

Periumbilical papules- A potentially sinister visage behind a benign facade with a surprise!

(Poster ID-9)

Tejas Vishwanath^a, Binny Binny^b, Kavita Ingle^b, Sunil Ghatge^b

^a- Department of Dermatology, Rajiv Gandhi Medical College and Chhatrapati Shivaji Maharaj Hospital, Kalwa, Thane-400605, India.

^b- Department of Dermatology, Rajawadi Hospital, Ghatkopar (East), Mumbai-400080, India.

Background:

An uncommon manifestation of pseudoxanthoma elasticum (PXE) is the **periumbilical variant** caused by stretching of abdominal wall connective tissue eg. obesity¹.

-Characterized by periumbilical **transepidermal elimination** of abnormal altered calcified elastic fibres.

- Presenting as indolent periumbilical papules, it can be confused with a variety of diseases and **may be associated with sinister internal manifestations like choroid streaks** which threaten eye sight in **22% cases**, **gastrointestinal bleeding** and **hypertension** among others

- **Histopathology** and recently **dermoscopy**² play important roles in diagnosis.

Cases:-

Case1: A 55 year old woman



- All three patients are **obese and multiparous**, presenting with **mildly pruritic** (cases 1 and 2) and **asymptomatic** (case 3) periumbilical yellow to erythematous and hyperpigmented papules with atrophic scarring. All were incidentally detected.

-1st degree relatives unaffected, no comorbidities.

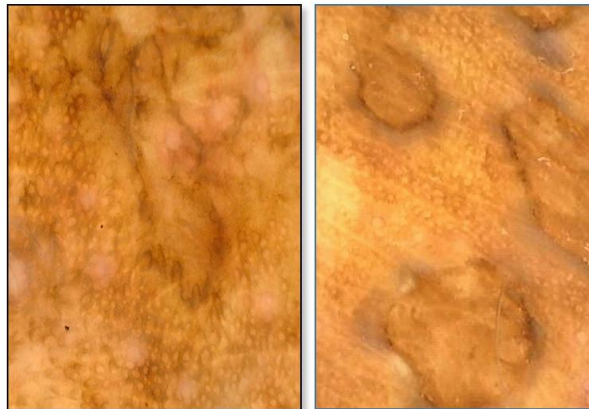
- Cases 1 and 2 are symptomatic since 5 years.

- Case 3 is has these complaints since **15 years**.

Dermoscopy (Dino-lite AM4113, polarized mode, 200x)

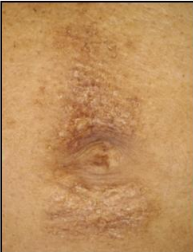
In all three cases

Case 2: A 60 year old woman



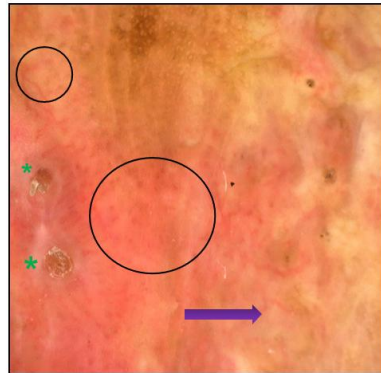
Multiple annular hyperpigmented lines

Case 3: A 65 year old woman



Dermoscopy

Cases 1 and 2



Keratotic plugs (*),
Telangiectasias (circle),
Brown background (arrow)

Differential diagnoses

1. Periumbilical pseudoxanthoma elasticum (PXE)
2. PXE like papillary dermal elastolysis
3. Focal dermal elastosis

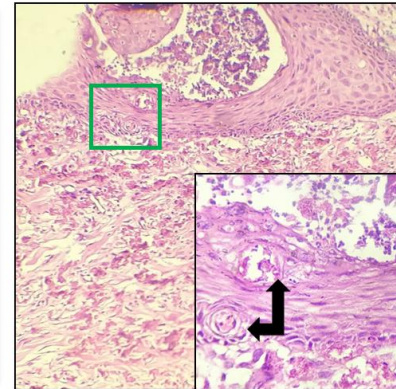
Beside and ancillary investigations:

Blood pressure- normal in all three cases

- Fundoscopy (for angioid streaks), serum calcium, ECG, stool examination, kidney function tests were within normal limits in all three patients.

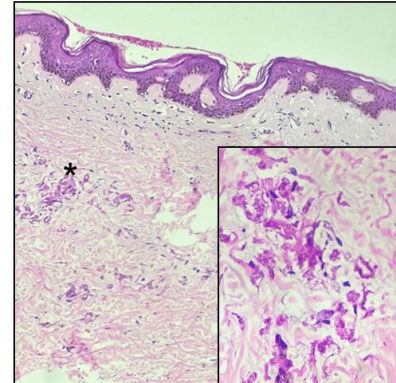
Histopathology

Cases 1 and 2:

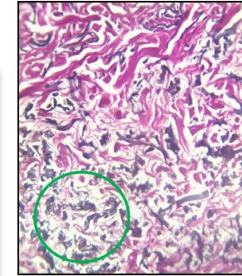


-**Transepidermal elimination (TE)** of altered upper dermal **ravelled wool elastic fibres** (square, inset- arrow), upper dermal lymphocytic infiltrate

Case 3- **ABSENCE** of TE of ravelled wool elastic fibres (*)

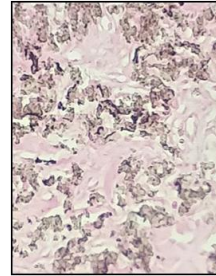


Verhoeff-Van Gieson stain



Ravelled wool elastic fibres (circled)

Von Kossa stain



Calcium deposits (black)

Dermoscopic feature	Histologic correlate
Keratotic plug (green *)	Transepidermal elimination of altered elastic fibres
Telangiectasias, dotted vessels (circled)	Vasodilation and inflammation in upper dermis
Yellow-brown background (purple arrow), arcuate hyperpigmented lines	Altered elastic fibres in upper and mid dermis

Diagnosis- Periumbilical Pseudoxanthoma elasticum

Learning points-

- 1) Role of histopathology (**confirmatory**), special stains and dermoscopy (**supportive**).
- 2) **Long standing non perforating variant** (very rare).

References:

- 1.Vishwanath T, Nagpal A, Shinde G. Periumbilical Papules in a Middle-aged Woman. JAMA Dermatol. Published online October 23, 2019. doi:10.1001/jamadermatol.2019.2963. (epub ahead of print).
2. Jha A, Zeeshan M, Sinha B, Singh A, Agrawal P. Periumbilical perforating pseudoxanthoma elasticum: a rare case report. Dermatology Practical & Conceptual. 2018;8(2).