

DERMATOPATHOLOGY

PERFORATING CALCIFIC ELASTOSIS- A FORGOTTEN ENTITY

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Introduction- Pseudoxanthoma elasticum though a inherited disorder has an acquired variety in the form of perforating calcific elastosis or periumbilical perforating pseudoxanthoma elasticum (PPPXE) which is most frequently found in obese, multiparous, middle-aged women and is characterized by lax, well-circumscribed, reticulated or cobble-stoned plaques over the periumbilical region with peripheral keratotic papules.

Case report- A 55-year-old non obese female, gravida 8, presented with pruritic plaque on the abdomen since 14 years. The lesions started over the umbilical region as a papule which within a span of 1 month lesions increased in number and size to burst open, discharging yellowish material and healed with scarring. Her family history was unremarkable.

On examination, multiple reticulated plaques with keratotic papules around the umbilicus and B/L iliac region with cobblestoning were present. Yellowish discharge was seen around the keratotic papules. The lesions were skin colored to violaceous with scarring of the involved area. Her laboratory investigations were normal. Abdominal ultrasonography, MRI and ocular examination revealed no abnormality.

Polarising dermoscopy from the centre of lesion showed reticulate pigmented network with yellowish background and hyperpigmentation at the border of the cobble stoned lesions. Erythema and telangiectasia with linear and branched vessels was also seen. The yellow structureless areas were visible at the edges as well.

Biopsy from a keratotic papule for histopathology with hematoxylin and eosin staining (H and E) revealed acanthotic epidermis with pseudoepitheliomatous hyperplasia. There were irregularly clumped, short, fragmented, basophilic elastic fibers in the mid-dermis. Staining with Ver hoeff's stain showed the fibers to be short, curled, and thick and stained positive for calcium with von Kossa stain.

On the basis of clinical manifestation, dermoscopy and biopsy diagnosis of perforating calcific elastosis was made and treatment in the form of topical retinoids were given.

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