

Teaching NeuroImage: Microangiopathic complications in pseudoxanthoma elasticum

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A 43-year-old woman without vascular risk factors presented with acute right sensorimotor deficit. Her medical history revealed pseudoxanthoma elasticum (PXE), diagnosed at age 29 by characteristic findings on skin biopsy (performed because of progressive skin lesions) and funduscopy (figure 1), complicated by peripheral artery disease and vision impairment.

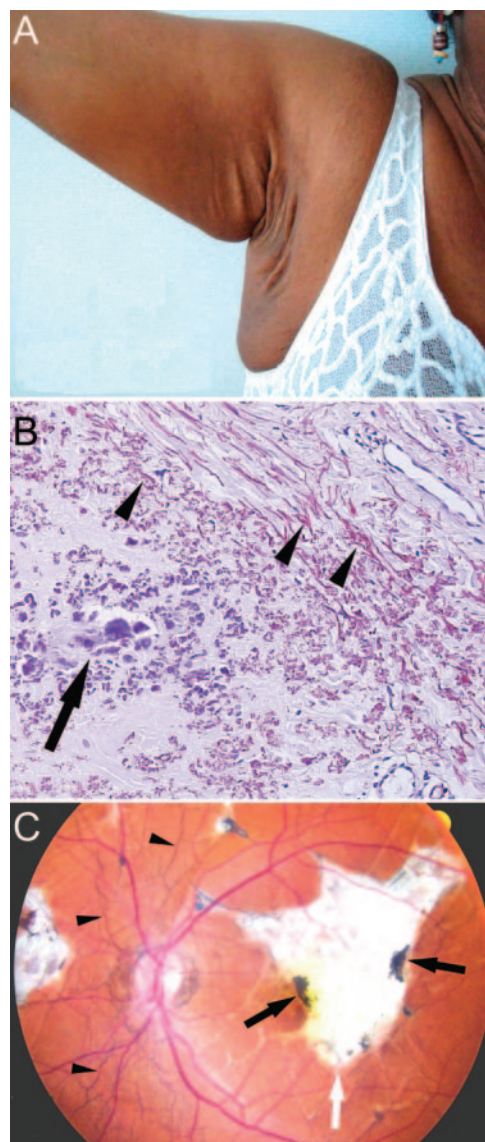
Brain MRI showed pontine lacunar infarcts and microbleeds (figure 2), concordant with earlier reported microangiopathic complications in PXE.^{1,2}

PXE is an inherited disorder characterized by progressive calcification and degeneration of elastic fibers, primarily affecting skin, eyes, and gastrointestinal and cardiovascular system. Management includes treatment of complications and strict control of cardiovascular risk factors.

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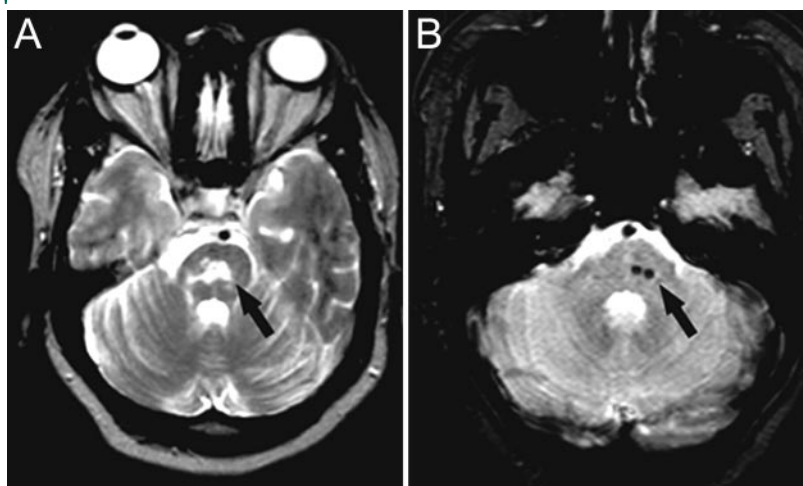
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Figure 1 Patient, skin histopathology, and eye fundus



(A) Redundant axillary skin folds demonstrate loss of elasticity. (B) Histopathology of the skin on orcein stain showing calcium deposits (arrow) and swollen, fragmented elastic fibers (arrowheads). (C) Fundus photography of the eye shows characteristic angioid streaks (arrowheads), and a large retinal scar (white arrow) due to a macular hemorrhage (black arrows).

Figure 2 Brain MRI



(A) MRI showing pontine ischemic T2 hyperintensities and (B) multiple foci of low intensity in the pons on echogradiant T2-weighted images consistent with microbleeds.

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