

AN ULTRASTRUCTURAL STUDY OF NECK SKIN FROM
A PATIENT WITH PSEUDOXANTHOMA ELASTICUM.

By

B.W. Oakes and M.L. Wahlqvist

Monash University.

Pseudoxanthoma elasticum is a familial connective tissue disease characterized by abnormal mineralization of the dermis and vascular system. This report describes the light and electron microscopic observations of the dermis obtained from a neck skin biopsy of a 28 year old man with the disease. He has bilateral femoral artery occlusions and marked femoral calcification together with classical fundal angioid streaks and lax skin of the neck and axilla.

Light microscopic examination of the skin revealed abnormal calcified elastic fibres in the mid-dermis. Ultrastructurally, electron-dense deposits were seen within the central region of these elastic fibres. Needle-like calcium apatite crystals were frequently observed within the electron-dense deposits both in the dermal elastin and also within and associated with numerous matrix vesicles in the internal elastic lamina of a dermal arteriole. In tissue stained with Ruthenium red, calcium apatite crystals were also seen, frequently associated with the surface of elastic fibres and throughout the bundles of collagen fibres. Cells in these areas of dense concentrations of calcium apatite crystals also contained similar needle-like crystals within peripheral vesicles. A dermal lymphatic vessel contained calcium apatite crystals within its lumen and similar crystals were seen within vesicles in the lymphatic endothelium. EDTA treatment of small blocks of dermis substantially removed the electron-dense material from the elastic fibres.

These observations indicate that although calcification occurs primarily within the mid-dermal elastic fibres, collagen or a collagen associated component also has the capacity to induce calcium apatite formation in the skin in this rare clinical condition.