Case 4

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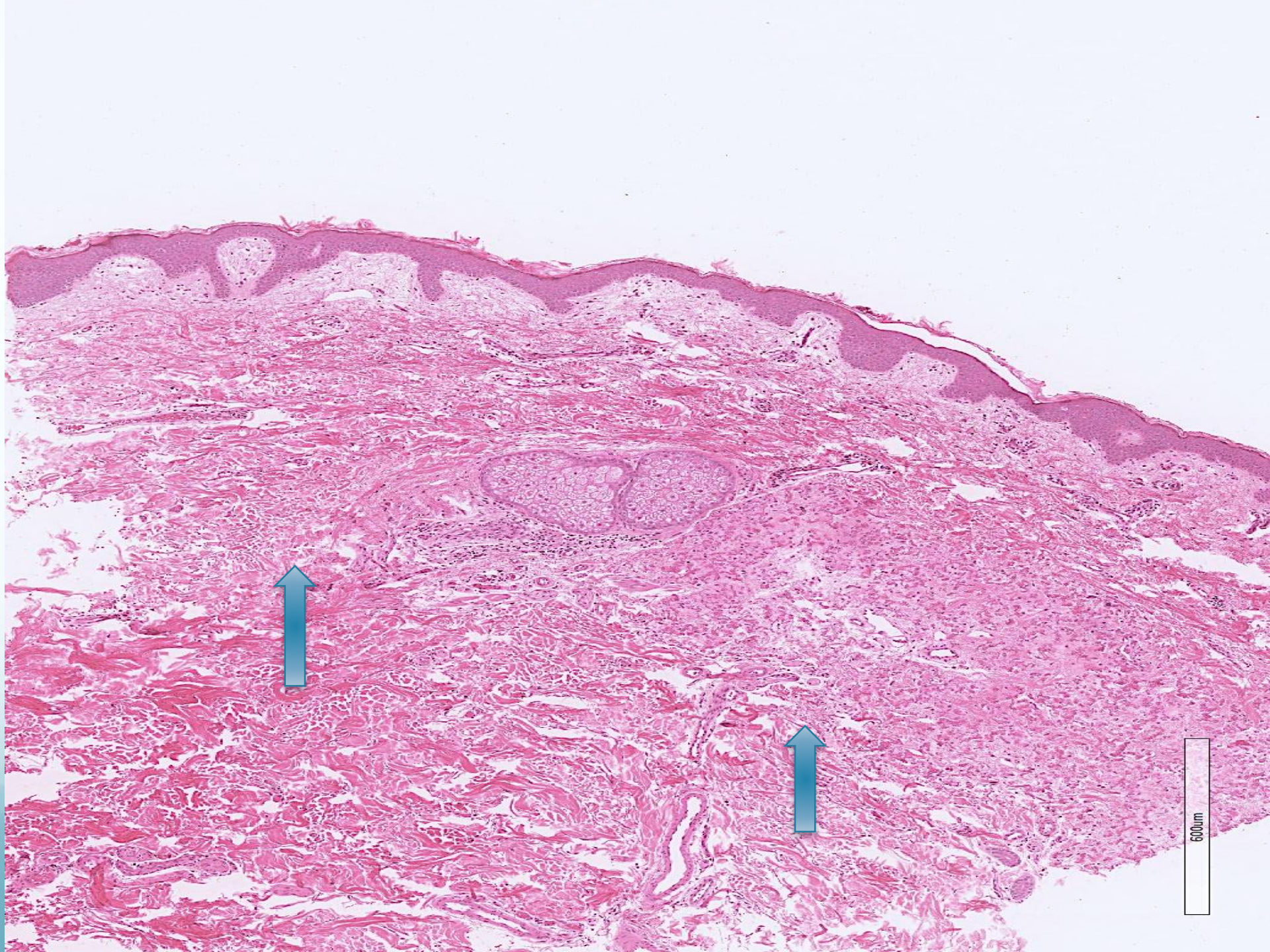
- Clinical History: 44 M
- Few areas on the skin of the back of neck with a ‘plucked chicken appearance’.
- Punch biopsy left side of the neck.


2mm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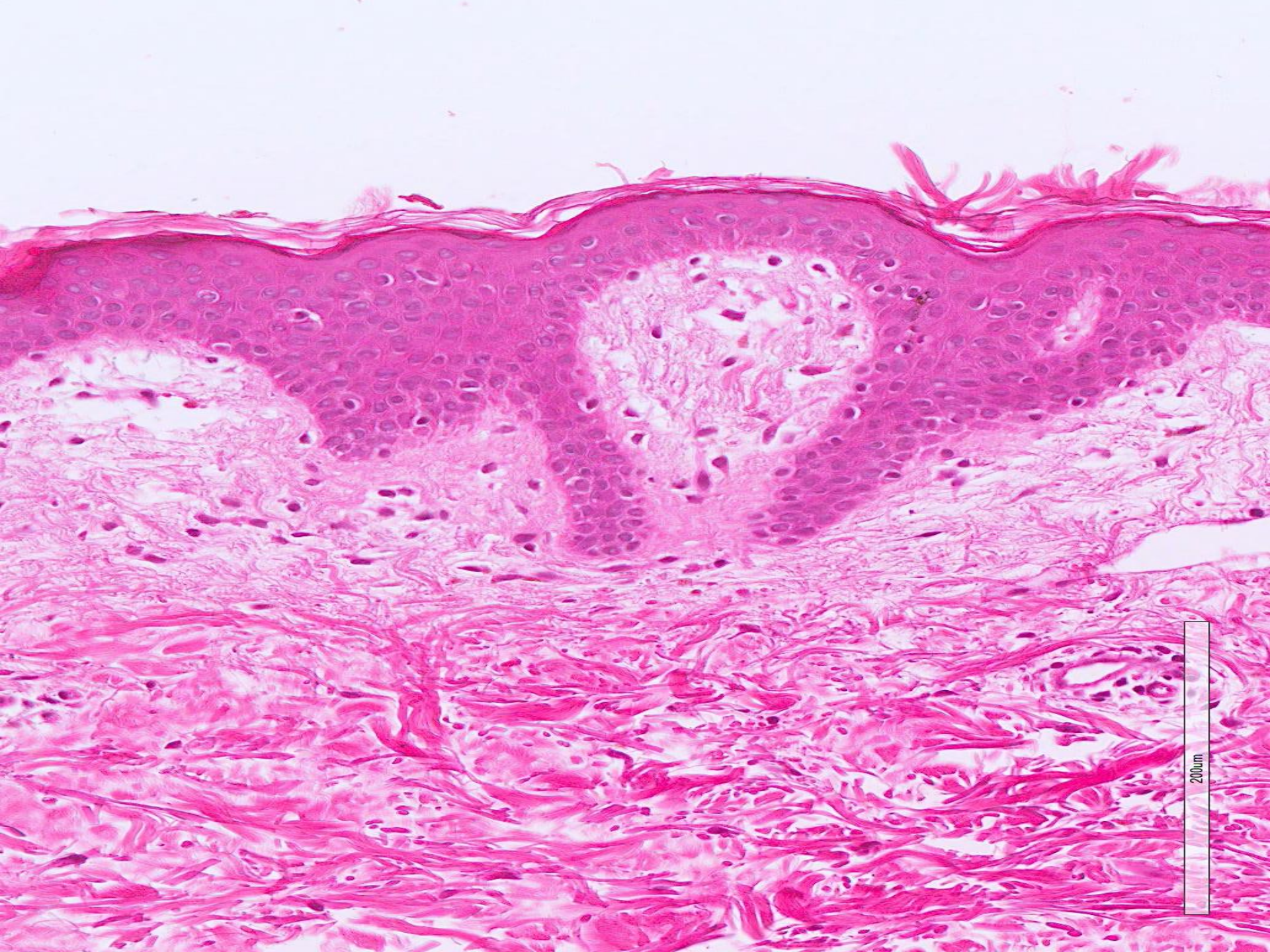
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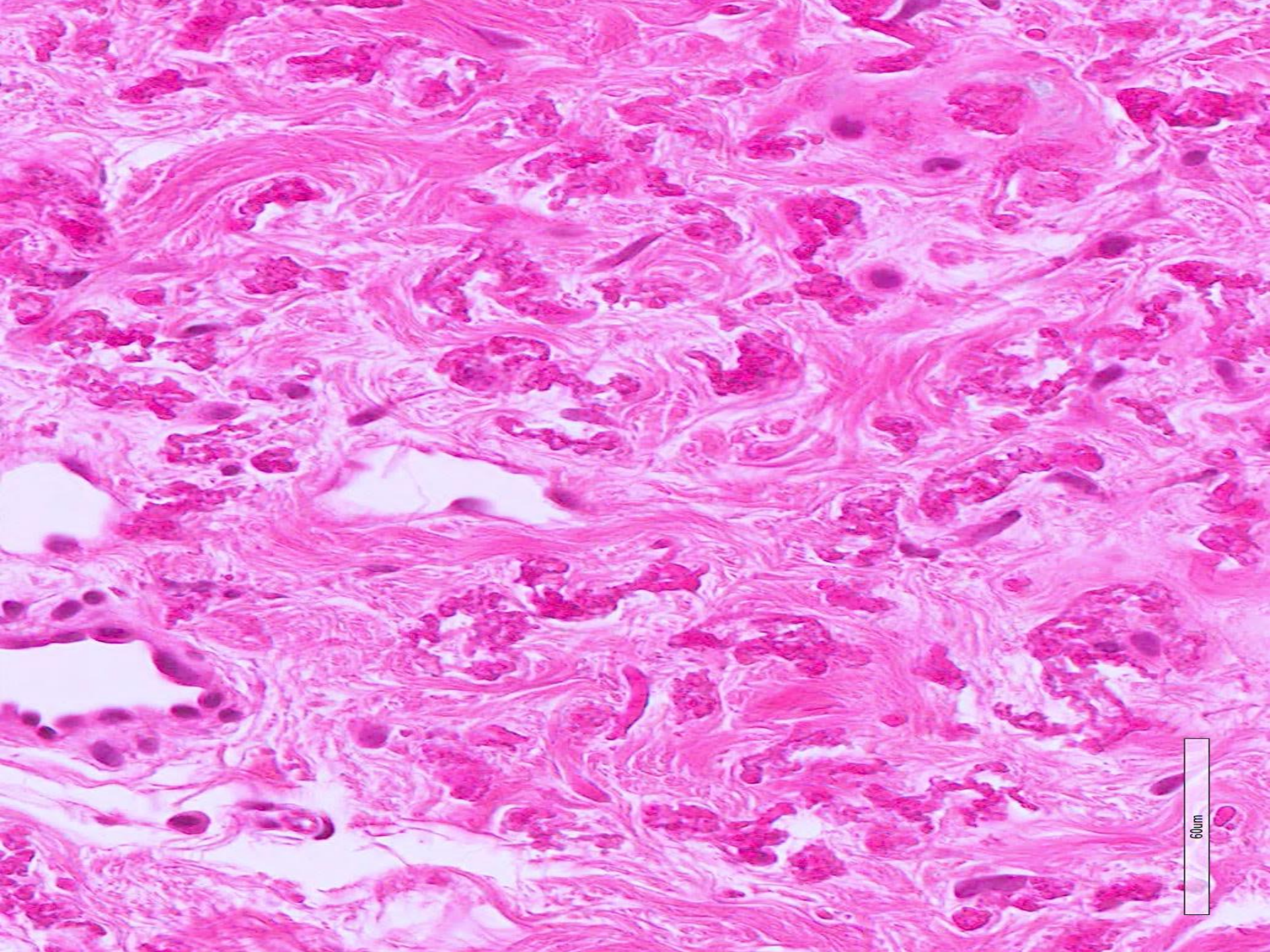
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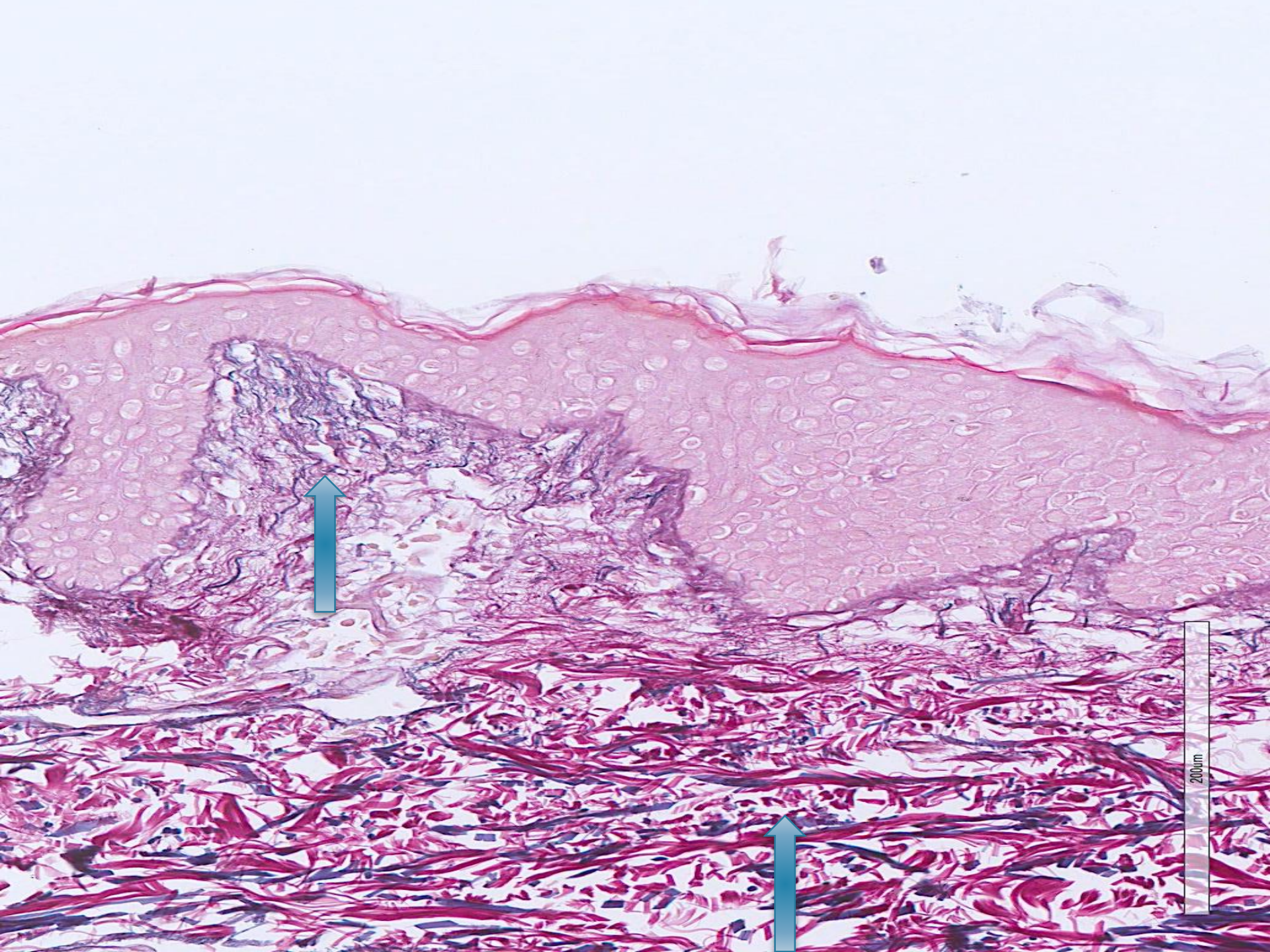
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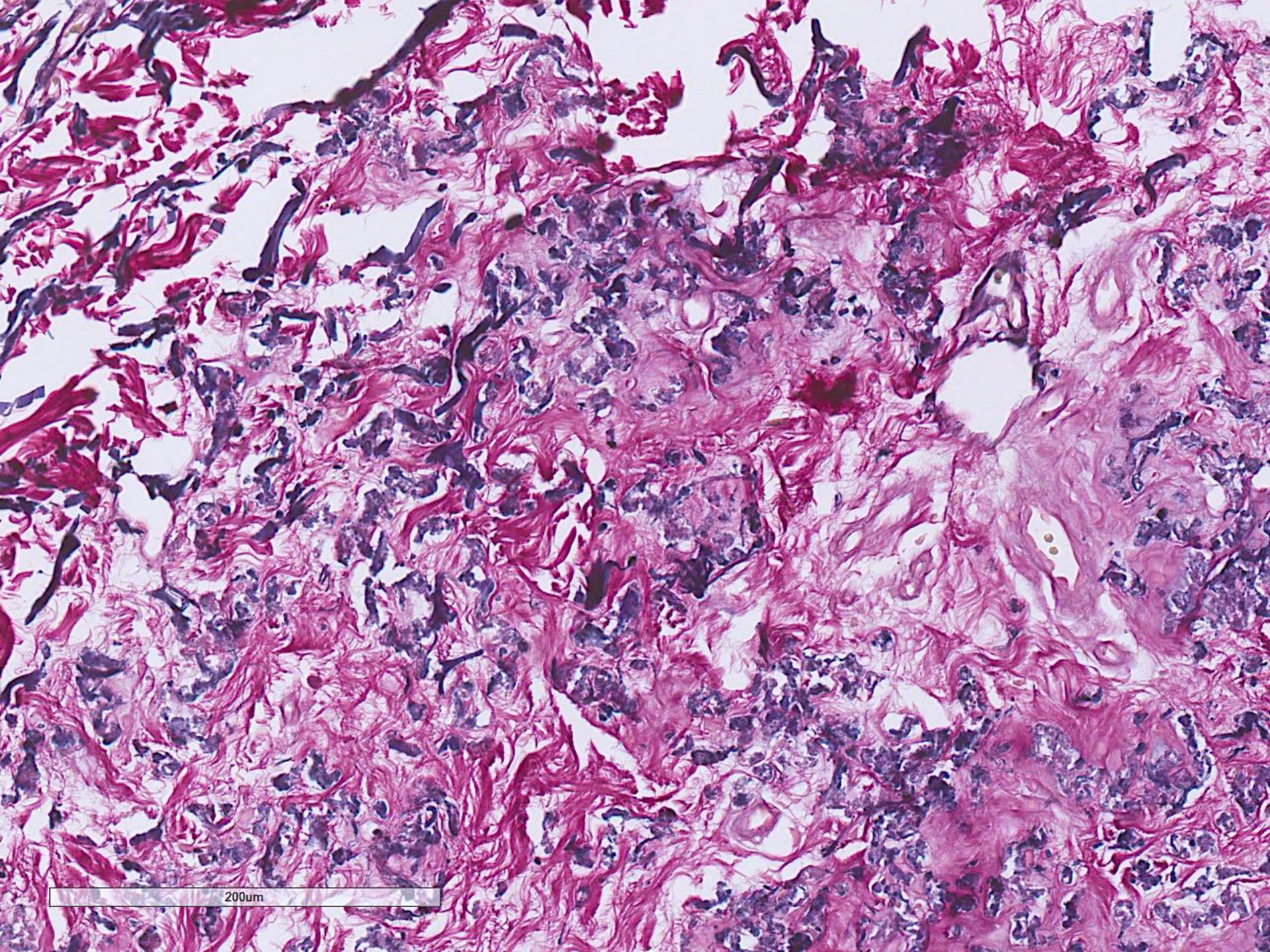
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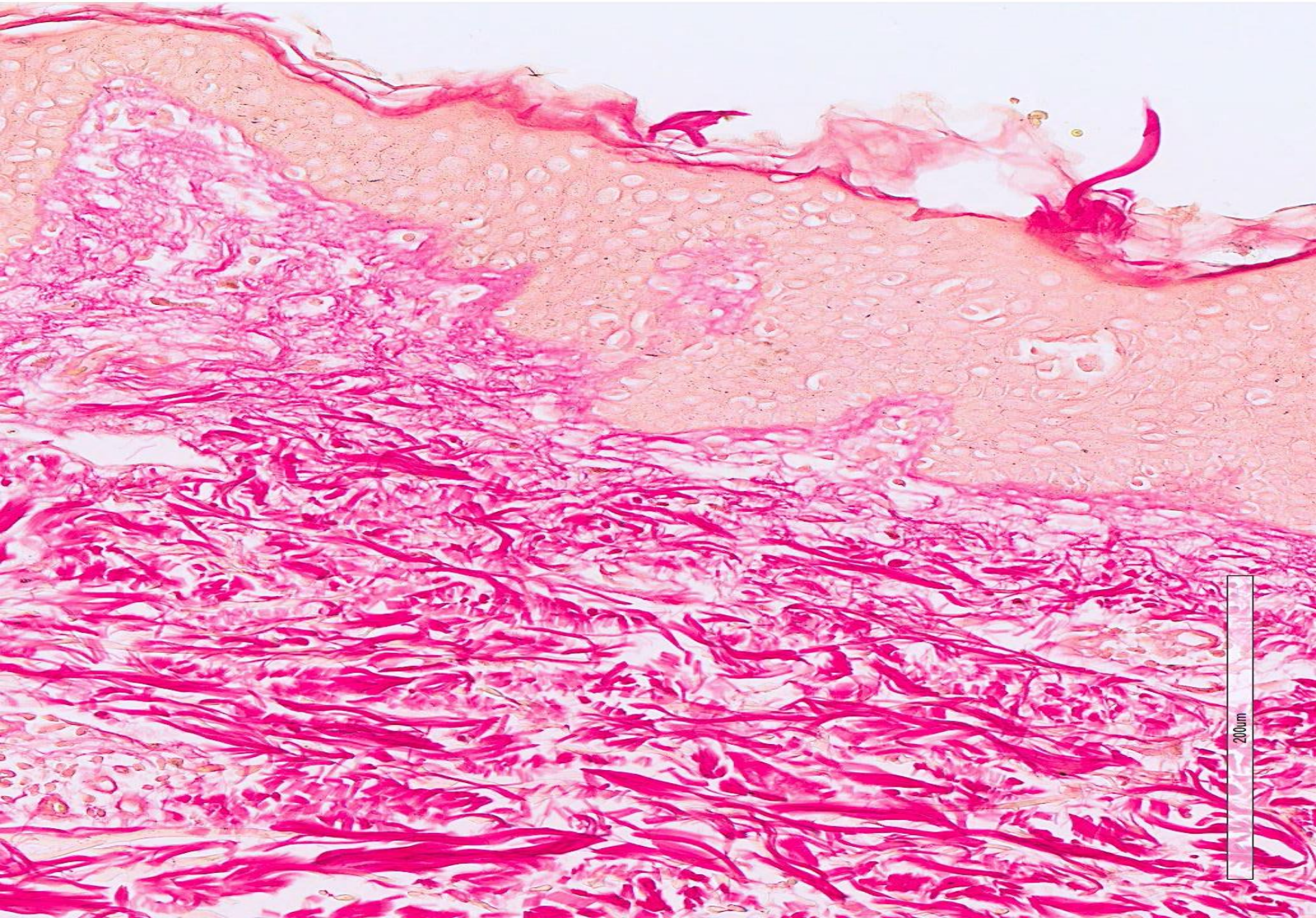
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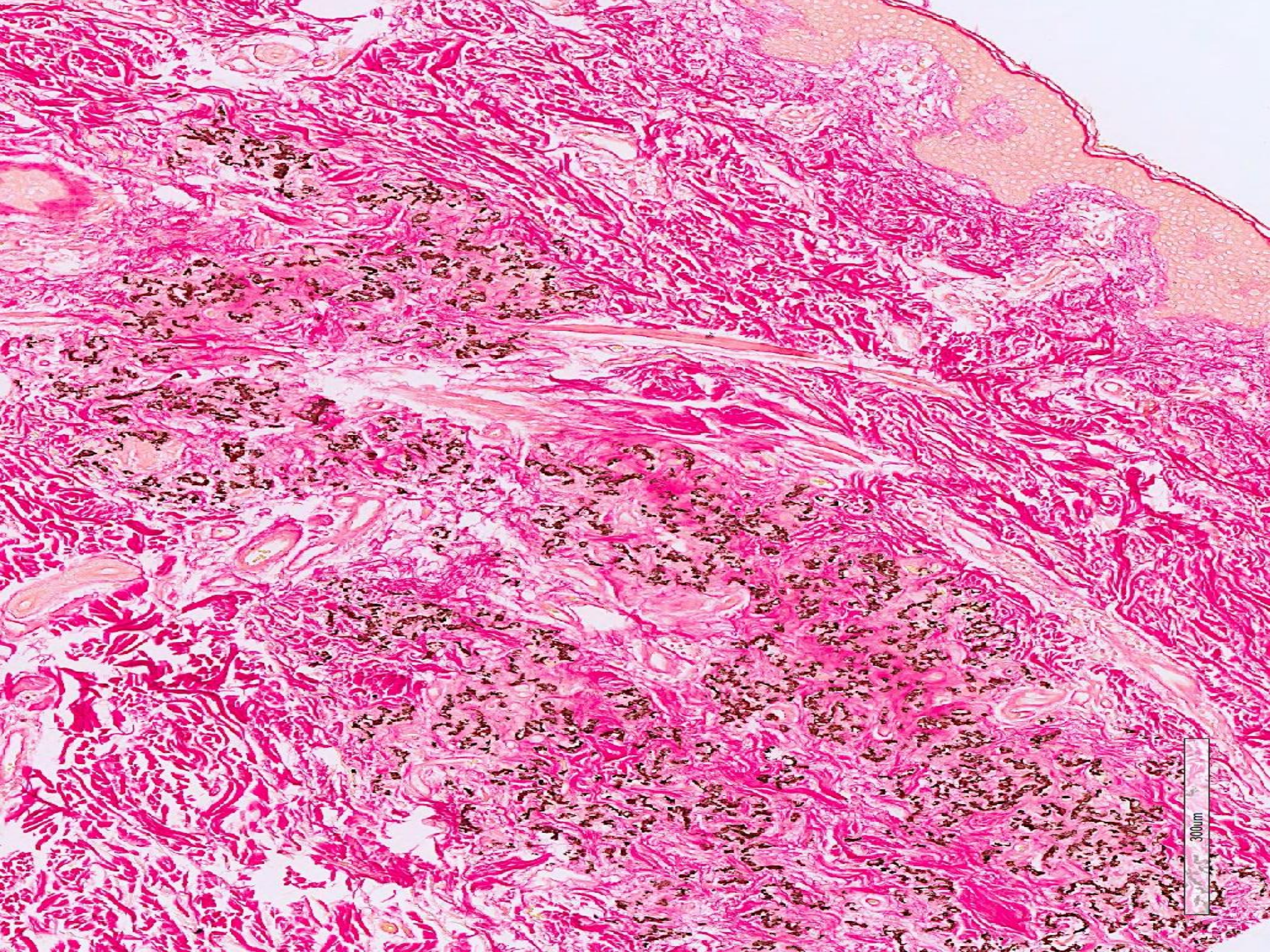
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DD

- 1: Calciphylaxis
- 2: Focal dermal elastosis
- 3: Mid-dermal elastolysis
- 4: PXE-like papillary dermal elastolysis
- 5: Pseudoxanthoma elasticum

Calciphylaxis

- Calcification of small to medium sized arteries and arterioles.

- Different clinical picture.

Focal dermal elastosis

- Rare disorder of elastic tissue characterised by a yellowish papular eruption.
- Elderly.
- Sides of neck and flexural areas.

- Local accumulation of elastic fibers in the mid and deep reticular dermis.
- Elastic fibers are normal!

Mid-dermal elastolysis

- Most commonly affected sites are trunk and upper arms
- 3 clinical subtypes of mid-dermal elastolysis described.
- Type I, the most common subtype, presents with asymptomatic, well-demarcated areas of fine wrinkling
- Type II, presents with looseness of skin around hair follicles, resulting in perifollicular papules
- Type III, presents with reticular erythema

- ***Biopsy***-
- Normal
- Patchy inflammation & loss of elastic fibers within the mid-dermis (EVG)

PXE-like papillary dermal elastolysis

- Rare, 40 cases
- Women in late adulthood.
- An acquired disorder characterised by multiple , asymptomatic or pruritic, yellow/skin coloured papules .

- Atrophic epidermis and band-like loss of elastic tissue in the papillary dermis.
- Clumping and fragmentation of elastic fibers may also be seen.

This Bx-

- Normal elastic fibers within the papillary and deep-dermis.
- Elastic fibers within the mid-dermis are polymorphous, mineralized and fragmented.
- Pseudoxanthoma elasticum

- Angioid streaks in the eyes. Normal vision.

- Cardiology clinic: Normal ECG, no significant murmurs

- PXE (Gronblad-Strandberg Syndrome) is a generalised degenerative disease of elastic tissue , AD inheritance.
- Prevalence varies 1:50000-70000
- Onset 2nd decade

- Characterized by dystrophic mineralization and fragmentation of elastic fibers and causes-

- 1. dermal (papular lesions flexural areas)
- 2. ocular (angioid streaks, subretinal neovascularization, and haemorrhage), and
- 3. vascular symptoms (coronary and peripheral vascular disease)

- *There is a striking variation in phenotypic expression*.
- Caused by mutations in the ABCC6 (ATP-binding casette subfamily C member 6) gene encoding a transmember transporter protein. The exact relation between the ABCC6 transporter & the elastic fiber abnormalities remains unclear.

References

- Laube S, Moss C Pseudoxanthoma elasticum Archives of Disease in Childhood 2005;90:754-756.

- M J. Hosen, A Lamoen, A Paepe, and O M. Vanakker, “Histopathology of Pseudoxanthoma Elasticum and Related Disorders: Histological Hallmarks and Diagnostic Clues,” *Scientifica*, vol. 2012, Article ID 598262, 15 pages, 2012. doi:10.6064/2012/598262


THANK YOU