

GRÖNBLAD-STRANDBERG SYNDROME

Authors: Fernanda Fernandes Gomes, Amanda Lamóglia Bittencourt, Luiza Gabriela Zain, Eric Vieira, Taís Câmara V. dos Santos, Jean Vitor Martins Alves, Francisco Grupenmacher, Alex Treiger Grupenmacher

PURPOSE

To report a case of pseudoxanthoma elasticum presenting with angioid streaks in the fundus examination.

METHODS

Examination of medical records, fundus photographs, and OCT images.

RESULTS

A 53-year-old male with no significant ophthalmological history, previously diagnosed with pseudoxanthoma elasticum, presented at Hospital Santa Casa de Misericórdia de Curitiba with complaints of diminished visual acuity. His medical records revealed only controlled dyslipidemia. Ophthalmological evaluation showed best-corrected visual acuity of 20/200 in both eyes. Biomicroscopy indicated discreet corneal conjunctivalization at 7 o'clock on the left eye with no additional findings. Fundoscopy revealed, in both eyes, a well-defined optic nerve with peripapillary ring-like atrophy with irregular radiations and macular atrophy. The left eye exhibited subretinal scarring on the superior temporal arcade. Optical coherence tomography (OCT) imaging revealed scarring with localized fibrosis in both eyes, an inactive neovascular membrane in the right eye, and an active neovascular membrane in the left eye. Upon general physical examination, the patient presented typical yellowish papules and plaques on his lateral cervical area.

DISCUSSION

Pseudoxanthoma elasticum is a rare hereditary disorder characterized by pathological calcification of elastic connective tissue. Its simultaneous presentation with angioid streaks defines the Gronblad-Strandberg Syndrome. This syndrome manifests as angioid streaks (rupture of Bruch's membrane) in fundus examination, characteristic skin lesions, and arteriosclerosis. Therefore, comprehensive cardiological and ophthalmological evaluations are crucial in patients diagnosed with pseudoxanthoma elasticum to detect and treat early signs of the disease, thereby preventing further morbidity and visual loss.

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IMAGE 1: FUNDOSCOPY OD



IMAGE 2/3: RETINOGRAFY OD



IMAGE 1: YELLOWISH PAPULES ON CERVICAL AREA

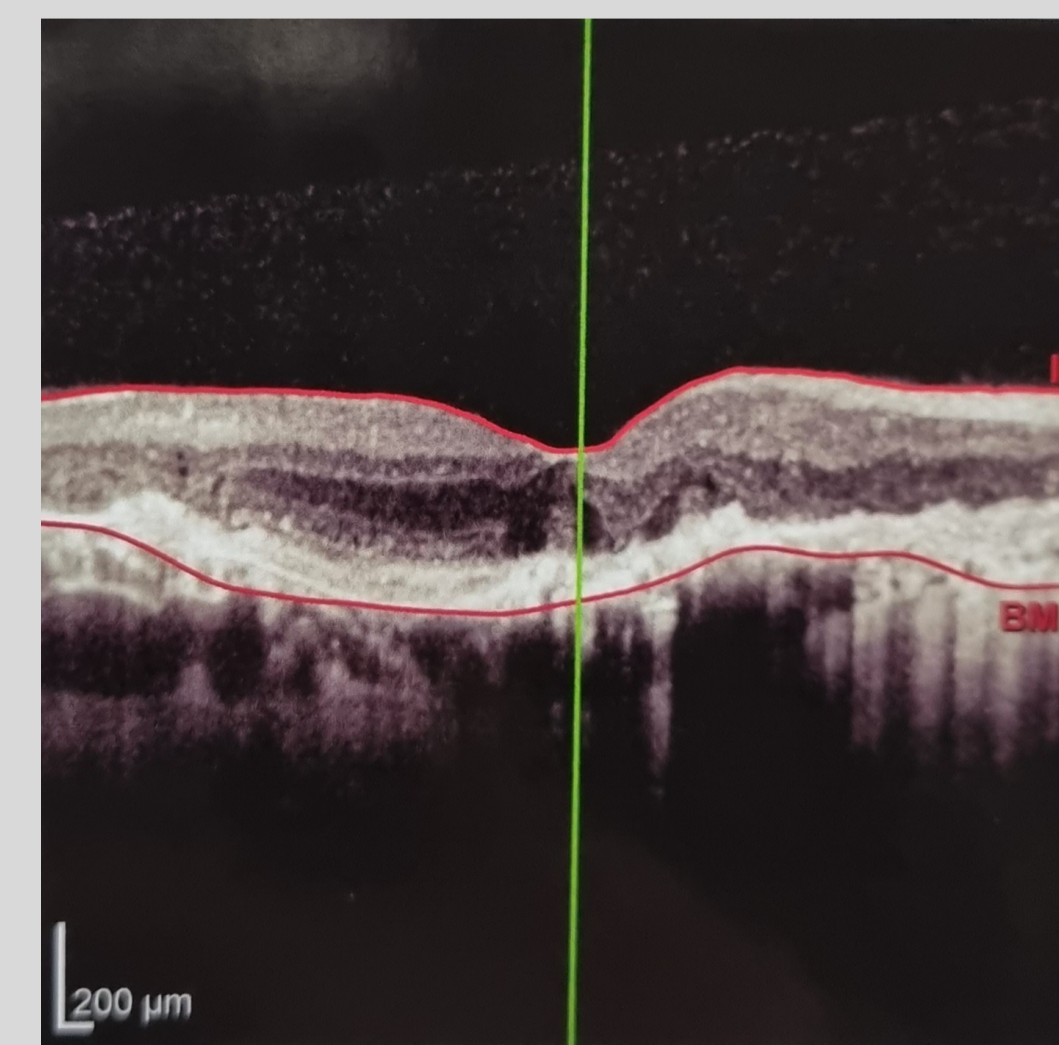


IMAGE 2: OCT OD

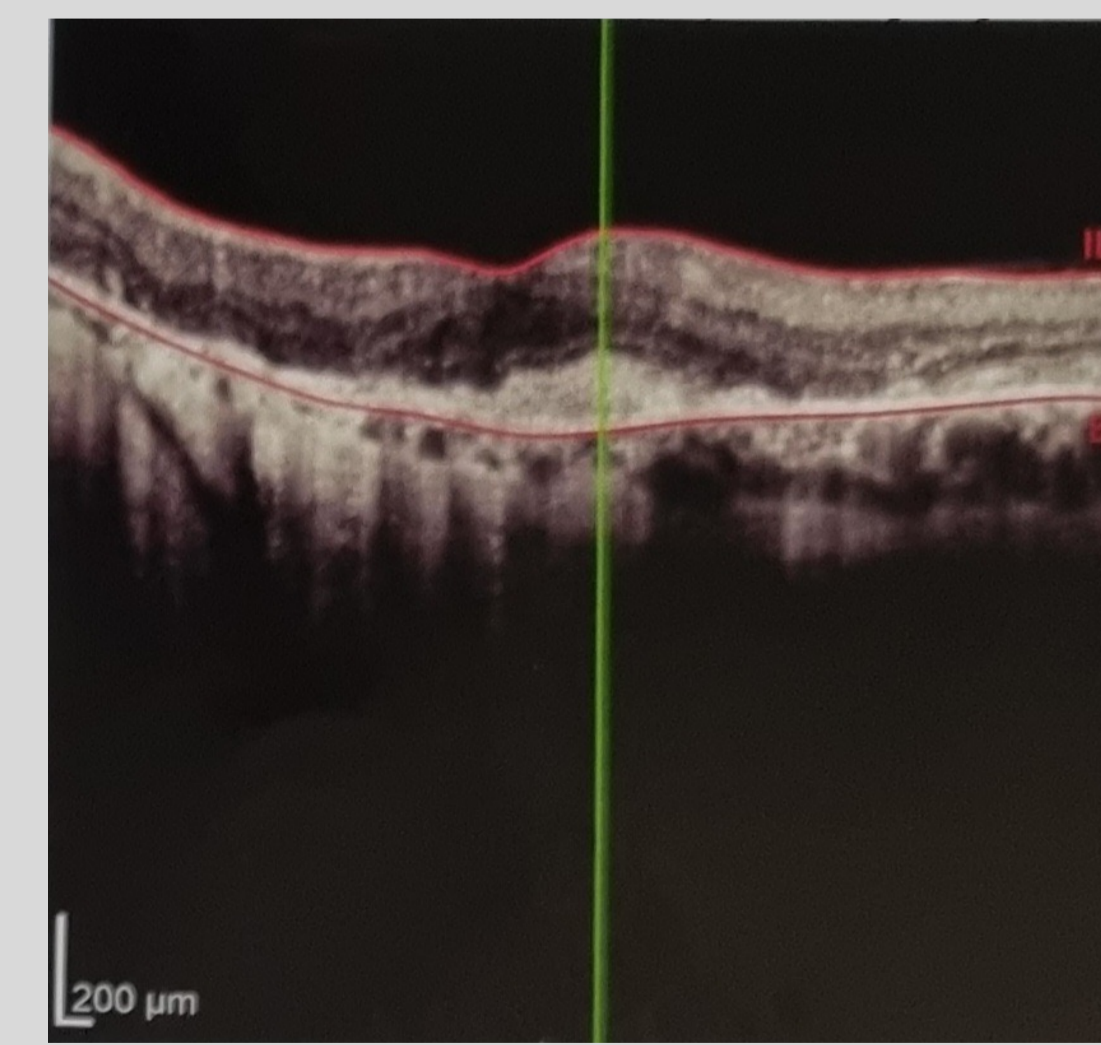


IMAGE 3: OCT OS