CLINICAL IMAGE

A CASE OF SKIN LAXITY

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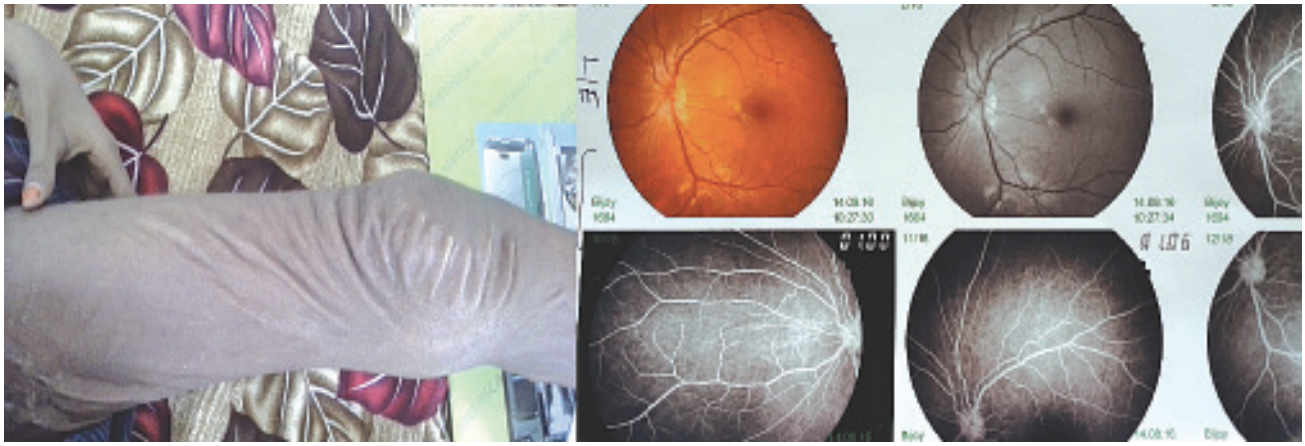

Master Bijoy, 16-year-old boy, second child of consanguineous marriage hailing from Comilla was admitted in Dhaka Medical College Hospital with the complaints multiple confluent papules mainly in the both armpits, flexural aspect of both elbow joints, groins, upper part of both legs, both angles of the lip and left side of the neck. They are not associated with itching, scaling or pain. He also experienced wrinkling of the skin over the left thigh and laxity of the skin of both armpits, flexural surface of elbow joint and groins. There was no scarring, hyperkeratosis or cicatrisation, no discharge or bleeding. Fundoscopy reveals evidence of optic nerve drusen. Evidence of angoid streak appearance of the left fundus. Thigh tissue for histopathology compatible with chronic abscess. Bone scintigraphy reveals dystrophic calcification. Skin biopsy reveals perforating pseudoxanthoma elaticum.

The term "pseudoxanthoma elasticum" was described by the French dermatologist Ferdinand-Jean Darier in1896[1]. Pseudoxanthoma elasticum (PXE) is a genetic metabolic disease with autosomal recessive inheritance caused by mutations in the *ABCC6* gene. The lack of functional ABCC6 protein leads to ectopic mineralization that is most apparent in the elastic tissues of the skin, eyes and blood vessels.[2]

References:

1. Darier J. Pseudoxanthoma elasticum. Monatshefte Prakt Derm. 1896; 23:609–17.
2. Dominique P. Germain. Orphanet Journal of Rare Diseases. 2017; 12:85.

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