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Ultrastructure of Angiokeratoma Vulvae

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Karlsmark T, Weismann K, Kobayasi T. Ultrastructure of angiokeratoma vulvae. *Acta Derm Venereol (Stockh)* 1988; 68: 80-82.

A case of vulvar angiokeratoma studied by electron microscopy is described. The patient, a 51-year-old woman, had noticed eruptions for the last 15 years, though without symptoms. Osmiophilic bodies with myelin-like figures were found in the vascular endothelial cells. The findings support previous opinion that vulvar angiokeratoma is a variant of scrotal angiokeratoma. (Received July 14, 1987.)

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Angiokeratoma vulvae is probably a variant of scrotal angiokeratoma Fordyce (1). Few papers have described the condition, though it seems to be more common than generally

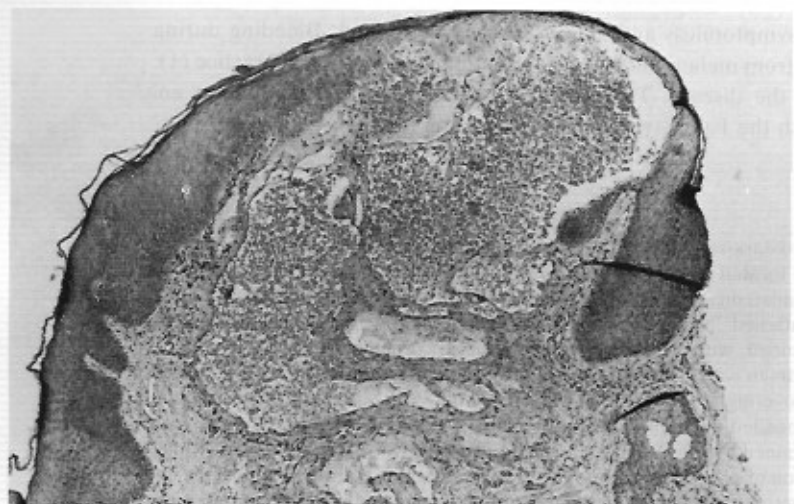


Fig. 1. Small blood vessels in the papillary dermis are dilated cavernously. The epidermis shows irregular thickening, with thin horny layers, $\times 290$.

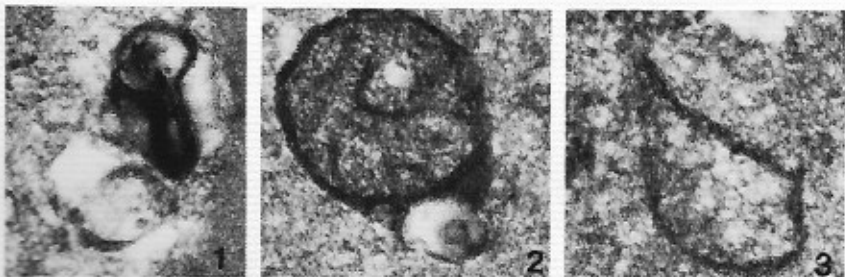
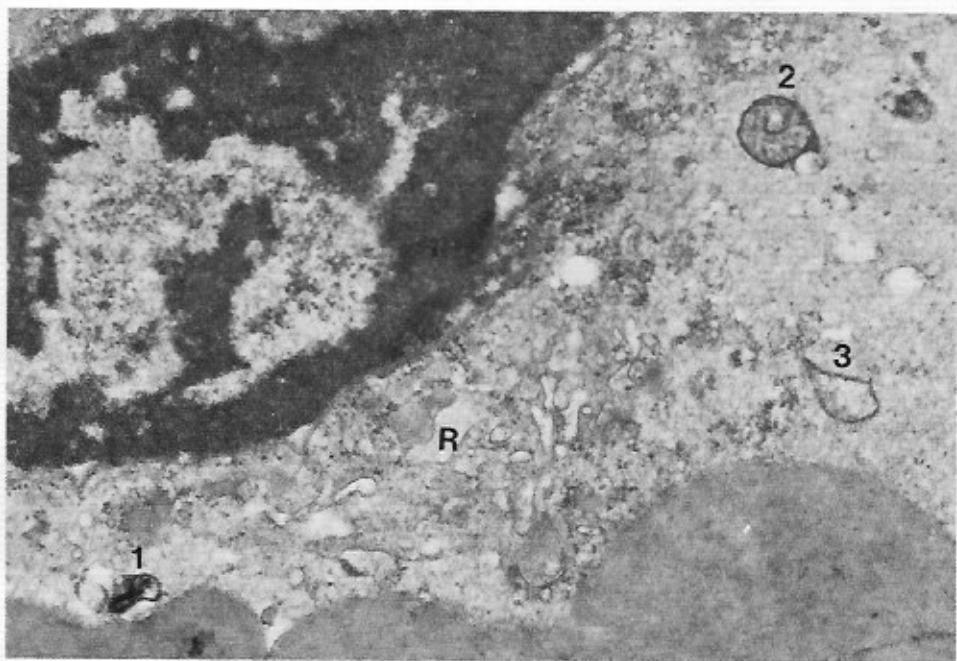


Fig. 2. An endothelial cell shows osmiophilic bodies (1, 2, 3) and dilated cisterna of reticula (R). $\times 20000$. Insets: Details of osmiophilic bodies (1, 2, 3) with myelin-like figures. $\times 60000$.

realized (2). The lesions are symptomless and may easily be overlooked. Bleeding during pregnancy and differentiation from melanoma may call attention in daily clinical practice (1). This paper reports a case of the disease. The ultrastructural changes in the vascular endothelium were compared with the Fabry-type of angiokeratoma (3).

CASE REPORT

A 51-year-old woman had observed dark-red papules on her vulva for the last 15 years. The papules were symptomless, 1–2 mm in size and located on both major and minor labia. Histological examination by light microscopy (Fig. 1) showed dilated capillaries in the papillar dermis filled with erythrocytes in the lumen. Endothelial cells were flattened. Neither inflammatory cells nor brown pigment-holding cells were seen. The epidermis was thinned, with a multiple-layered stratum corneum.

For electron microscopy a specimen was fixed in a 6% glutaraldehyde solution in cacodylate buffer, pH 7.4, with 7.5% sucrose at 4°C overnight. Post-fixation was carried out in 1% osmium tetroxide in the same buffer. Specimens were embedded in Epon. Ultrathin sections were cut from selected areas with a Reichert ultramicrotome and stained by a combined technique using uranyl acetate and lead citrate. A Jeol 100CX transmission electron microscope was used for the examination. Ultrastructurally, the endothelial cells contained small electron-dense bodies with myelin-like figures and dilated cisterna of endoplasmic reticula. Ribosomes were spread in the cytoplasm, few were attached to the endoplasmatic reticula (Fig. 2).

COMMENTS

Angiokeratoma is known to occur in five different clinical types (4). Especially diffuse angiokeratoma corporis of Fabry has been studied by electron microscopy. Vascular endothelial cells have been shown to contain osmiophilic bodies with myelin-like figures. The bodies are large and numerous, which is a valuable sign for the diagnosis (3). In the present paper we report a finding of similar bodies in the endothelial cells, but they appear smaller and are less frequent.

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Fig. 1. Dilated capillaries in the papillar dermis filled with erythrocytes. The endothelial cells are flattened. The epidermis is thinned with a multiple-layered stratum corneum.