines was 26 Å, of the osmiophobic ones 36 Å, which means a periodicity of 62 Å. These lamellar granules often enclosed a granular or homogenous electron-opaque centre. The size of the corpuscles varied from 0.2 to 1.4 µ with an average of approx. 0.5 µ. They showed a close resemblance to lamellar osmiophilic granules demonstrated in Tay-Sachs disease, but differed from those by a number of

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Fig. 1. Lamellar osmiophilic bodies (type 3) within a fibrocyte. $\times 17,200$.

ultra-structural details. Attention is drawn to the marked predominance of osmiophilic granules of the types 1 and 2 in the formol fixed autopsy material. Unfavourable conditions (unsuitable formol fixation, autolysis) may have been partly responsible for this phenomenon.

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