Case 3

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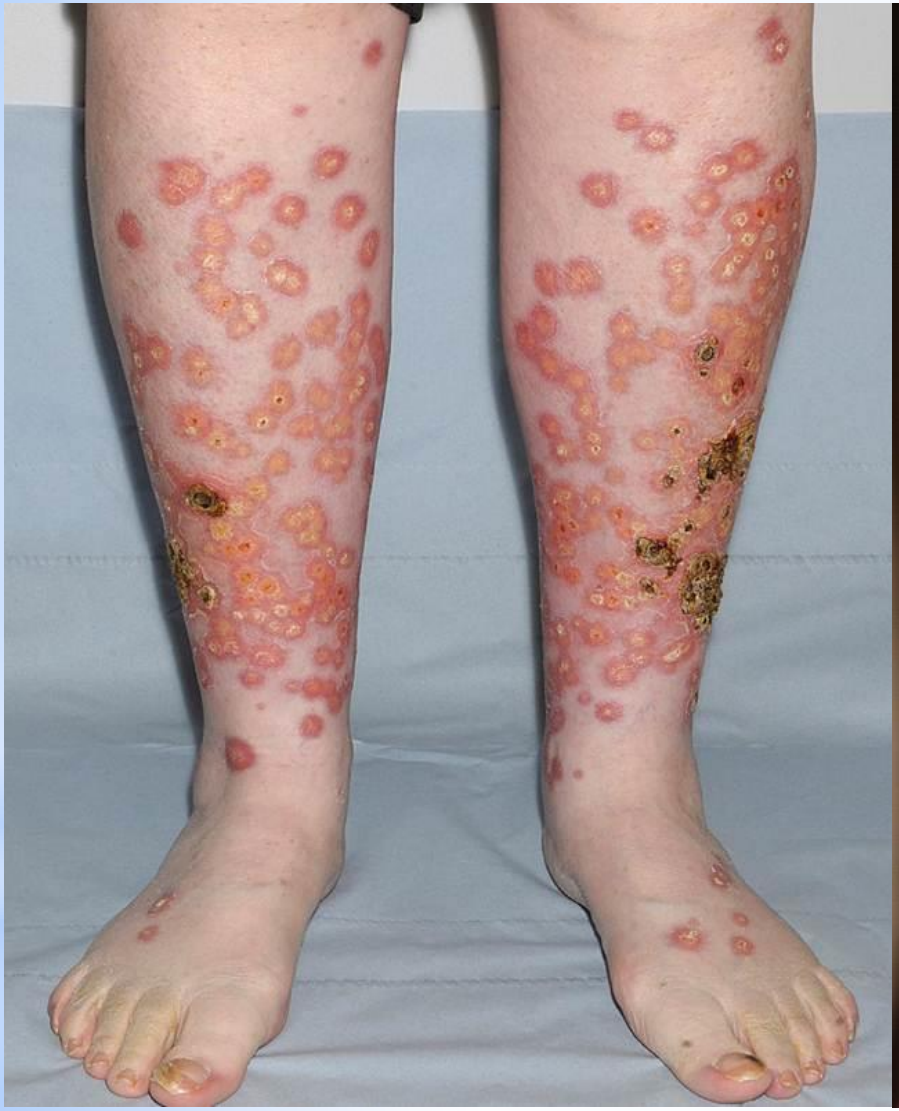
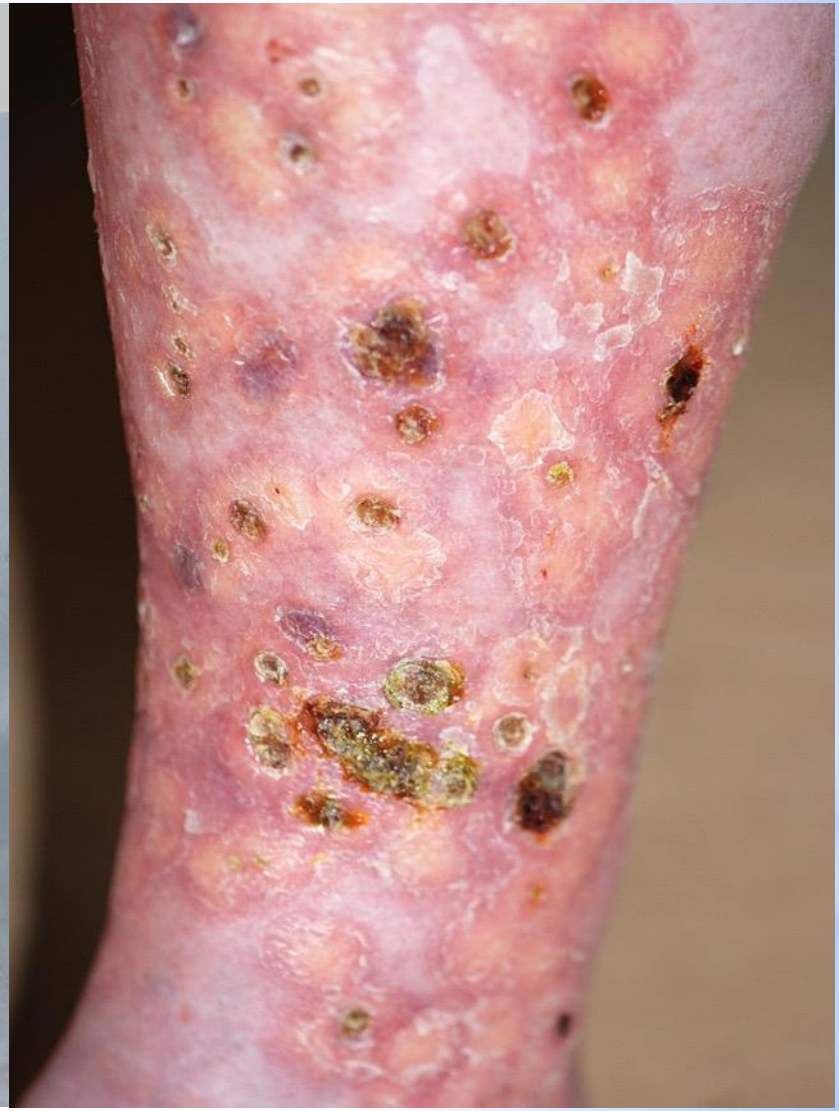
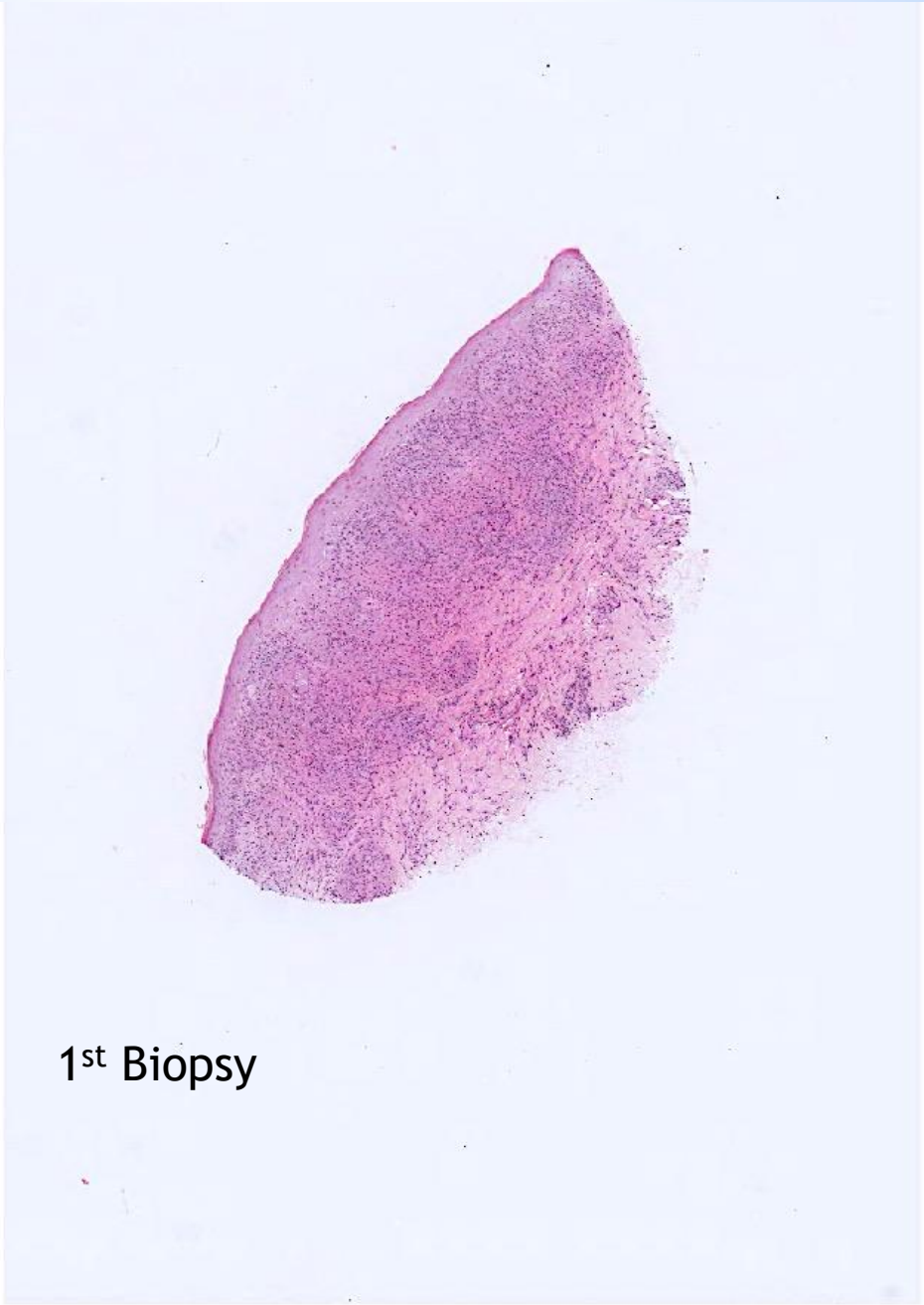
* 50 F, known to have lichen plano pilaris

* Monomorphic, painful papules and plaques on lower legs.

* Punch biopsy lower leg

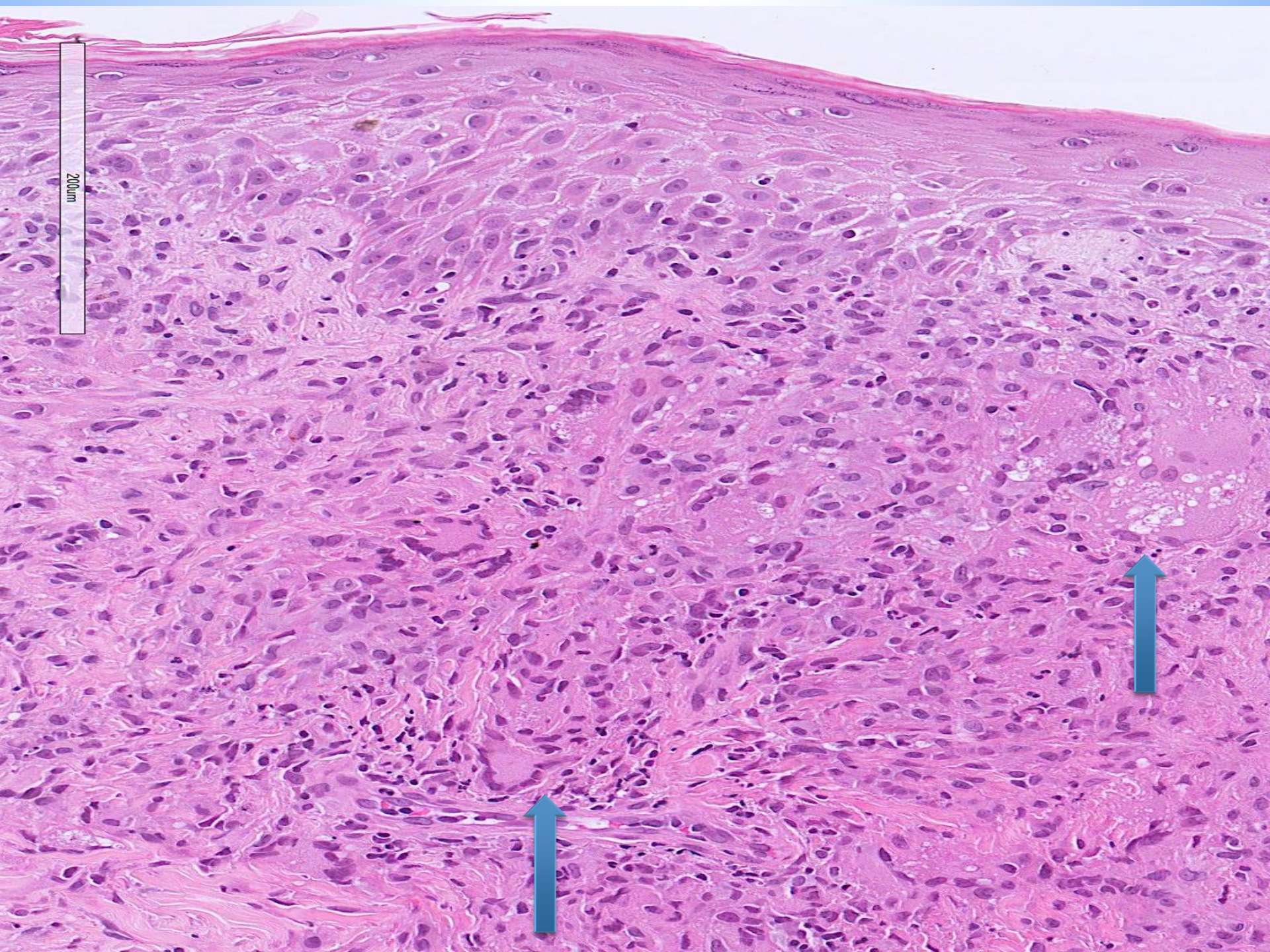






1st Biopsy

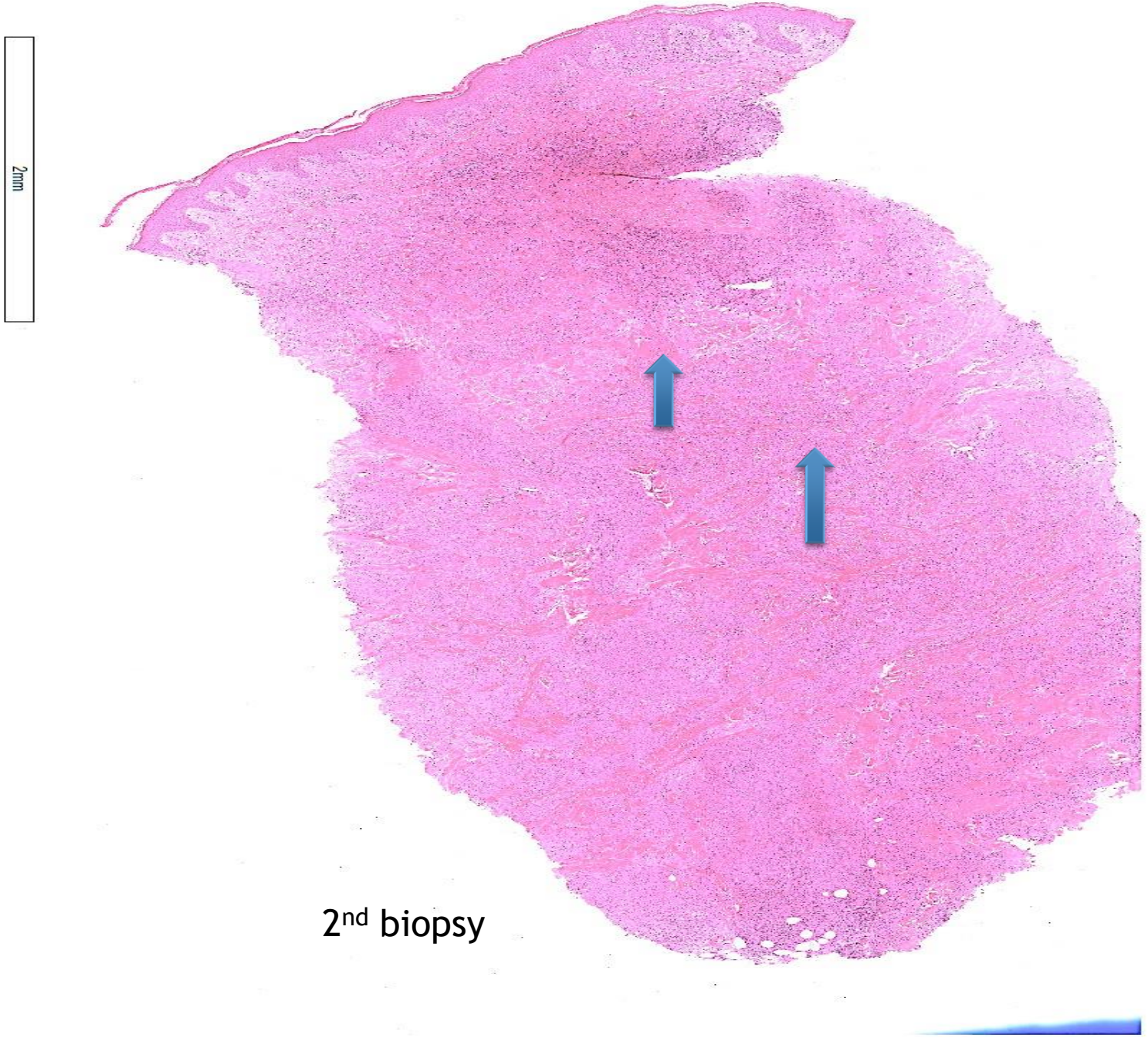
2mm


200um

* Shallow punch biopsy
* Granulomatous infiltrate

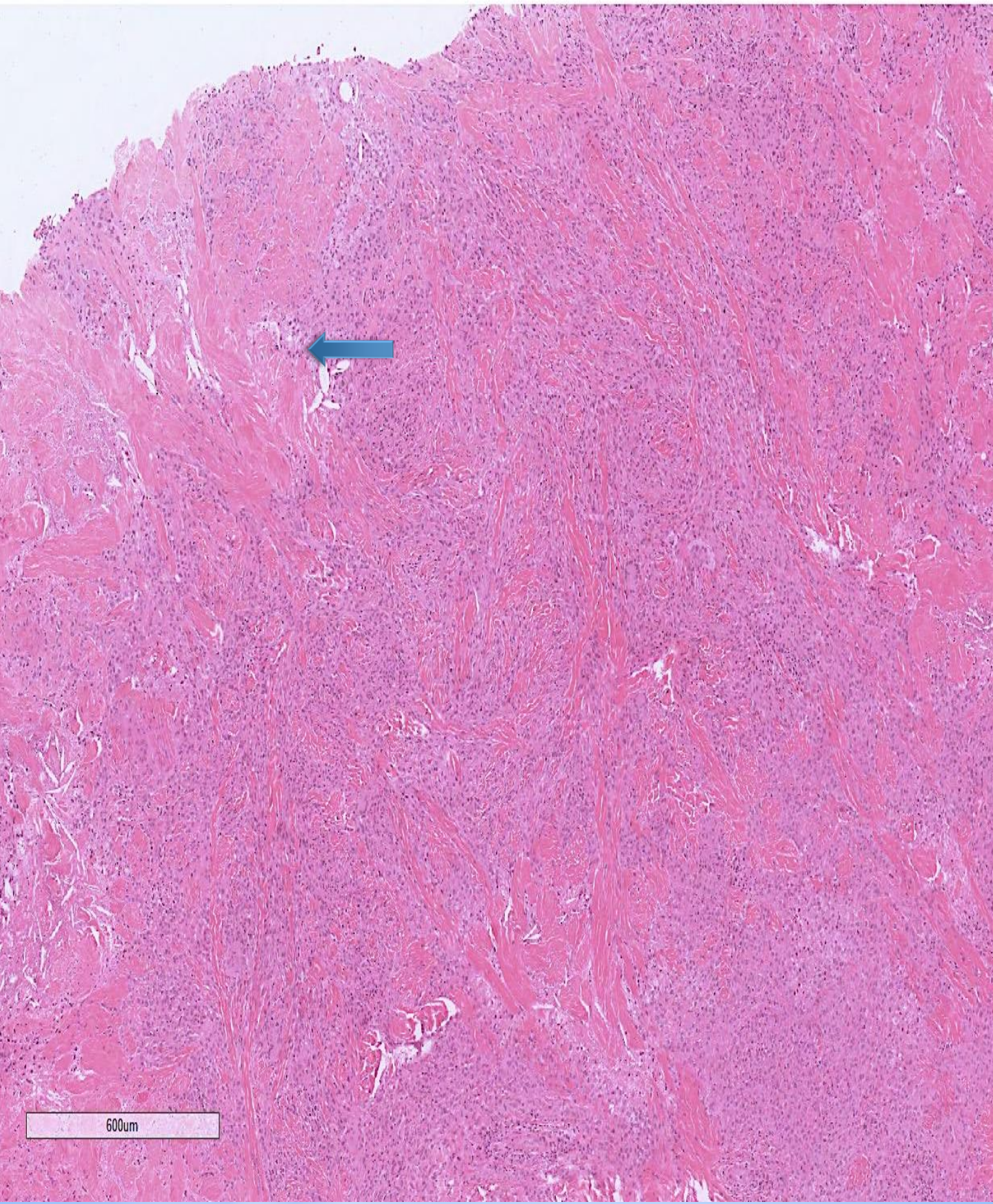
* JXG

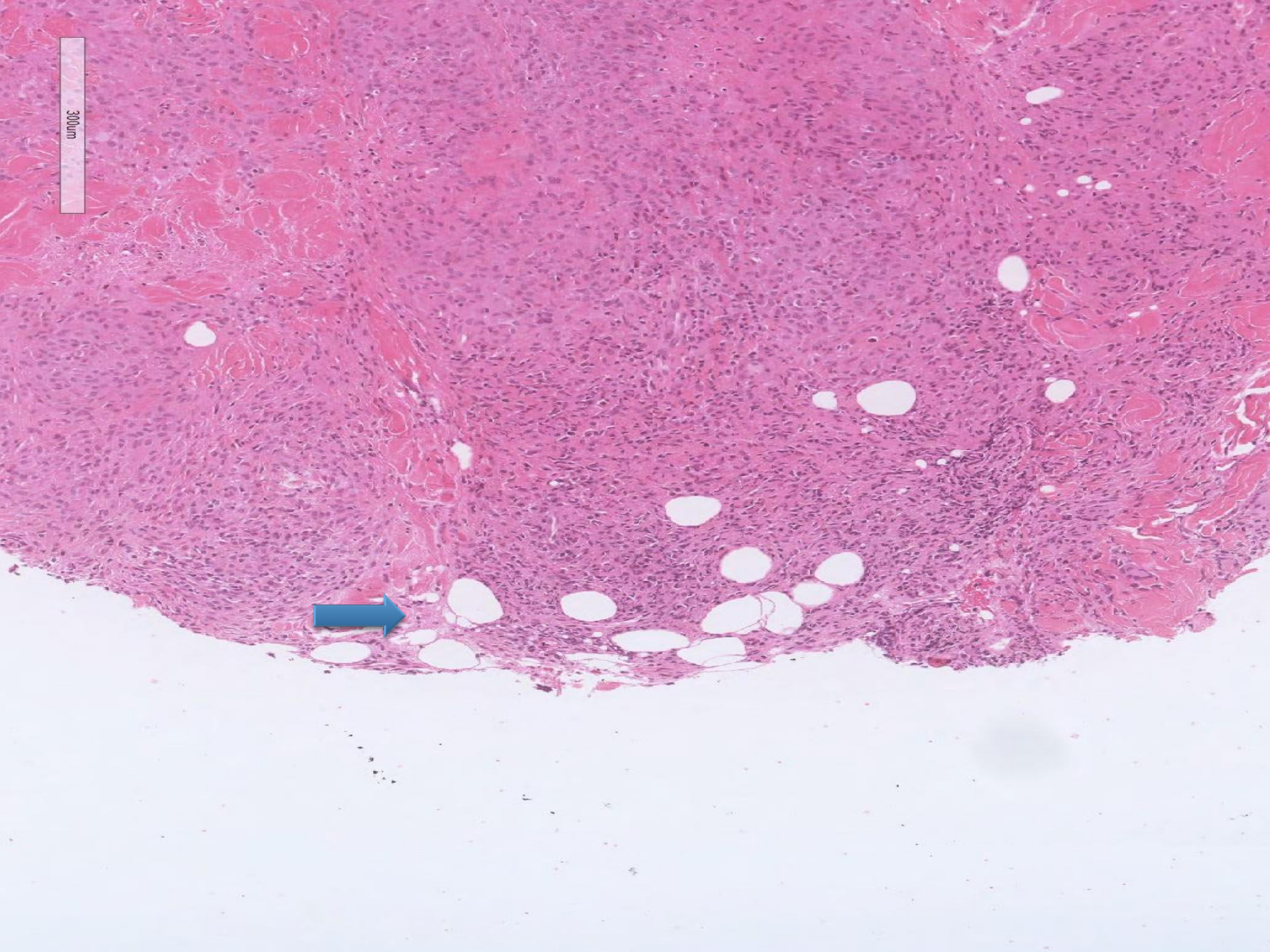
* Please repeat biopsy

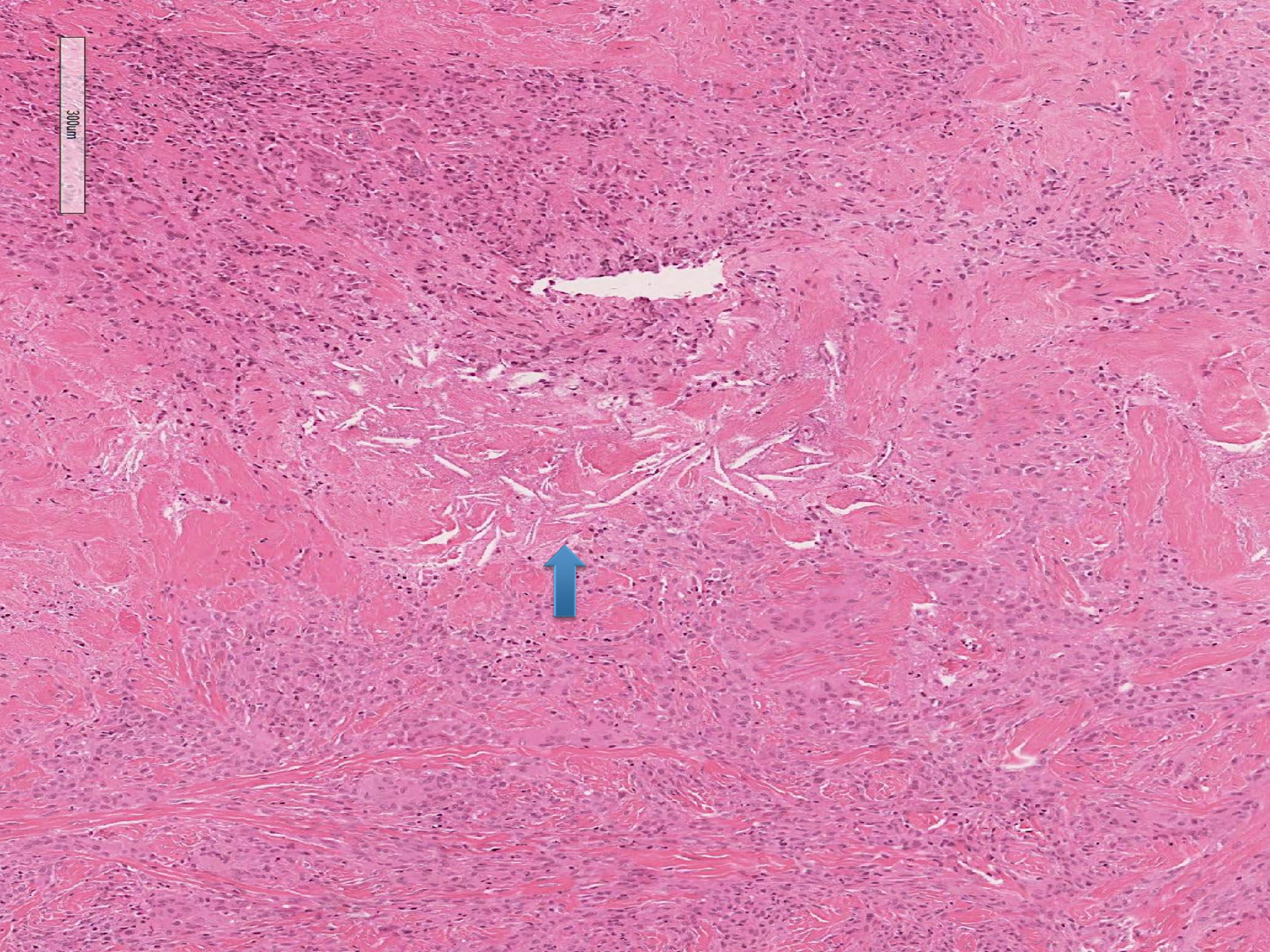


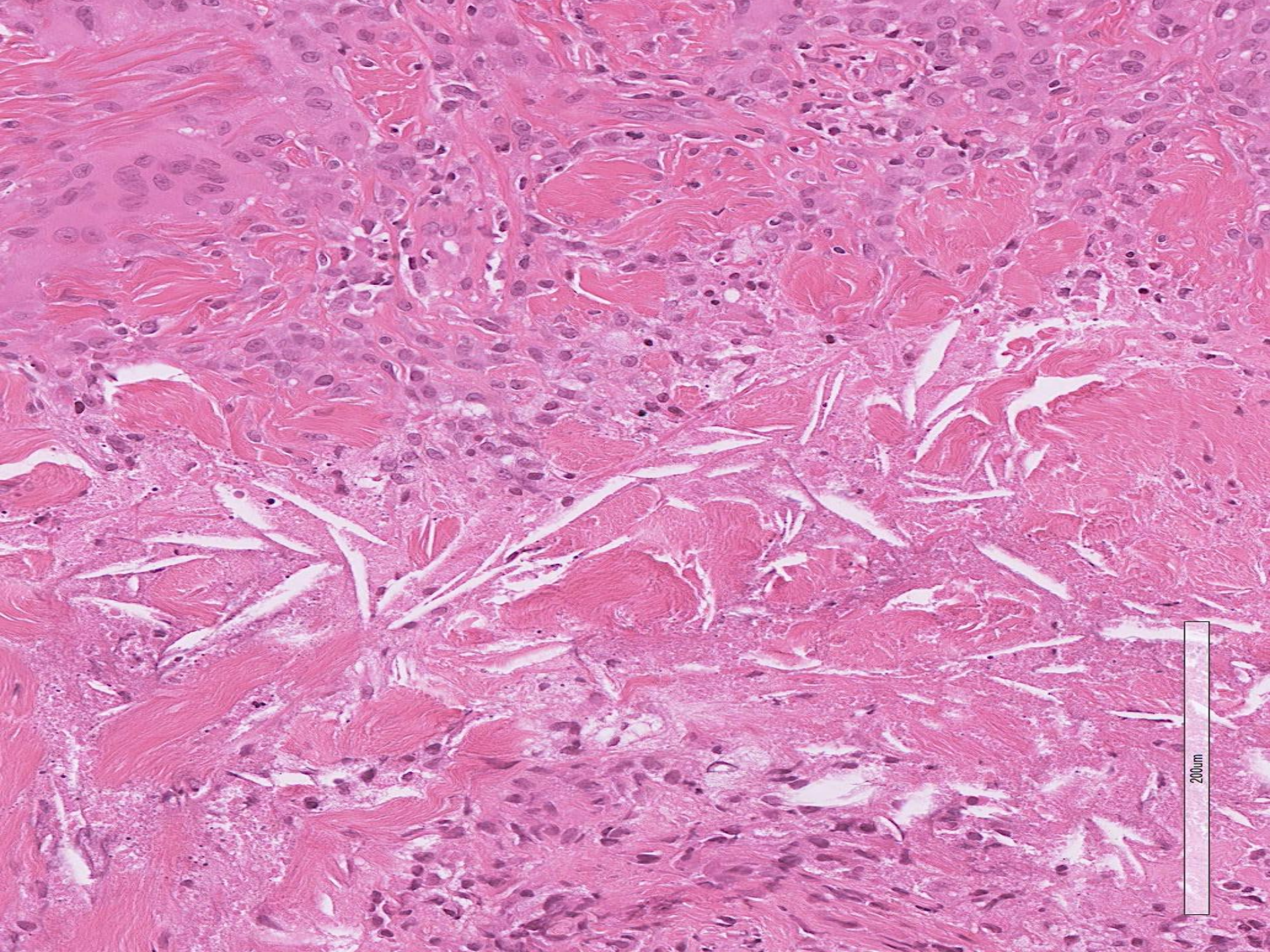
2nd biopsy

600um


600um


300um


300µm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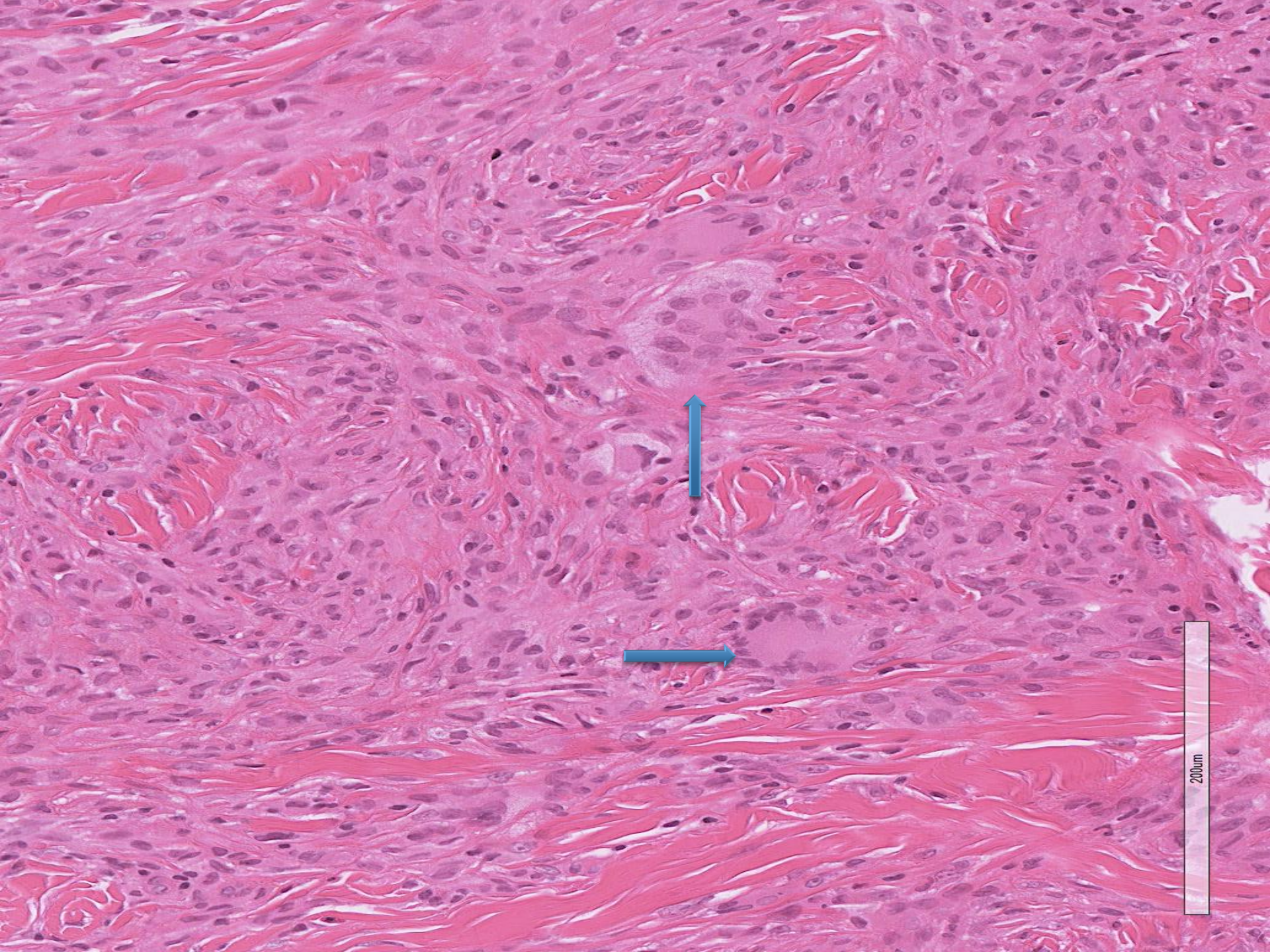
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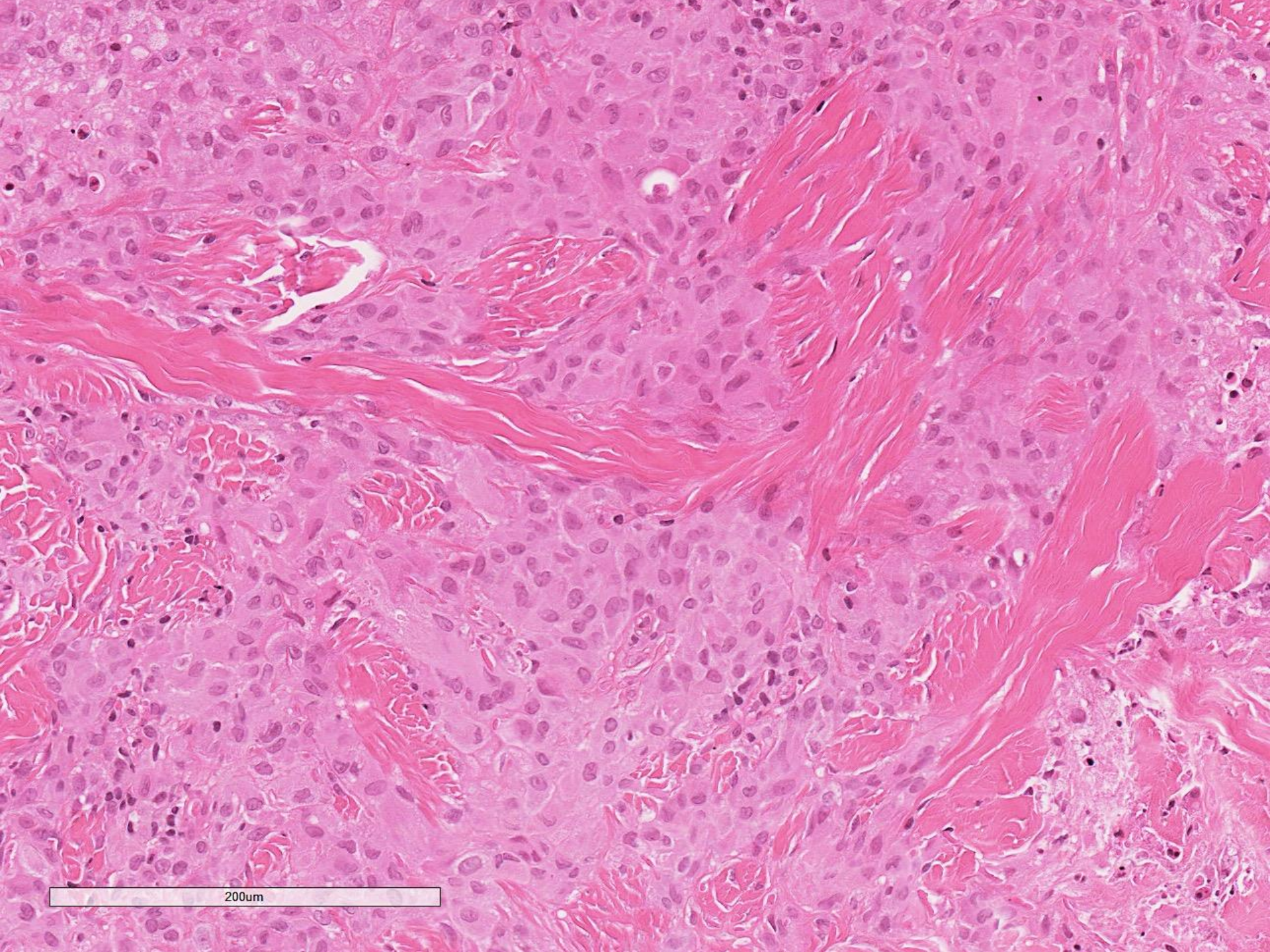
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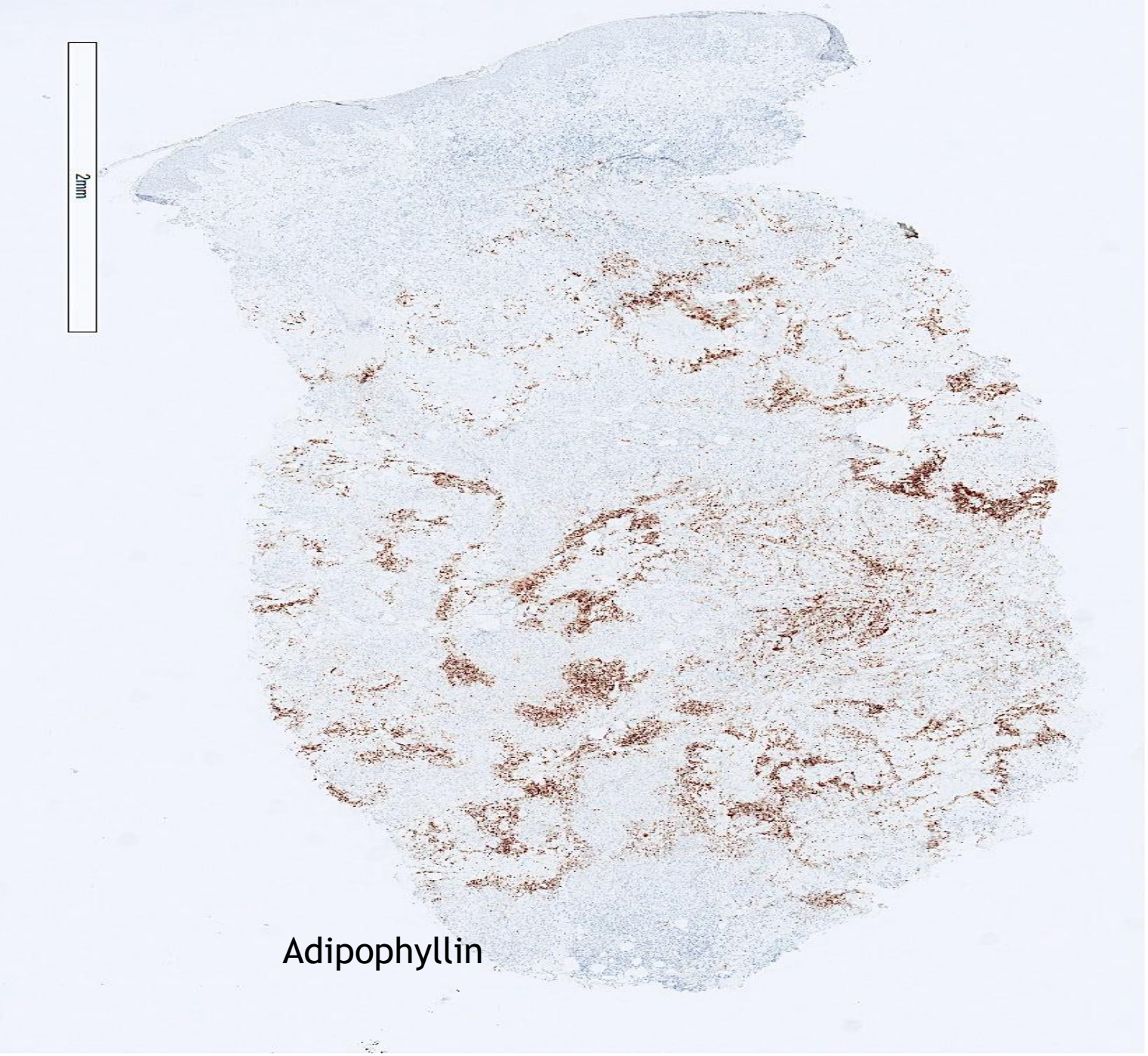


Adipophyllin

* 1: Juvenile xanthogranuloma
* 2: Granuloma annulare
* 3: Necrobiosis lipoidica
* 4: Histiocytic sarcoma
* 5: Necrobiotic xanthogranuloma

Juvenile Xanthogranuloma

* Most common in the first five years
* Multiple skin lesions much less common
* Mixed infiltrate including lymphocytes, eosinophils and plasma cells
* Necrobiosis and cholesterol clefts XX

Granuloma annulare

* Generalised form- papular lesions, nodules can occur
* Necrobiosis is circumscribed/ill defined
* Lipid and giant cells are rare

Necrobiosis lipoidica

* B/L , single-multiple, often symmetrical lesions on the lower extremities.
* Characteristic lesion is a sclerodermatous plaque, round/oval shape and an elevated rim.
* Papulonecrotic and ulcero-nodular lesions are rare.

* Cholesterol clefts are rare and only exceptionally prominent.

Histiocytic sarcoma

* Extremely rare
* Advanced stage, red-violaceous nodules
* Pleomorphic histiocytic cells with multiple and frequent mitoses.

*Necrobiotic Xanthogranuloma

* Rare, non-Langerhans cell histiocytosis, first described by Kossard and Winkelmann in 1980.

* 100 cases, worldwide

* The most common site of involvement is the face especially periorbital areas. In some patients this feature is absent and lesions can be found restricted to extremities & trunk.

* NXG is now considered as a systemic disease. Internal organ involvement have been reported, in most cases asymptomatic and diagnosis established only at post-mortem.

* Haematological and lymphoproliferative malignancies are the most important associated systemic disorders.

* Develop approximately 2.4 years after onset of skin lesions.

* 80-90% of cases demonstrate monoclonal gammopathy (IgG kappa 60% > IgG lambda 26%)

* 10% cases develop multiple myeloma

* Clinical course, chronic, progressive and indolent.

* Patients should undergo long term surveilance for haematologic and other malignancies.

Investigations

* **Serum Free light chains:** Free Kappa 36.8 (3.3 -19.4)Free Lamda 8.7 (5.7 – 26.3)Kappa:Lamda 4.23 (0.26 – 1.65)

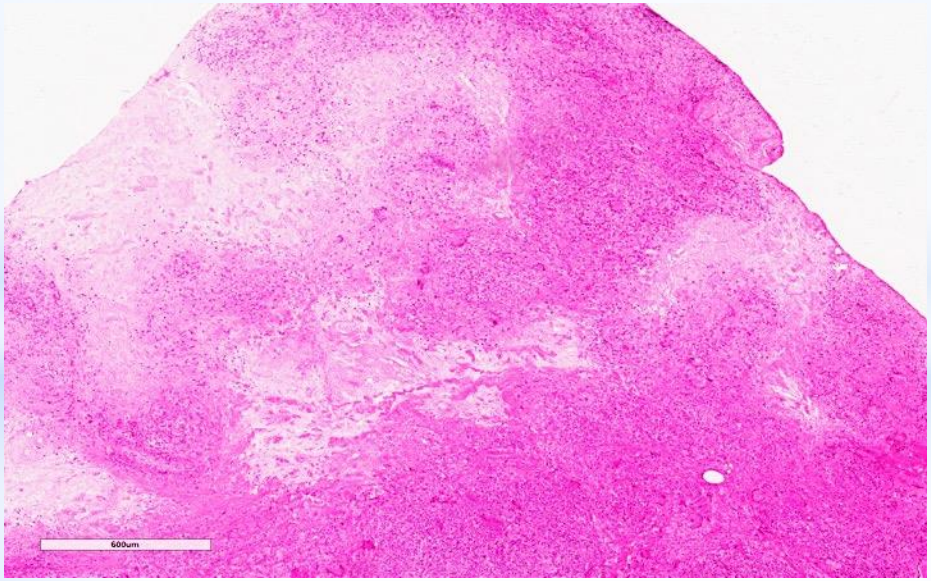
* **Bence Jones proteins:** Monoclonal Kappa detected

Management:

- Commenced on course of topical PUVA for symptom control
- Analgesia
- Discussed with Haematology:
  - BM biopsy performed; plasma cell myeloma with approx. 30-50 cellularity.
  - Reclassified as ‘smouldering myeloma’
  - Discussed potential treatment with alkylating agent for both haematological and dermatological manifestations.

* A few months later patient developed orbital/peri-orbital plaques

* Histology showed features typical of NXG



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