

Relationship between ankle brachial index and arterial remodeling in pseudoxanthoma elasticum

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Résumé en anglais	ObjectivesPseudoxanthoma elasticum (PXE) is an inherited metabolic disease characterized by elastic fiber fragmentation and calcification in the cutaneous, ophthalmologic, and vascular tissues. Cardiovascular manifestations such as peripheral arterial disease (PAD) are frequent in PXE. Because of the changes in the elastic properties and medial calcification of the arterial wall in PXE, the impact of the arterial remodeling on the ankle brachial index (ABI), a well-established diagnostic method for the detection and follow-up of PAD, remains to be determined in this disease. Methods This was a cross-sectional, comparative, open study, which took place at the PXE Consultation Center, University Hospital of Angers. The subjects were 53 patients (mean age, 49 ± 14 years; 35 females) with PXE clinically proven on the basis of established criteria (skin changes, angioid streaks, and skin biopsy). The ABI at rest, symptoms of intermittent claudication (IC), carotid intimamedia thickness (IMT), carotid-femoral pulse wave velocity (c-f PWV), compliance (CC), and β stiffness index were measured in a single-center cohort. Results Forty-five percent of the PXE patients had an ABI ≤ 0.90 , but only one patient had an ABI > 1.40 . IC was found in 23% of the patients with a low and normal ABI in terms of IMT (P = .566) or β stiffness index (P = .194), but differences were significant for c-f PWV (P = .010) and CC (P = .011). Adjusted multivariate linear regression for the Framingham-Laurier score showed that patients with a low ABI had less compliant carotid arteries (B = 0.318, P = .039). Conclusions PAD detected by a low ABI is very frequent in PXE, although with limited prevalence of symptomatic claudication. Unexpectedly, ABI was low in such calcifying PAD and associated with lower CC, independently of atherosclerosis risk factors. These findings demonstrate that PXE represents a unique monogenic model of PAD in which the specific arterial wall remodeling could change the diagnostic value of the ABI to detect PA	

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