


An Unusual Cause of Hyperkeratosis

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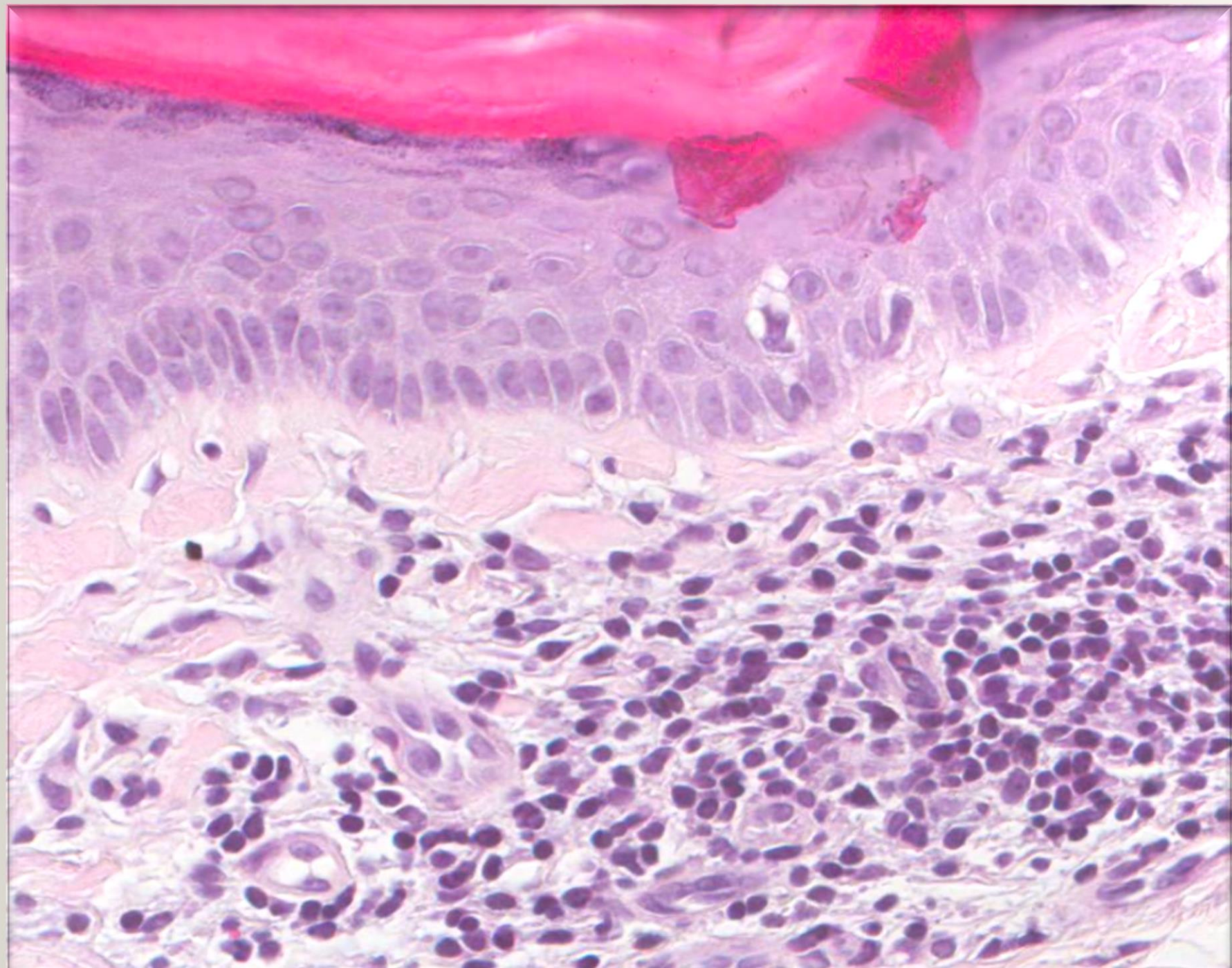
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Clinical Presentation

- 43-year-old 
- Two and a half year history
- Pink/light brown papules, some of which appeared scaly and keratotic
- Started on her ankle and lower legs and spread to her thighs, arms and neck

Clinical Presentation



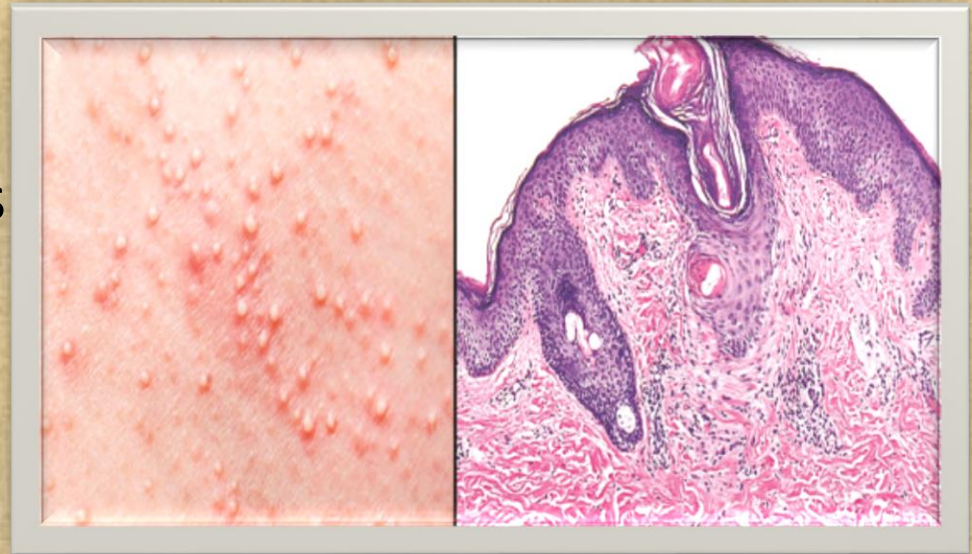


Differential Diagnosis

- 1) Keratosis pilaris
- 2) Lichen planus
- 3) Hyperkeratosis follicularis et parafofollicularis in cutem penetrans (Kyrle's disease)
- 4) Hyperkeratosis lenticularis perstans (Flegel's disease)
- 5) Lichenoid drug reaction

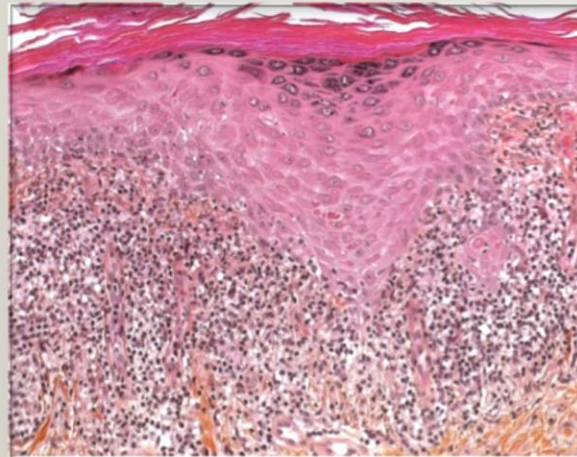
Keratosis Pilaris ¹

- Peak in adolescence
- Association with ichthyosis vulgaris
- Pruritic, follicular papules & pustules
- Lateral aspect arms & thighs
- Histology – follicular dilatation & keratin plugs
- May contain distorted hair shafts



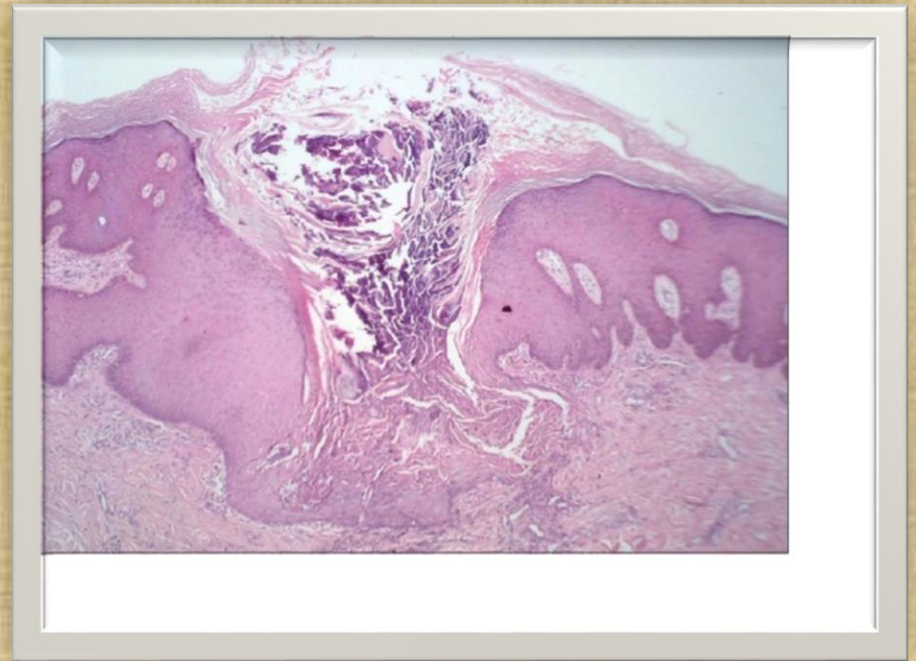
Lichen Planus ³

- Violaceous papules
- Intensely pruritic
- Flexor aspects wrists & forearms, extensor aspects hand & ankles
- Histology - Interface dermatitis
- Irregular acanthosis
- (Wedge-shaped) Hypergranulosis



Kyrle's Disease ⁵

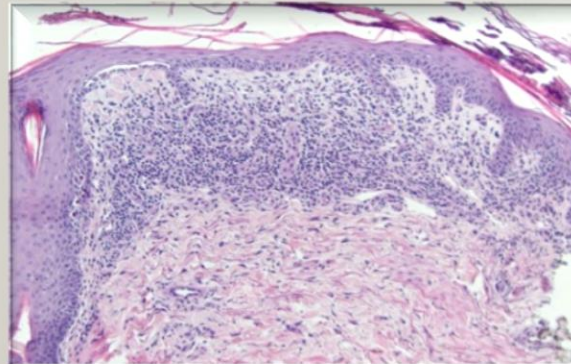
- Similar clinical presentation to our patient (age, gender, distribution), palms & soles classically spared
- Histology- epidermal invagination containing keratotic plug & basophilic debris
- Eventual perforation into dermis with associated foreign-body reaction & subsequent transepidermal elimination



Lichenoid Drug Reaction ⁷

- Clinically similar presentation to lichen planus
- Gold, anti-malarials, penicillamine, captopril, β -blockers, TNF- α inhibitors
- May be a long latent period between starting the drug & development of a rash
- Histology similar to lichen planus

8



Flegel's Disease ⁹

- Hyperkeratosis lenticularis perstans – described in 1958
- M=F, 4th – 5th decade
- Characterised by protracted clinical course (years to decades)
- Most cases sporadic, some evidence of AD inheritance in a subset of cases
- Unknown aetiology

Flegel's Disease ⁹

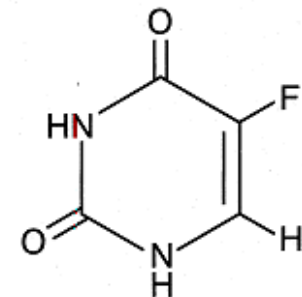
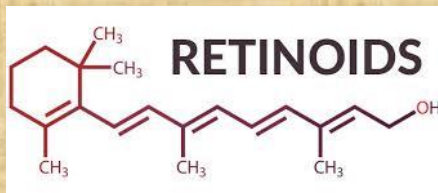
- Large numbers, small, discreet, grey/grey-brown/red-brown, circular papules with a well-developed scale
- Dorsum of foot, lower legs, upper arms, pinnae, buttocks, trunk, dorsal aspects of hands, palms & soles
- Removal of the scale associated with pinpoint bleeding

Flegel's Disease ⁹

- ❖ Histologically characterised by focal areas of abnormal hyperkeratinisation (**sharp cut-off from adjacent epidermis**)
- Hyperkeratosis
- Occasional parakeratosis
- Epidermal atrophy
- Loss of granular cell layer
- Lichenoid/perivascular chronic inflammatory infiltrate in papillary dermis
- Basal vacuolar degeneration, occasional cytoid bodies

Flegel's Disease

- A number of treatments have been used with varying degrees of response
- At present all of these are considered unsatisfactory due to high rates of recurrence¹⁰



5-Fluorouracil

References

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Questions?

