



The contribution of arterial calcification to peripheral arterial disease in pseudoxanthoma elasticum

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BACKGROUND AND AIMS: The contribution of arterial calcification (AC) in peripheral arterial disease (PAD) and arterial wall compressibility is a matter of debate. Pseudoxanthoma elasticum (PXE), an inherited metabolic disease due to ABCC6 gene mutations, combines elastic fiber fragmentation and calcification in various soft tissues including the arterial wall. Since AC is associated with PAD, a frequent complication of PXE, we sought to determine the role of AC in PAD and arterial wall compressibility in this group of patients. **METHODS AND RESULTS:** Arterial compressibility and patency were determined by ankle-brachial pressure index (ABI) in a cohort of 71 PXE patients (mean age 48 +/- SD 14 yrs, 45 women) and compared to 30 controls without PAD. Lower limb arterial calcification (LLAC) was determined by non-contrast enhanced helicoidal CT-scan. A calcification score (Ca-score) was computed for the femoral, popliteal and sub-popliteal artery segments of both legs. Forty patients with PXE had an ABI<0.90 and none had an ABI>1.40. LLAC increased with age, significantly more in PXE subjects than controls. A negative association was found between LLAC and ABI ($r = -0.363$, $p = 0.002$). The LLAC was independently associated with PXE and age, and ABI was not linked to cardiovascular risk factors. **CONCLUSIONS:** The presence of AC was associated with PAD and PXE without affecting arterial compressibility. PAD in PXE patients is probably due to proximal obstructive lesions developing independently from cardiovascular risk factors.

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