

Panniculitis update

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Update in Dermatopathology
Liverpool, Nov 28 2017



Icahn
School of
Medicine at
**Mount
Sinai**

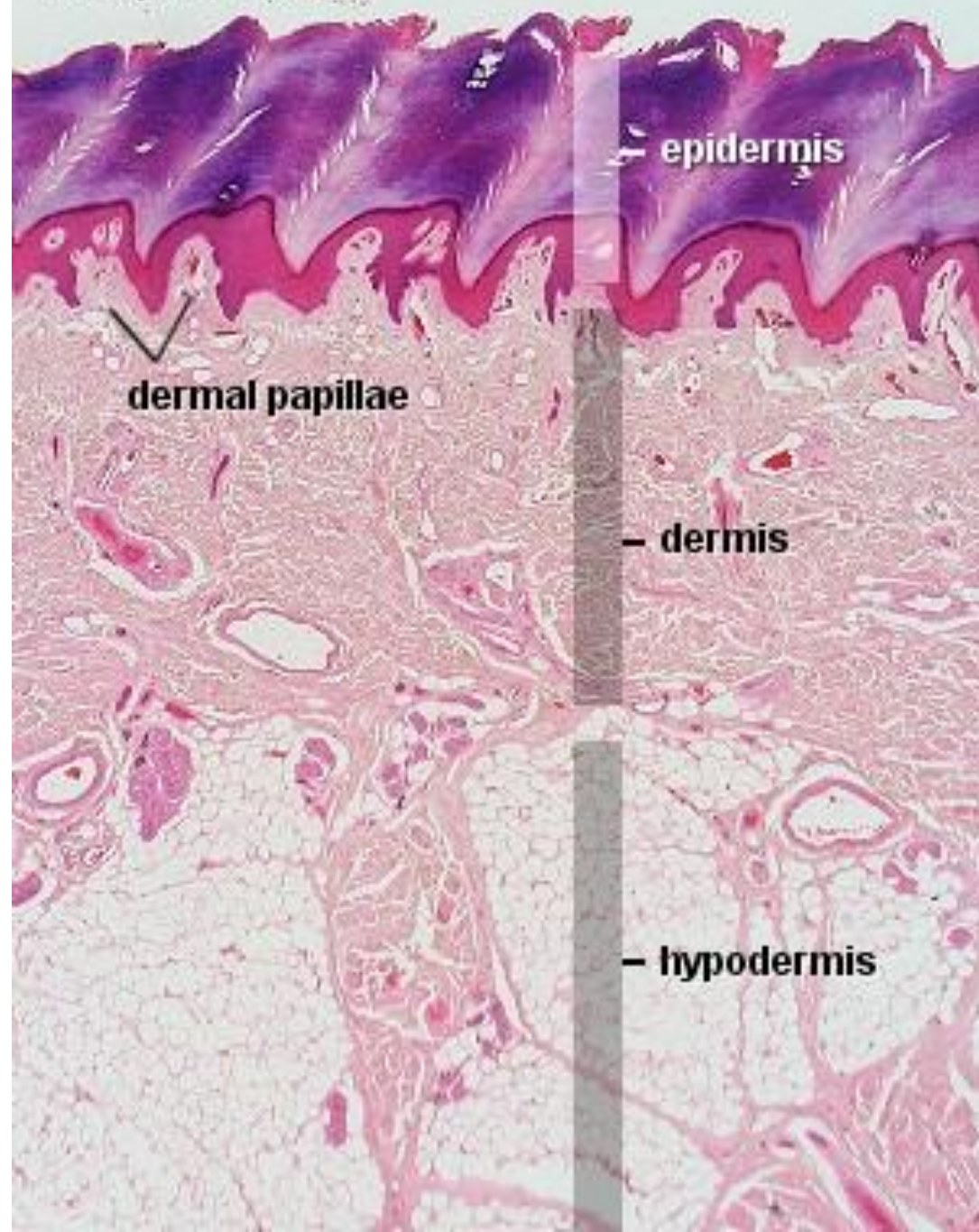
Clinical presentation of panniculitis

- Large often deep seated nodules
- More apparent by palpation than inspection
- May be tender or ulcerate
- Anatomic site can be relatively specific (tibia, calf, proximal limbs)

Anatomy of adipose tissue

- Divided into lobules
- Lobules separated by septa
- Septa contains nutrient vessels

Skin, thick H&E

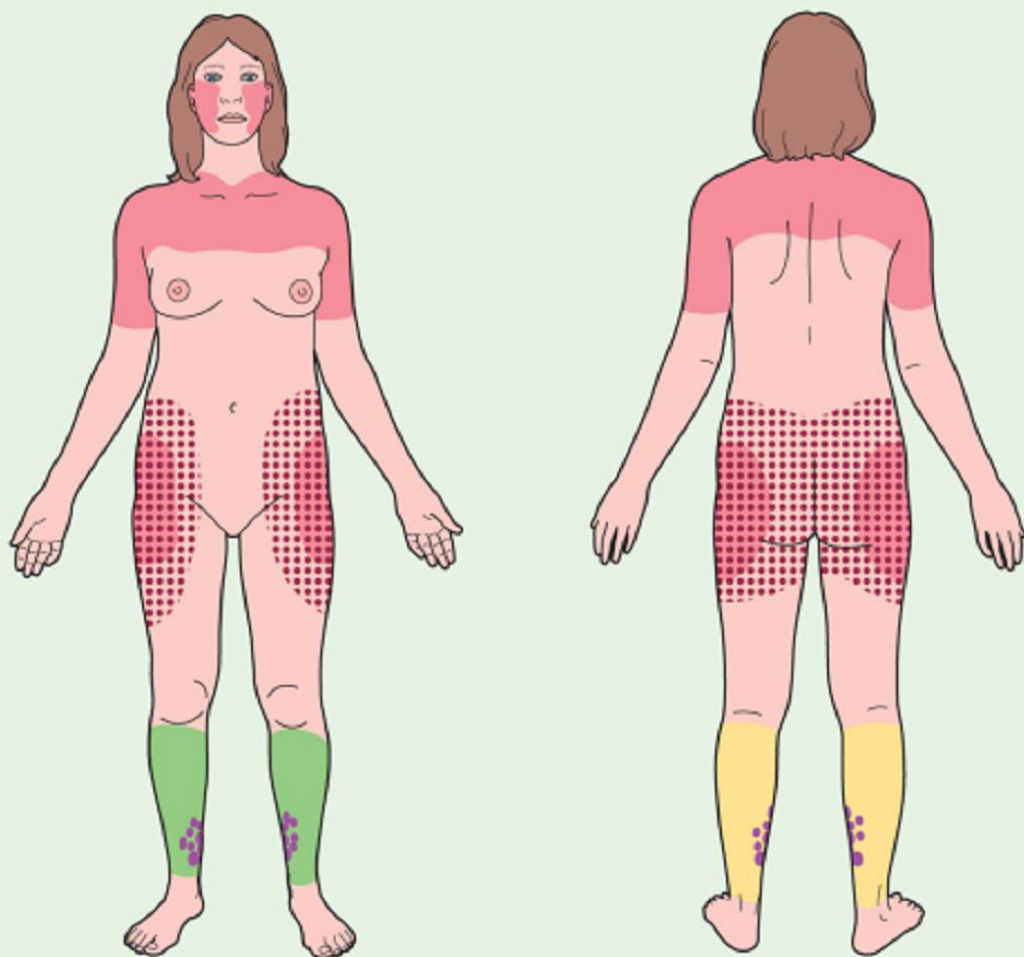


Classification of panniculitis

Clinical Algorithm

- Determine panniculus is primary site
- Look for predominant site of involvement

MOST COMMON LOCATIONS FOR SEVERAL FORMS OF PANNICULITIS



- | | | |
|--|---|--|
|  Erythema nodosum |  Erythema induratum |  Lipodermatosclerosis |
|  Lupus panniculitis |  α_1 -antitrypsin deficiency panniculitis | |

Classification of panniculitis

- Determine panniculus is primary site

Pathology Algorithm

- I. Septal predominant –erythema nodosum
- II. Lobular predominant – lupus panniculitis
- III. With a vasculitis – erythema induratum

Classification by pattern

- Panniculitides evolve with time
- Much overlap exists
- A high proportion cannot be classified
- Distinctions are often artificial

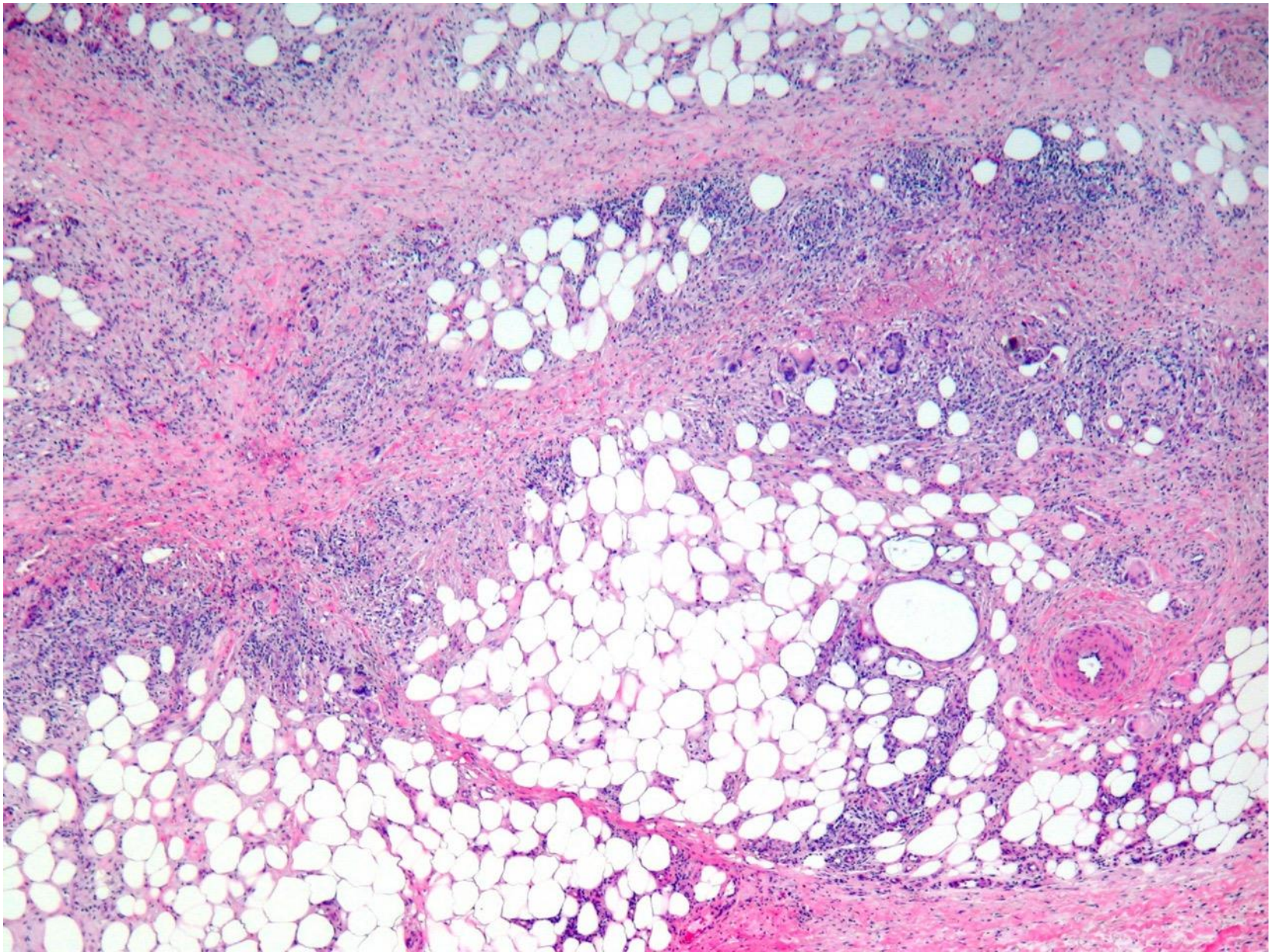
Case 1

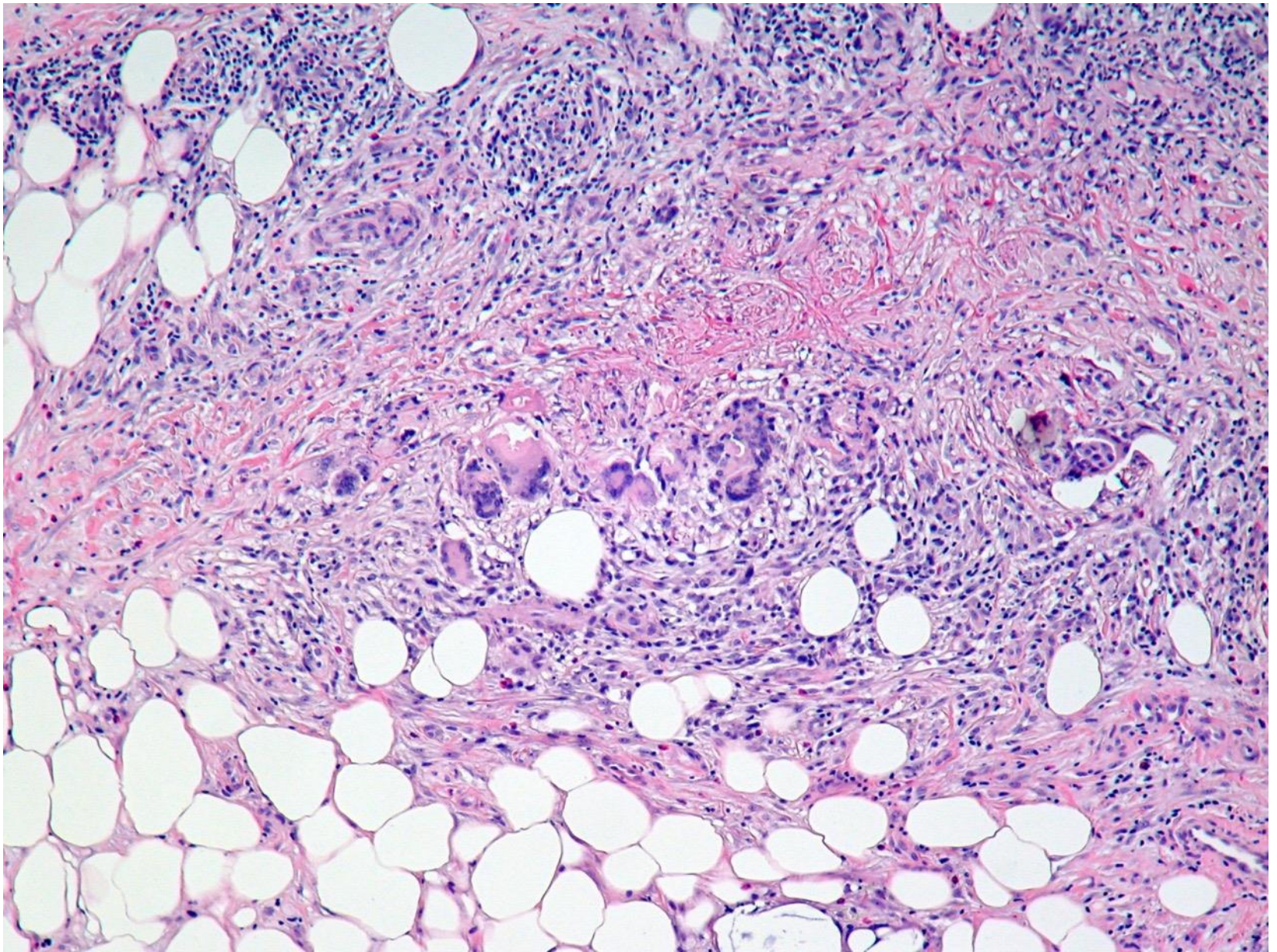
Chief Complaint

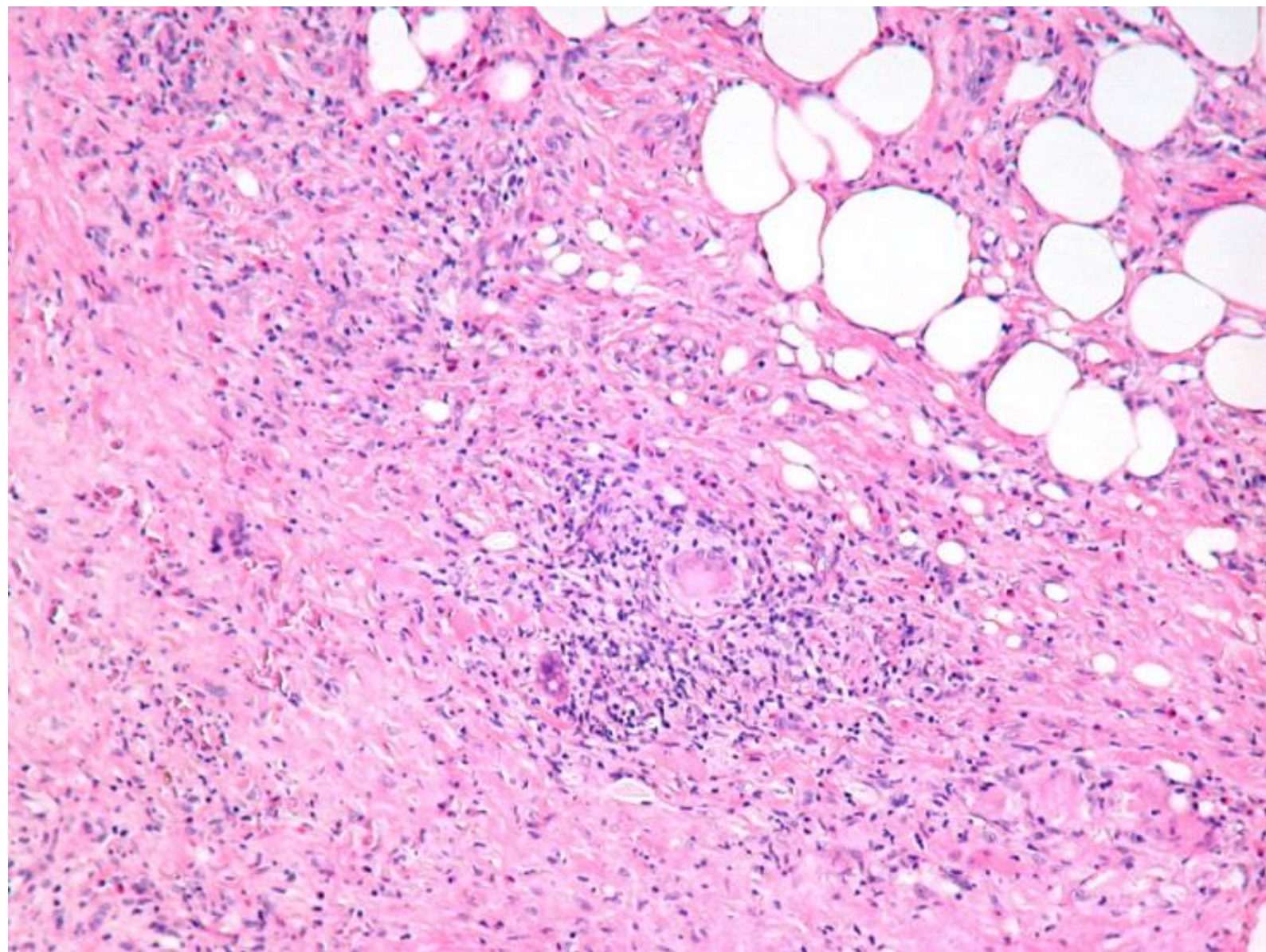
48 y/o woman c/o discoloration of lower legs for a couple of years that does not go away.

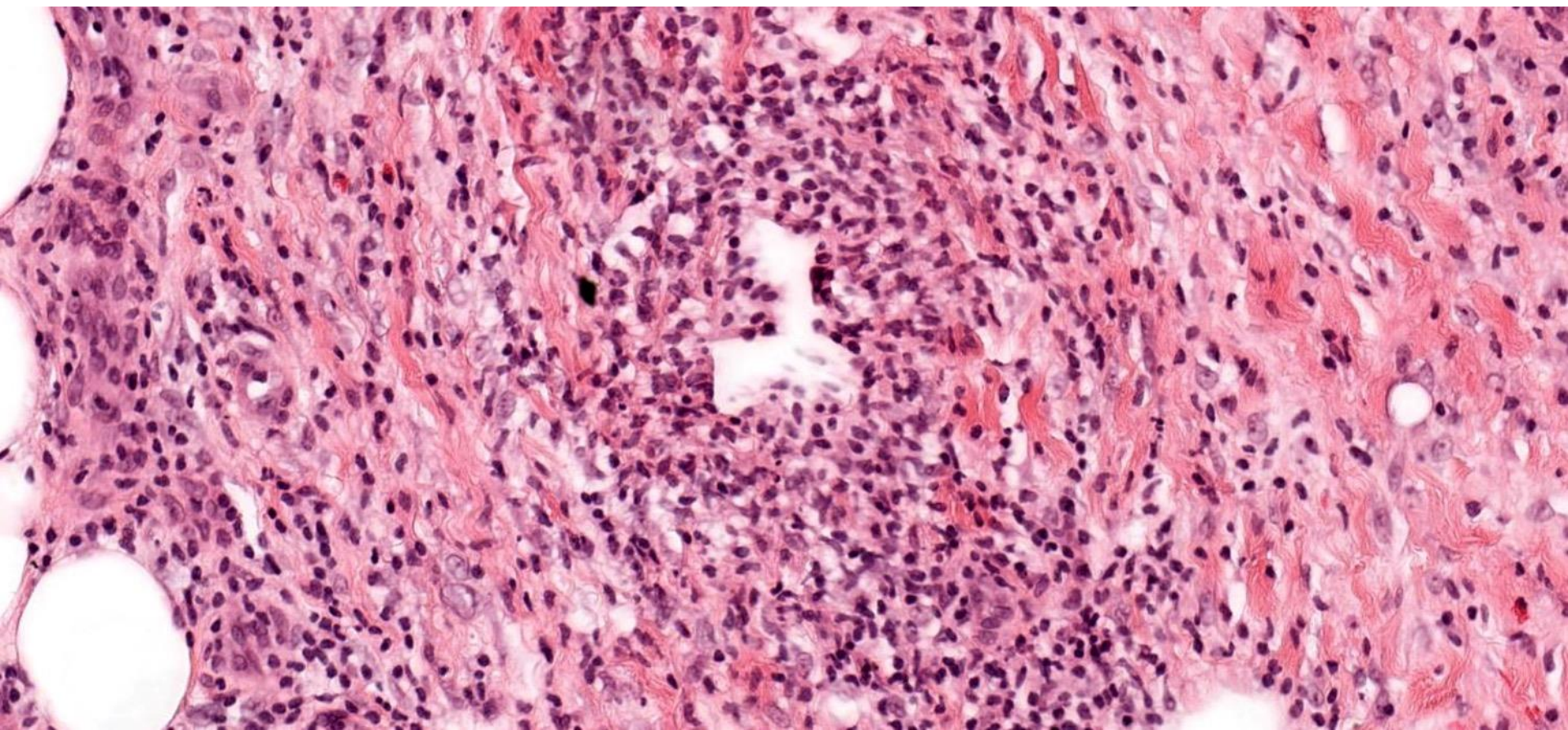


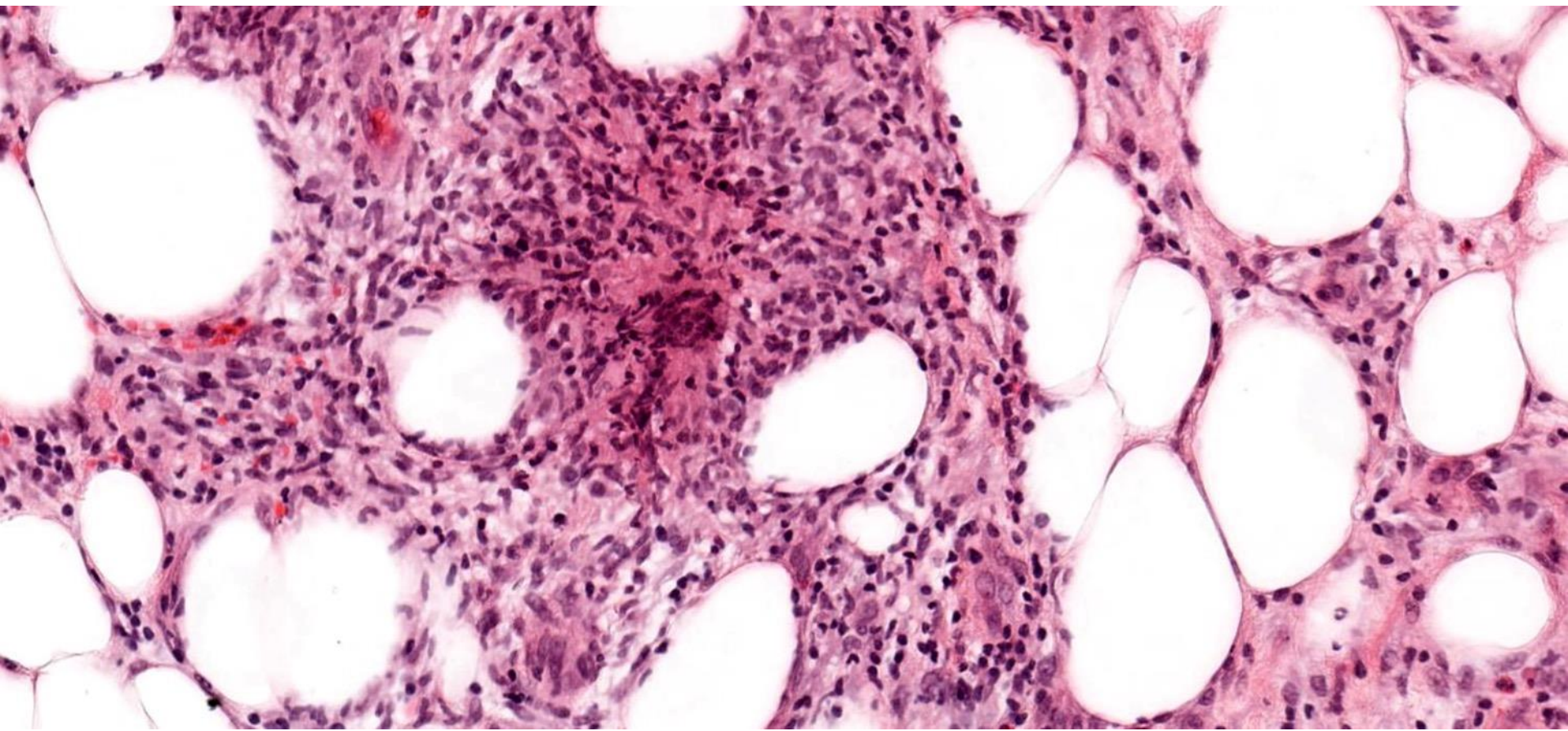


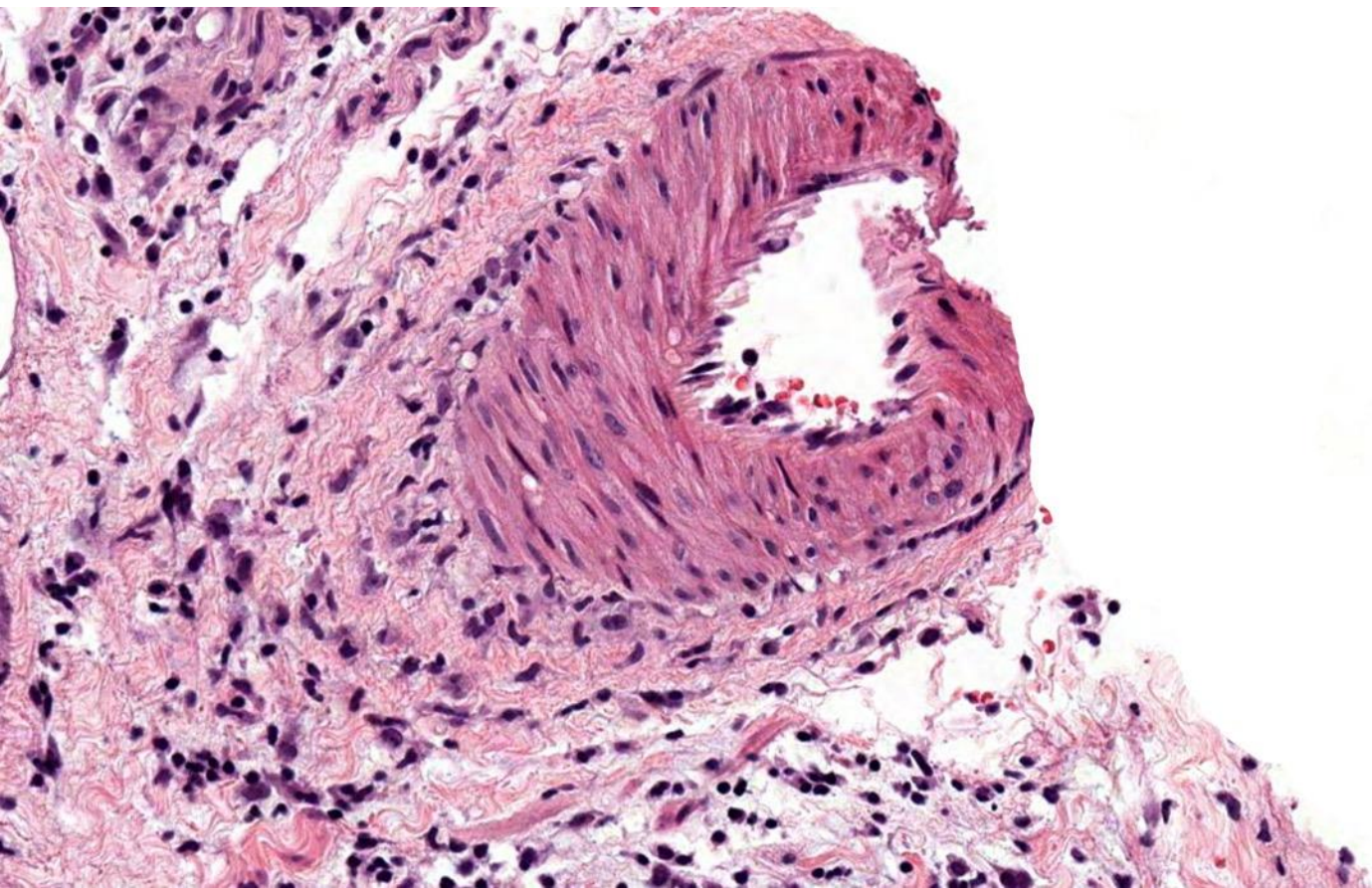












Erythema nodosum

Clinical presentation

- Usually occurs bilaterally over pretibial surfaces, other sites, acutely (specific)
- Slow involution
- Unilateral variants exist
- Tender, bruise like appearance
- No ulceration
- May have fever, arthralgia, malaise

Associations

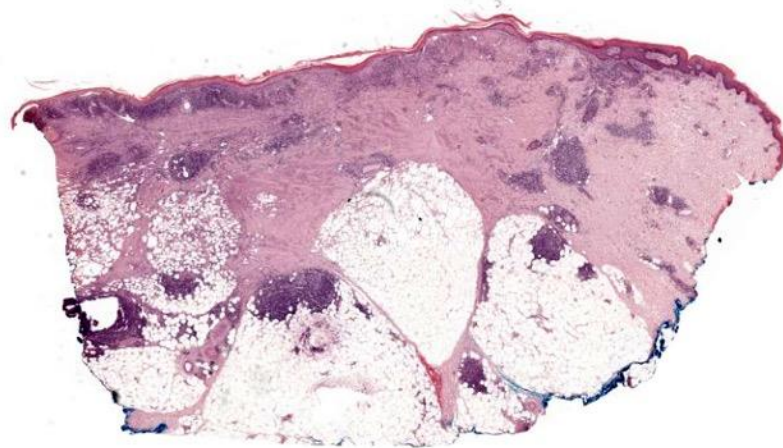
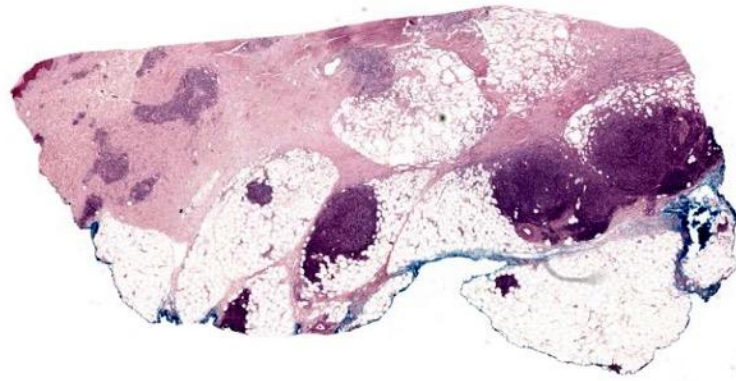
- Reaction to many things
- Infections (strep)
- Drugs (OC)
- Sarcoidosis
- Inflammatory bowel disease
- 1/3 no antecedent can be found

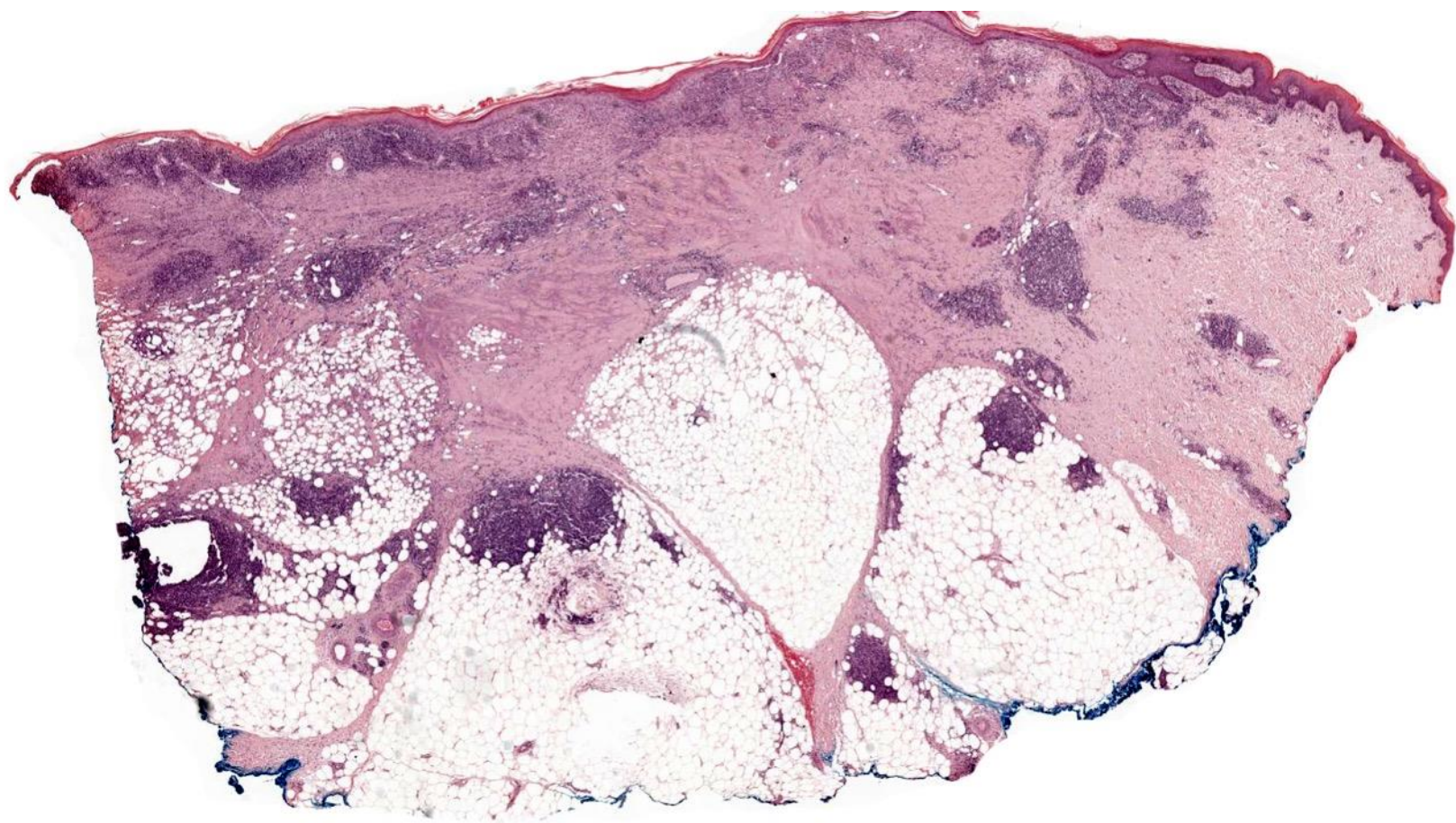
Pathology

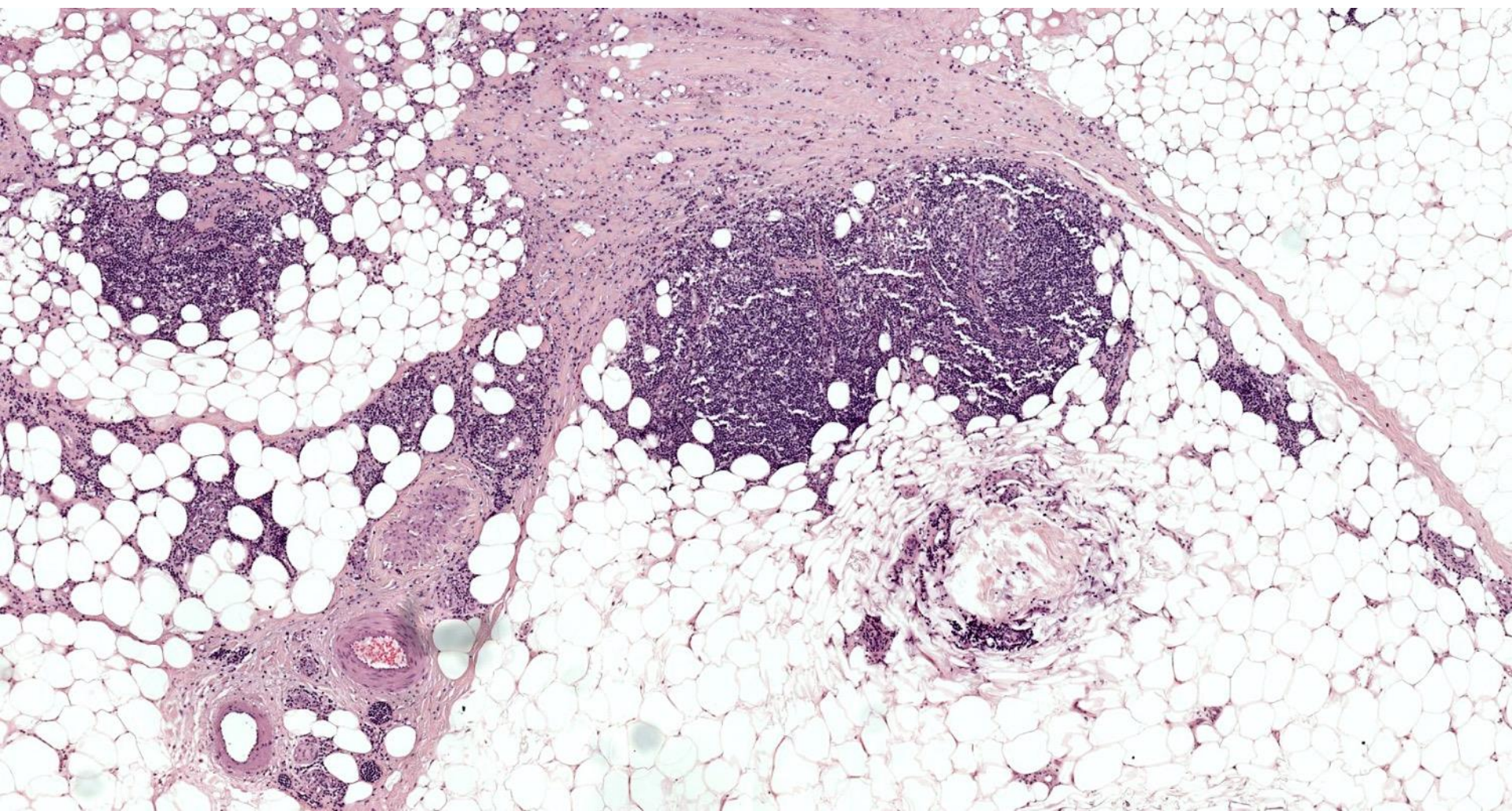
- Prototype of septal panniculitis (I)
- Widened septa evolve through stages
 - Fibrin neutrophils
 - Multinucleated giant cells
 - Resolution
- Granulomas at the periphery: Miescher's granulomas-evolve through stages
- Venulitis in a small percentage (not considered a true vasculitis)

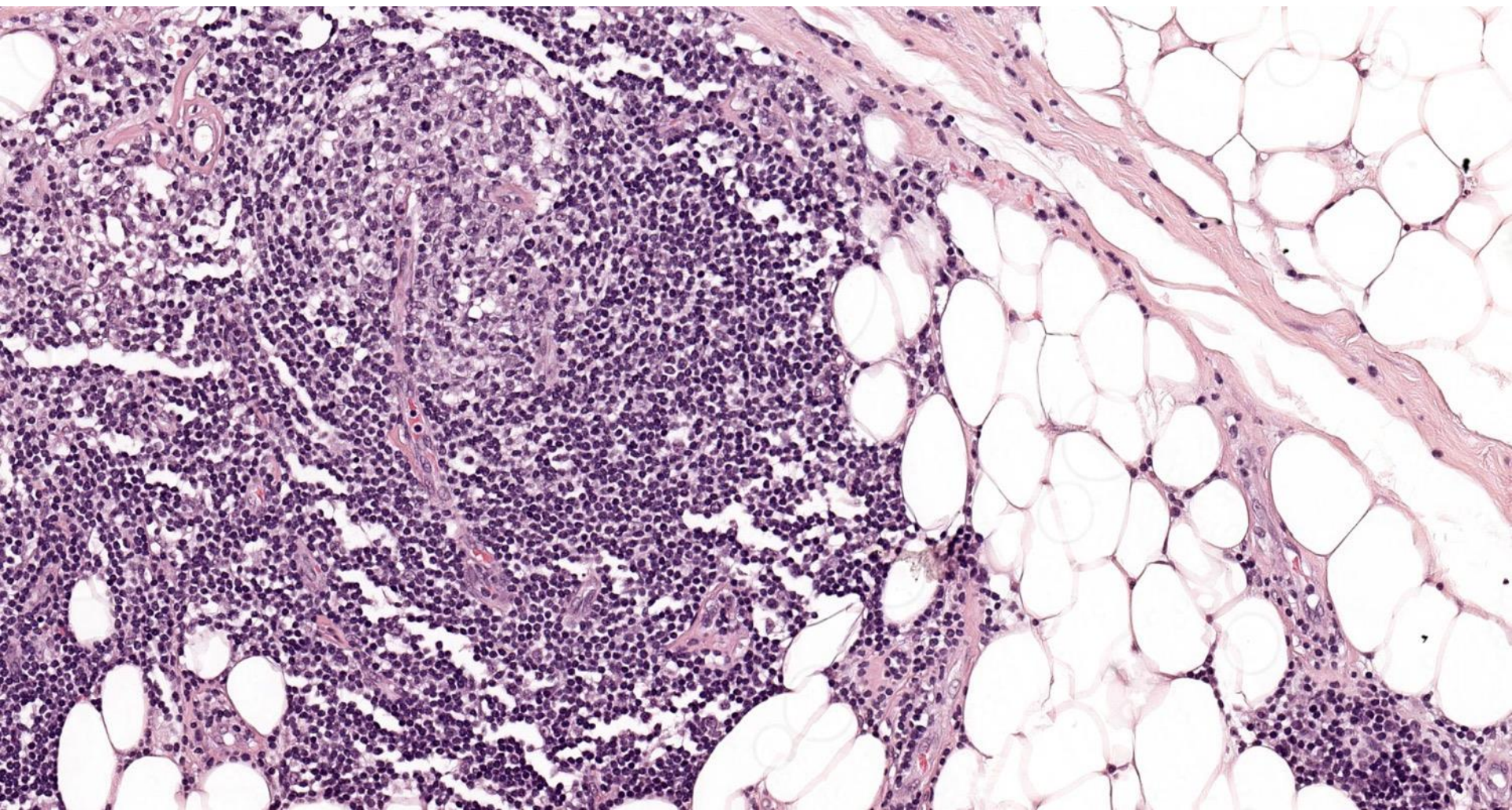
Case 2

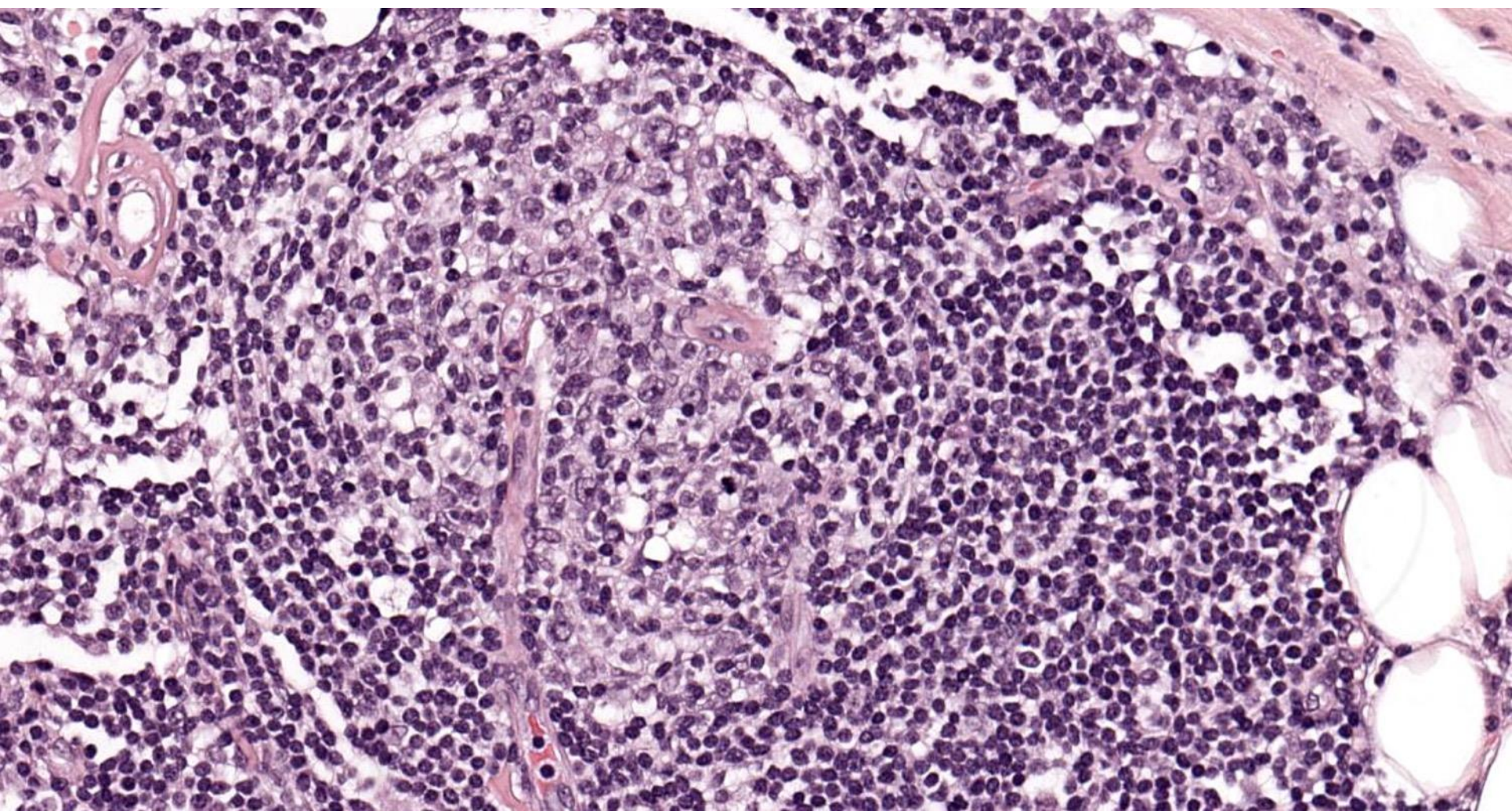


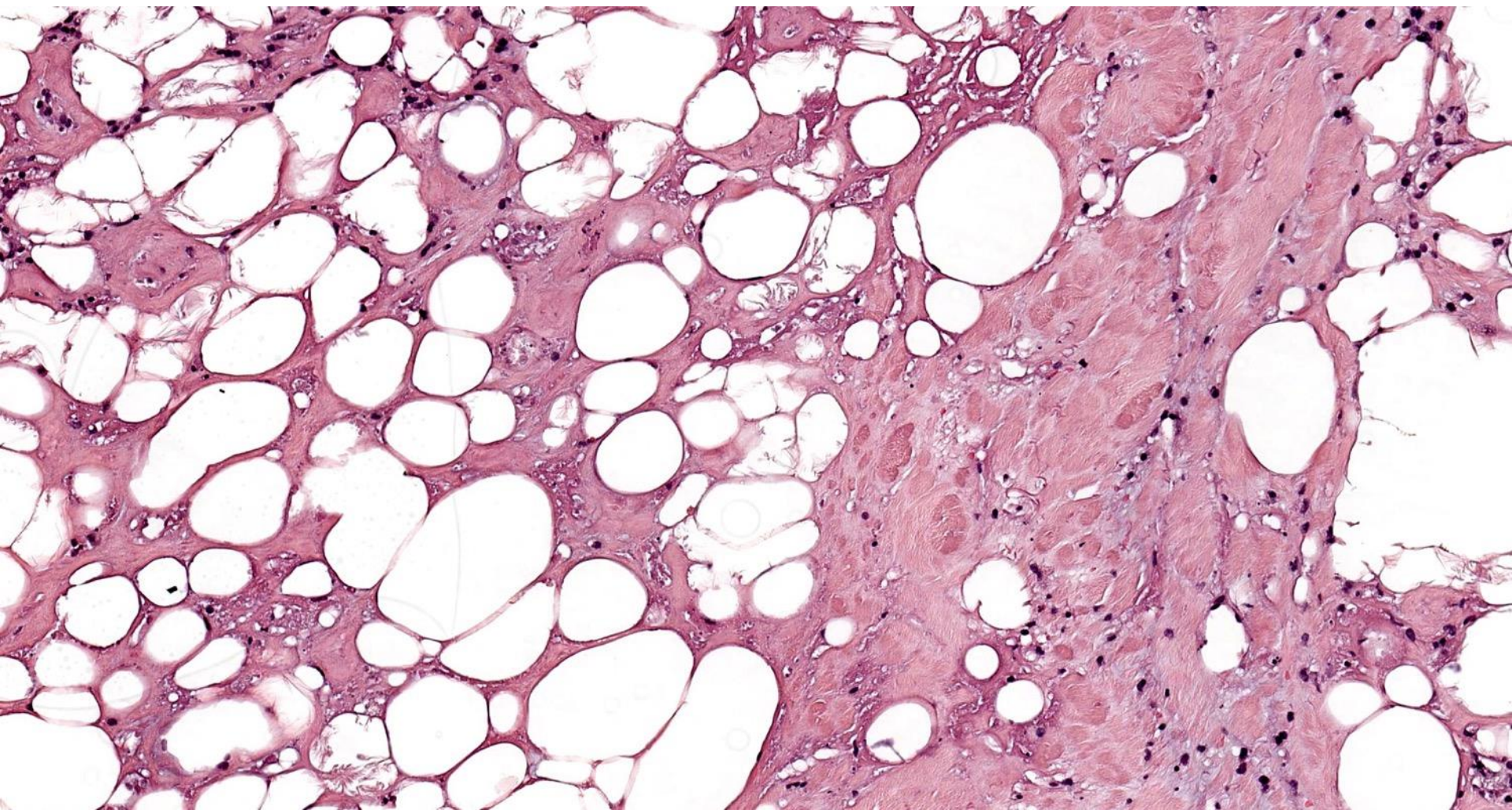


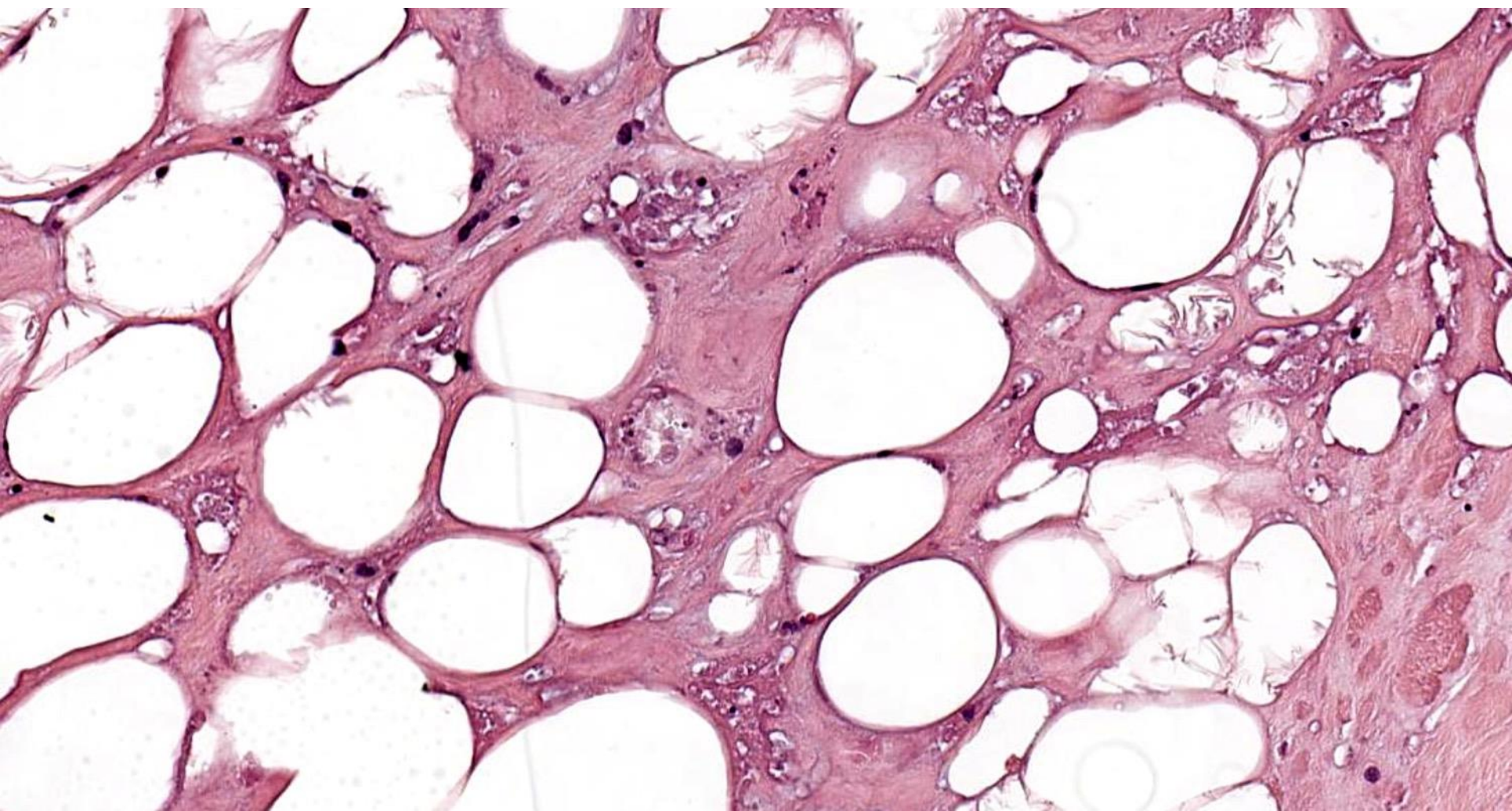


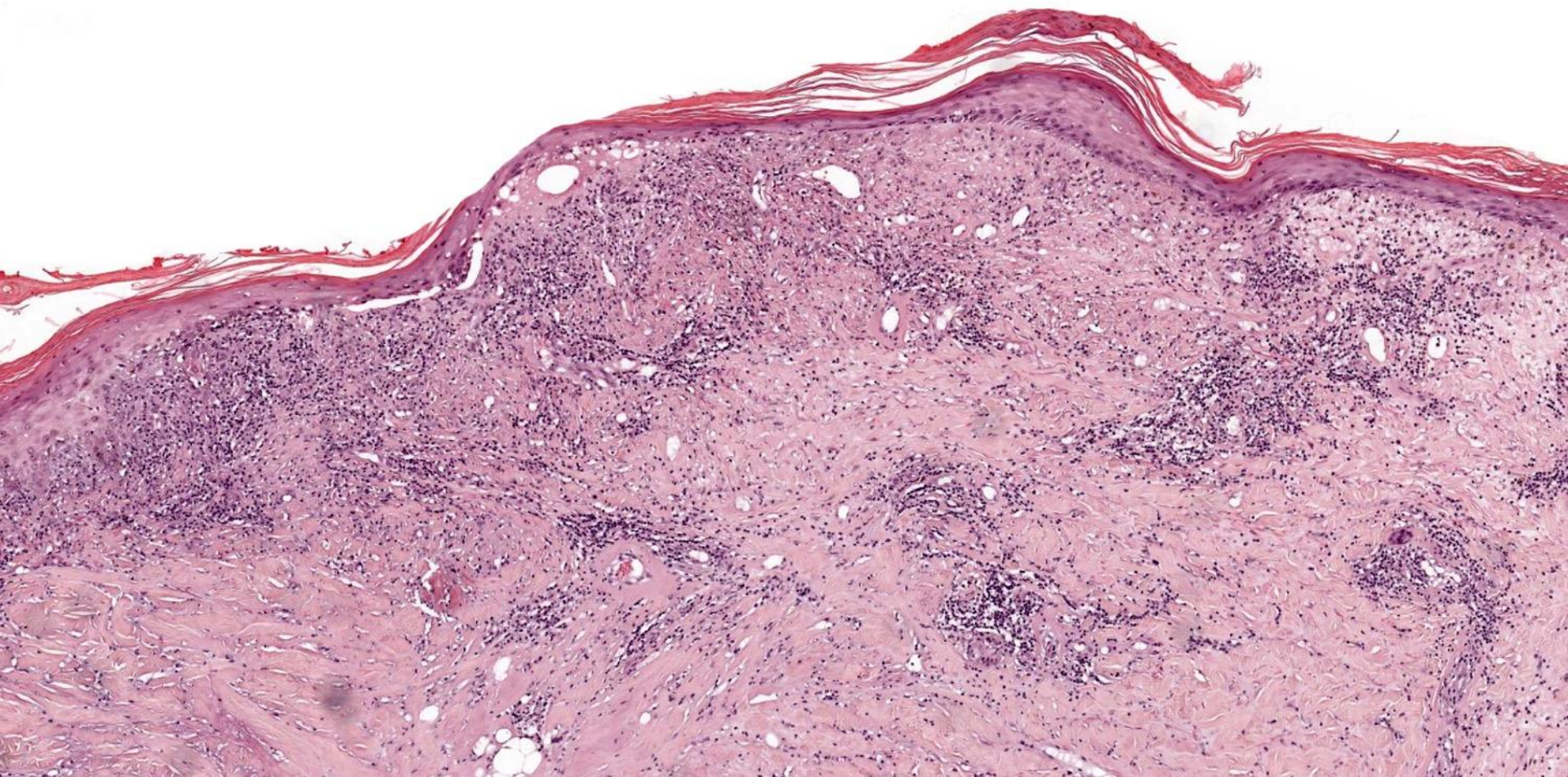


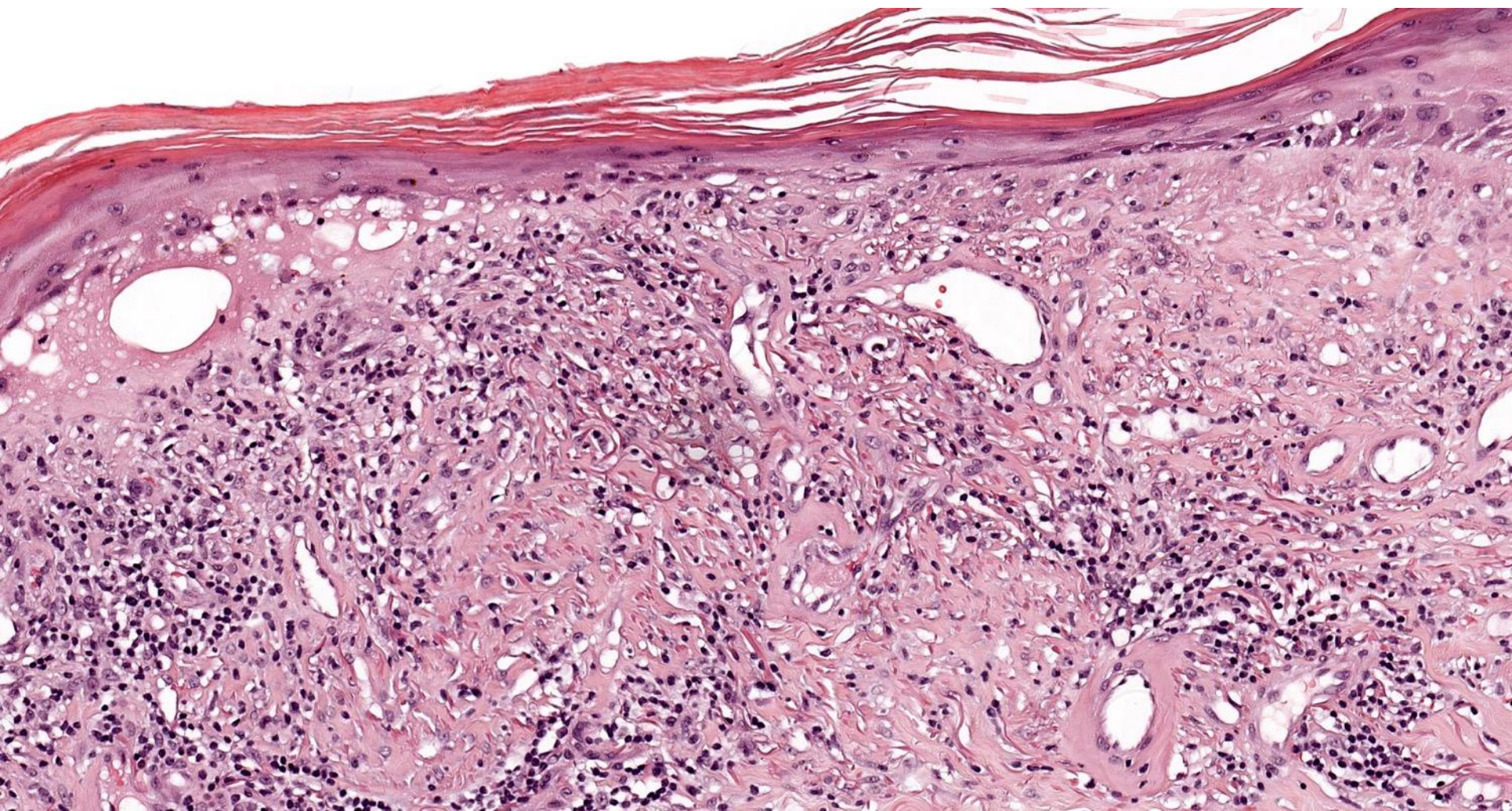












Lupus profundus/panniculitis

Clinical features

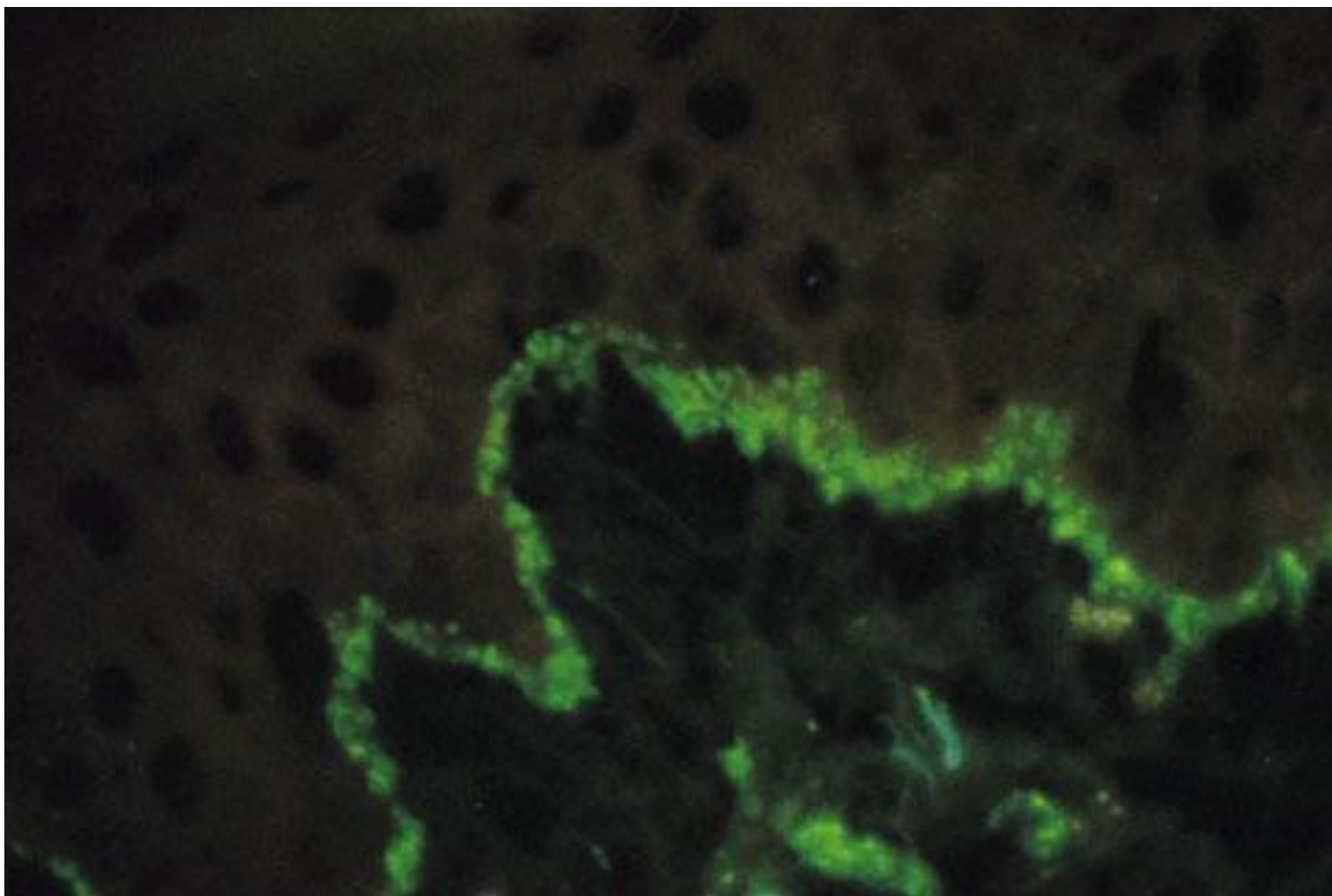
- Proximal and trunk, face
- Plaques and tumors
- Ulcerate and sometimes calcify, often disfiguring
- Heal with scarring
- Often associated with discoid lupus and not with systemic disease

Pathology

- Lobular panniculitis (II)
- Lymphoid aggregates with germinal centers (“connective tissue panniculitis”).
- Hyalinized adipose tissue
- Mucinosi, calcium deposition
- Sometimes surface changes of lupus in epidermis and dermis (DLE)

Immunopathology

- CD123 often positive in lupus
- May have positive lupus band



Case 3

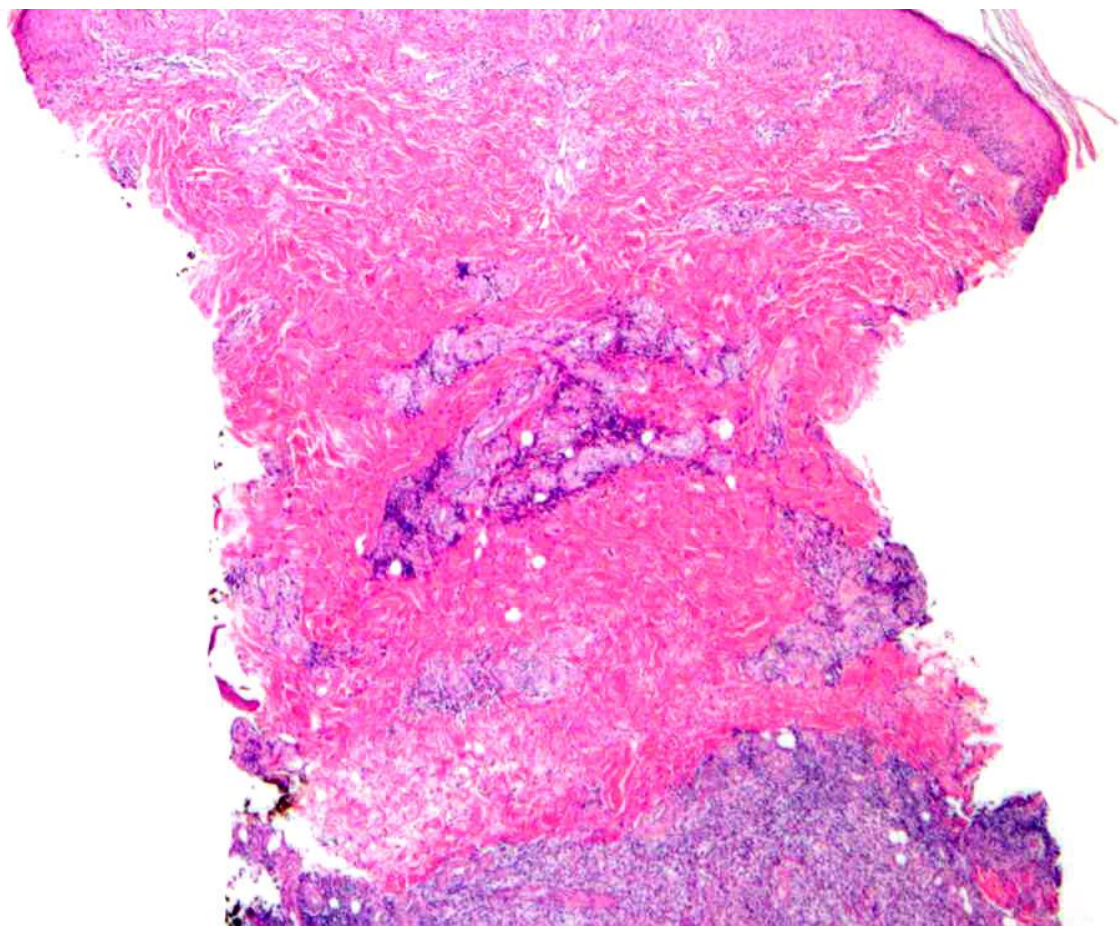
45 y/o male with CC of tender rash on lower legs present for 4 years

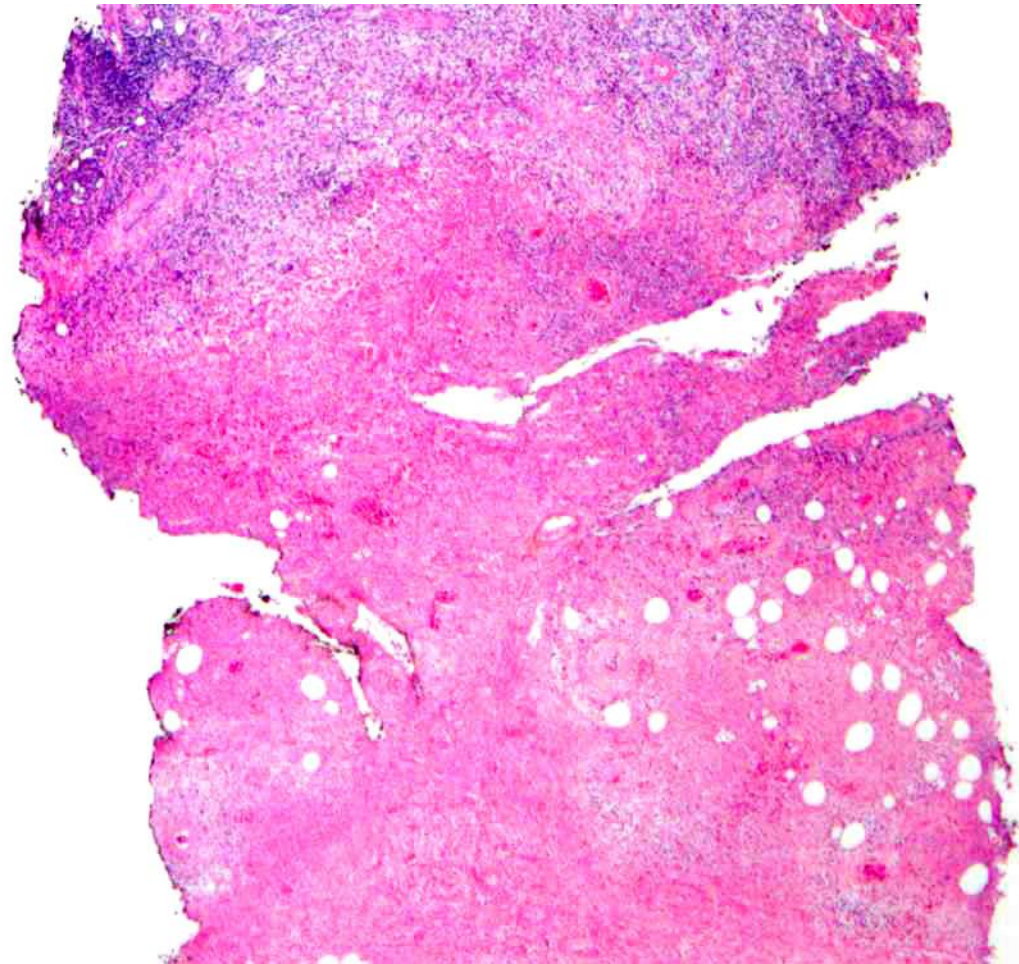


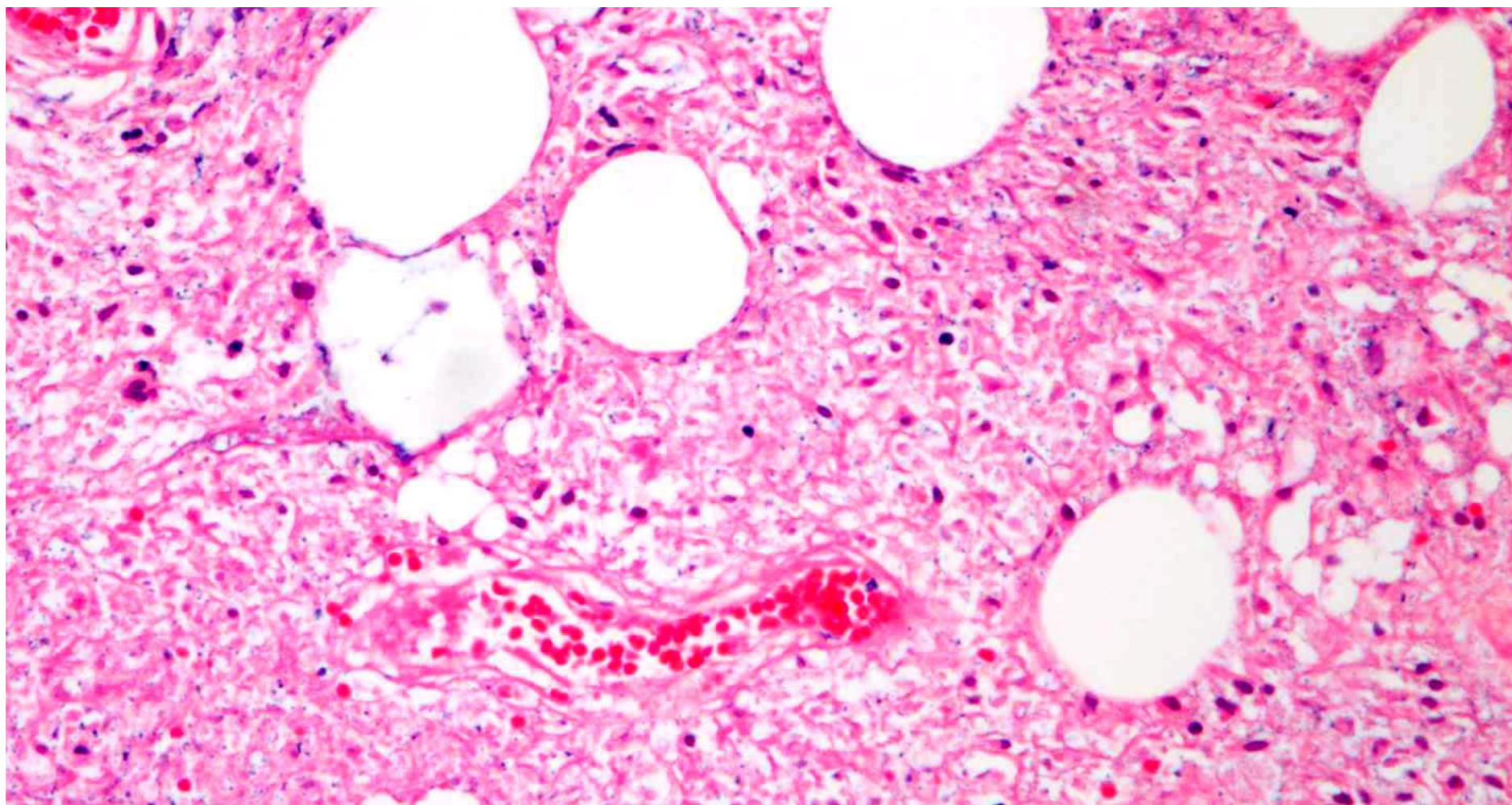


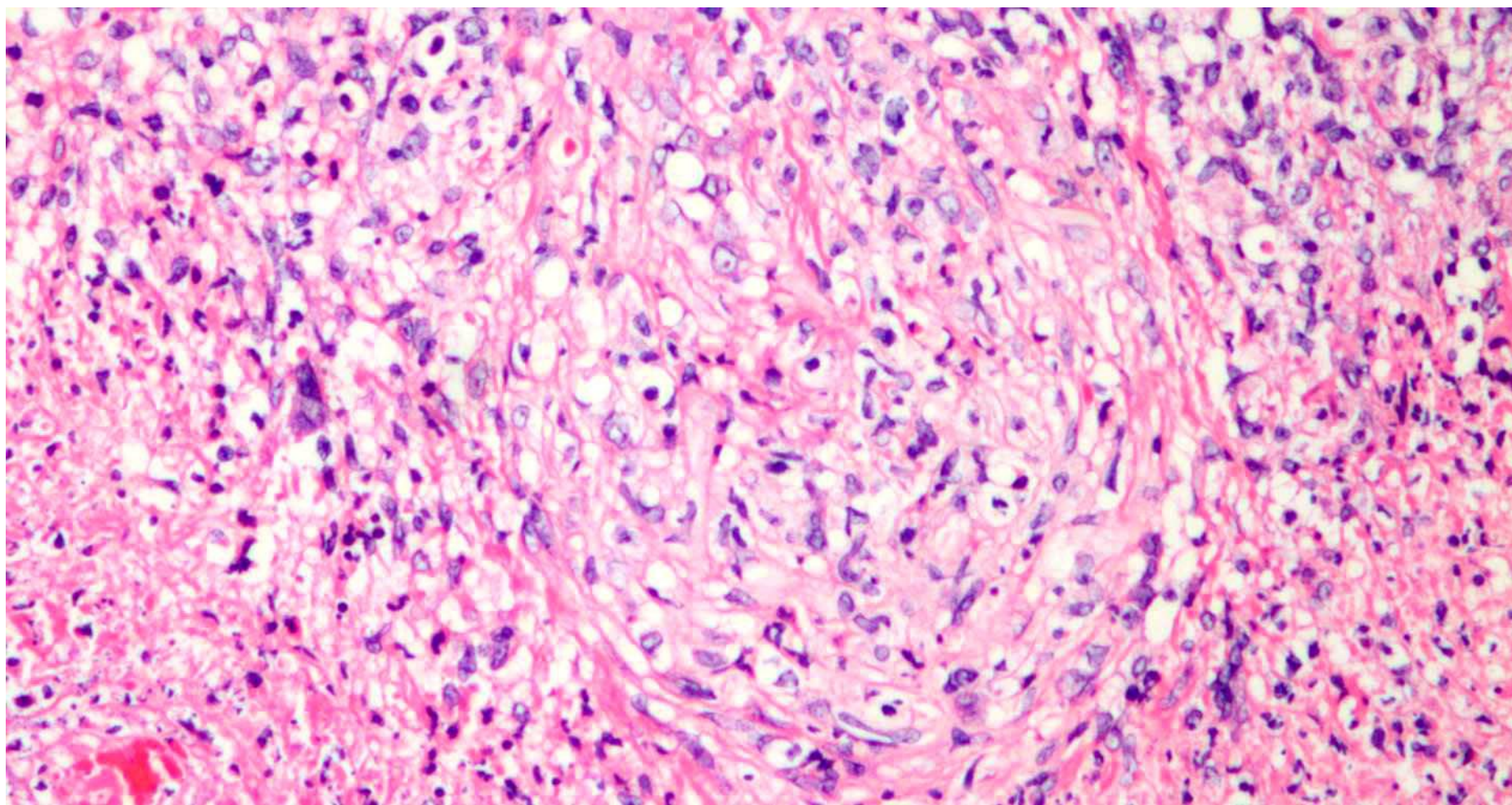


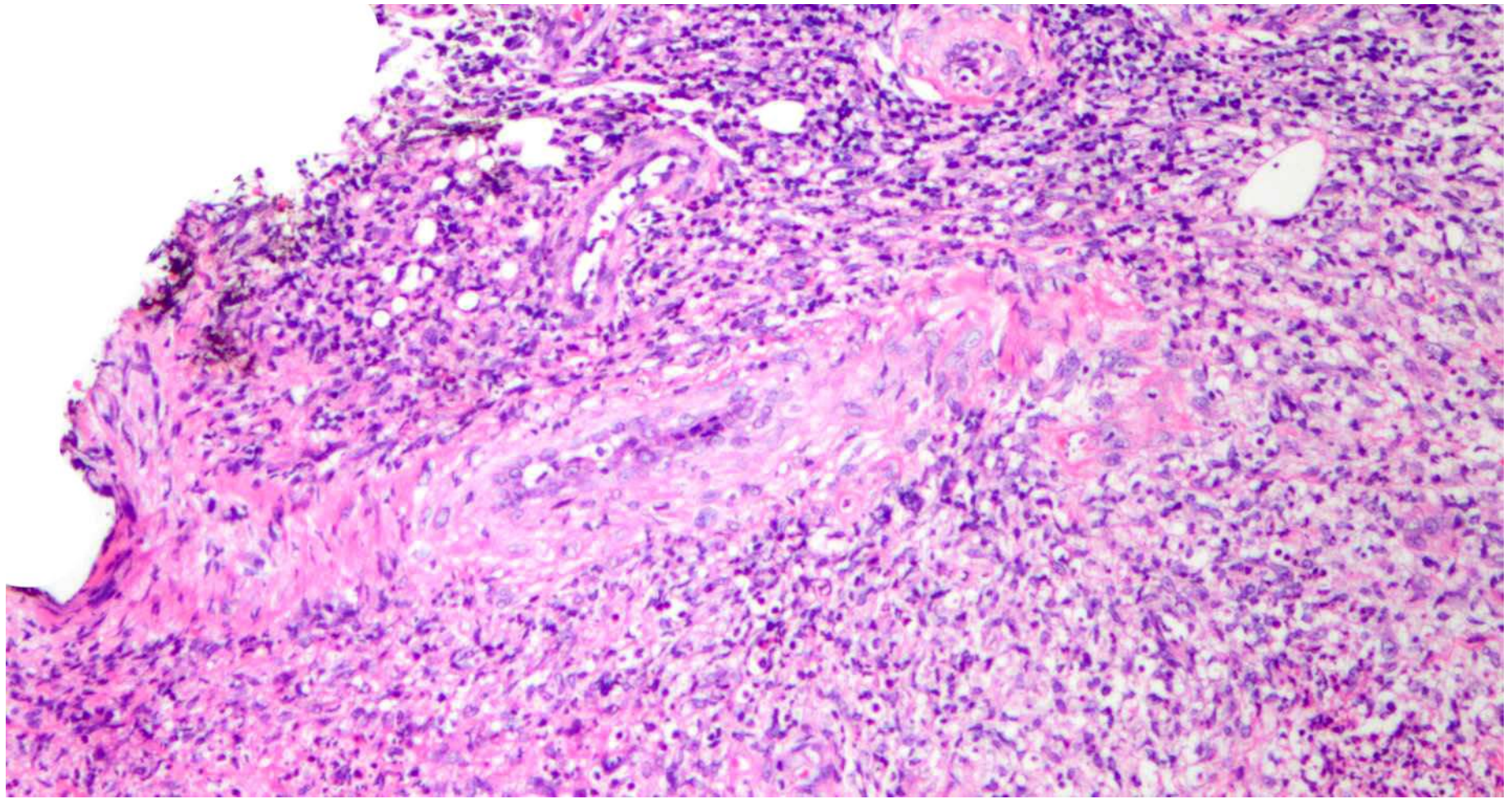












Erythema induratum/nodular
vasculitis

Clinical

- Erythematous tender nodules or plaques on posterior lower legs, calves
 - Often recurrent
- Young to middle-aged women
- +/- ulceration
 - Ulcerated nodules display overlying crust with rolled erythematous blue-tinged borders
- Individual lesions heal over several months with atrophic hyperpigmented scars

Pathology

- Histo: lobular panniculitis + vasculitis (II + III)
 - Neutrophils are sparse; mostly lymphocytic
 - Granulomas along with lymphs, plasma cells, eos and foci of extravascular caseous or fibrinoid necrosis
 - Vasculitis of medium sized vessels (veins or arteries)
- Often paucicellular; negative acid fast for bacilli, negative culture

EI Association with TB

- Originally classified as a tuberculid
- Mycobacterial DNA in lesions detected by PCR
 - Considered hypersensitivity to mycobacterial Ags
- Recommend testing in patient PPD, CXR, QG
- Non-TB causes: infections (eg nocardia) and drugs (eg propylthiouracil)

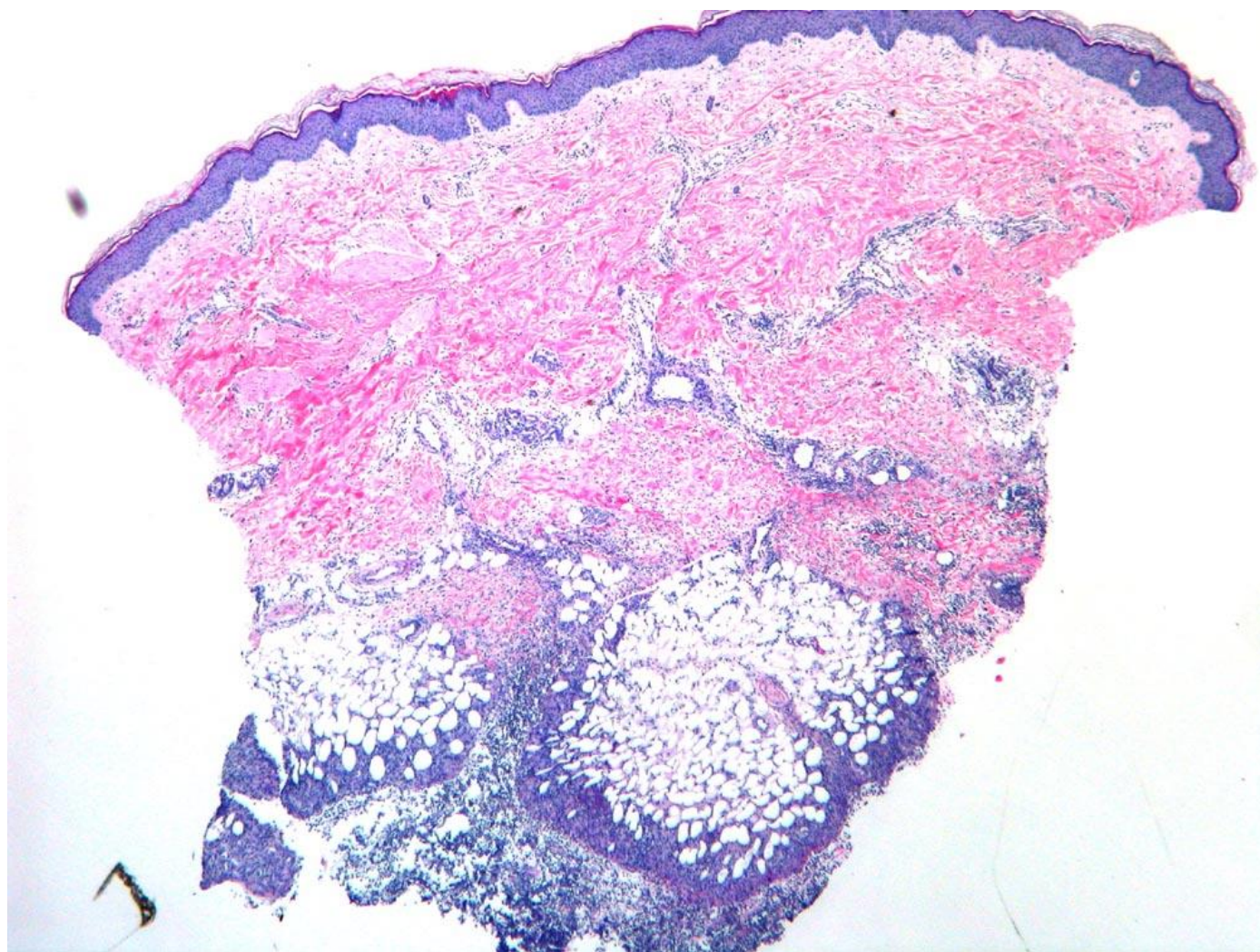
Case 4

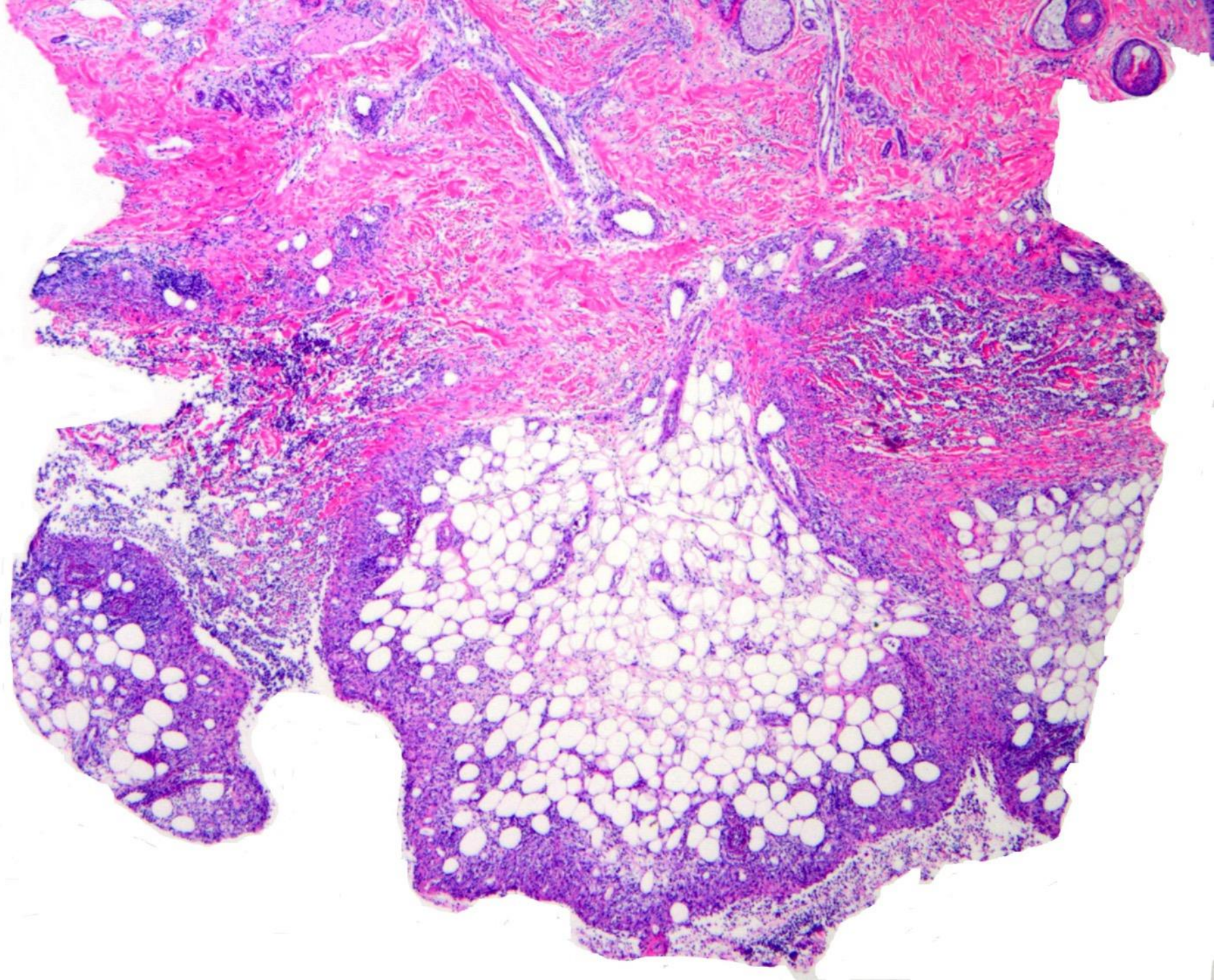
41 year old female with history
of multiple firm nodules over
many years

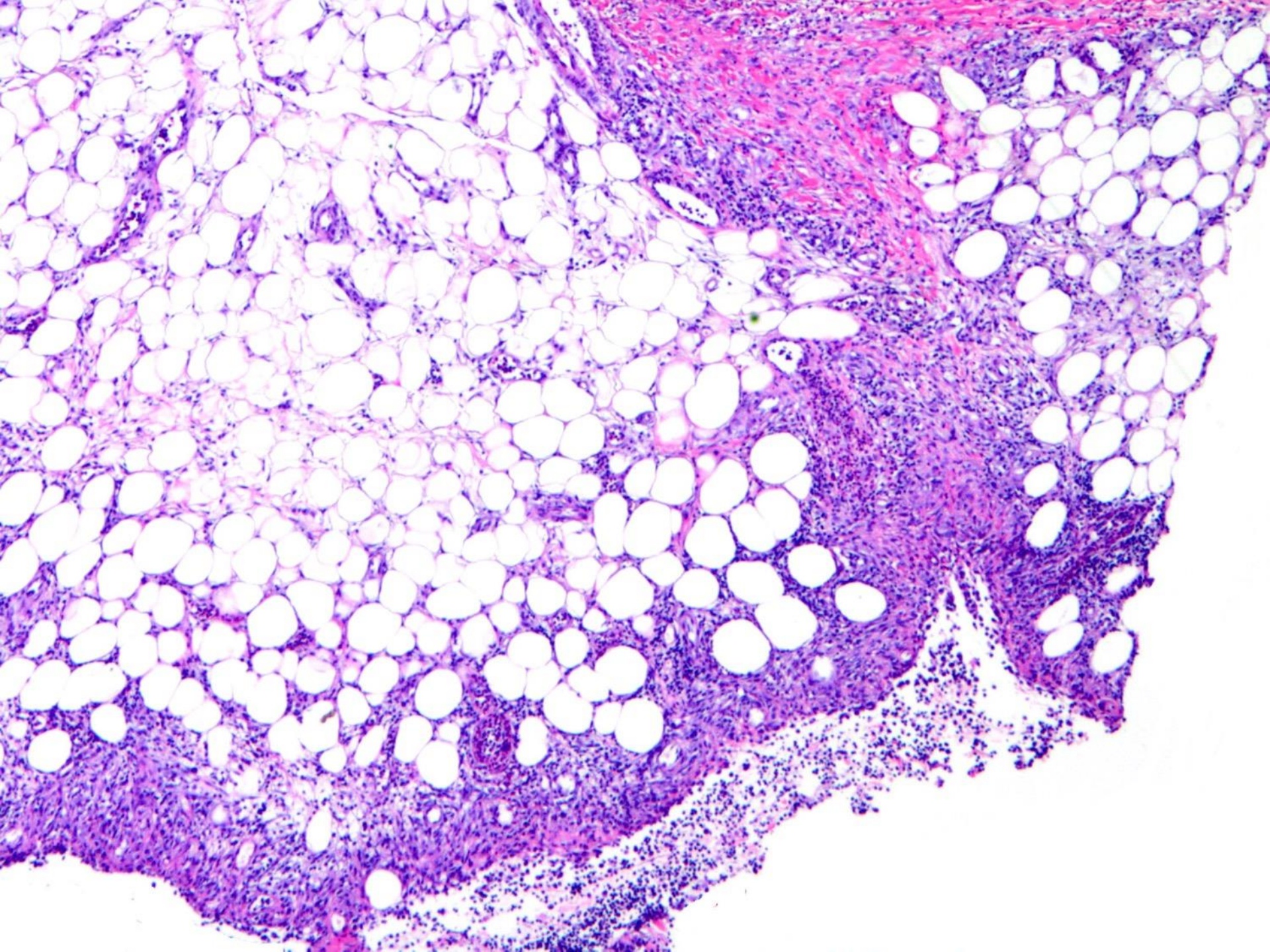


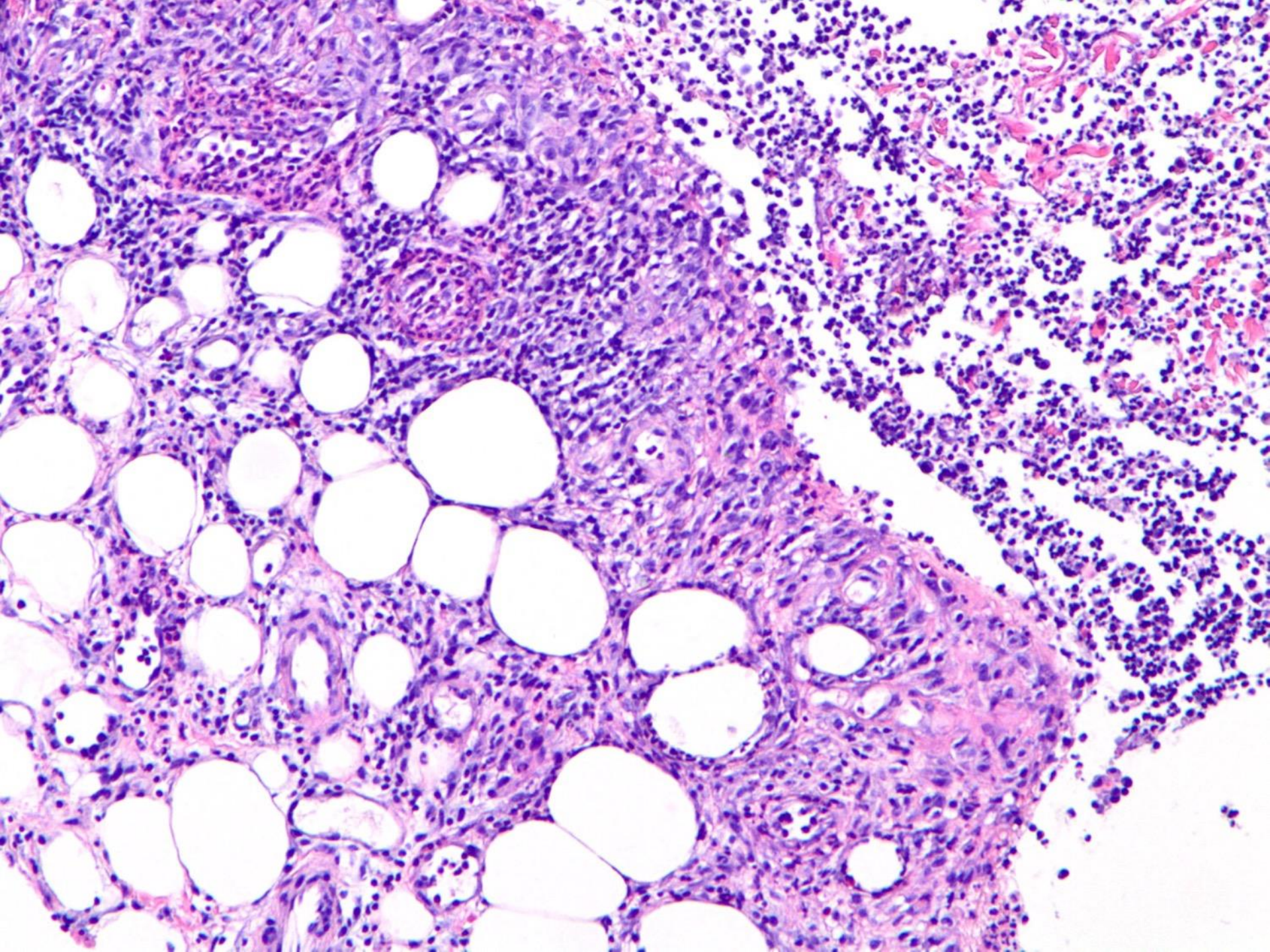


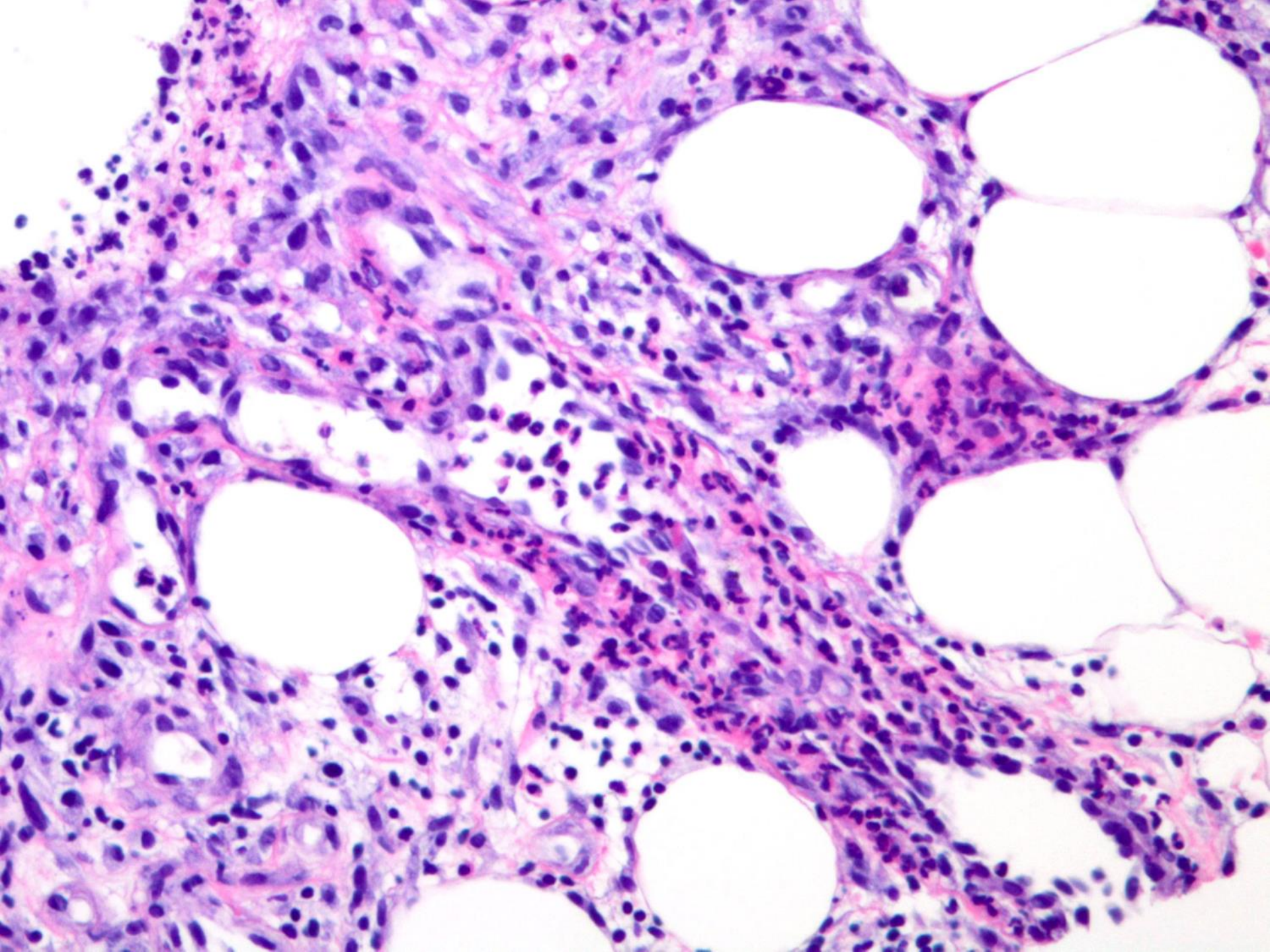


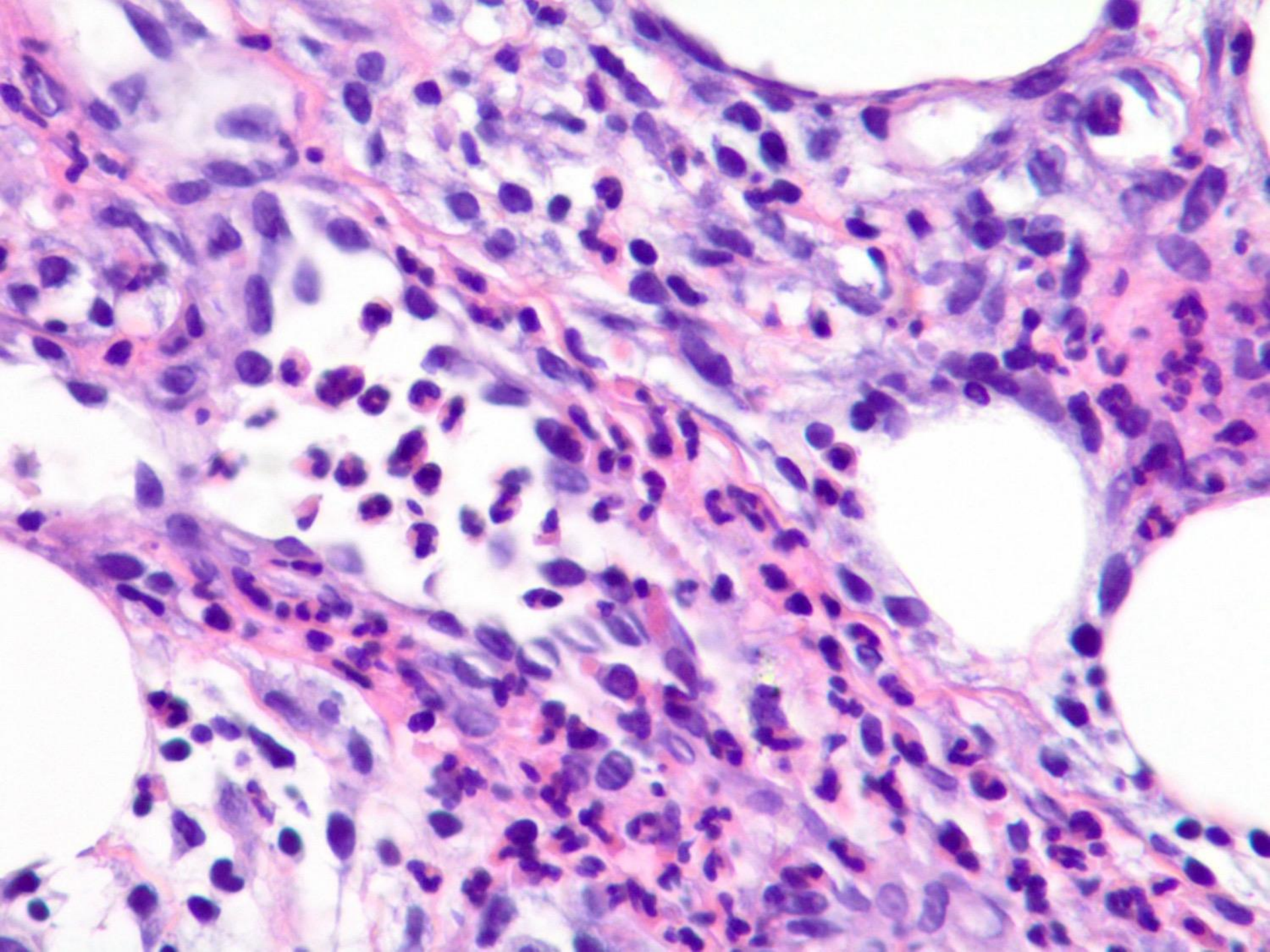


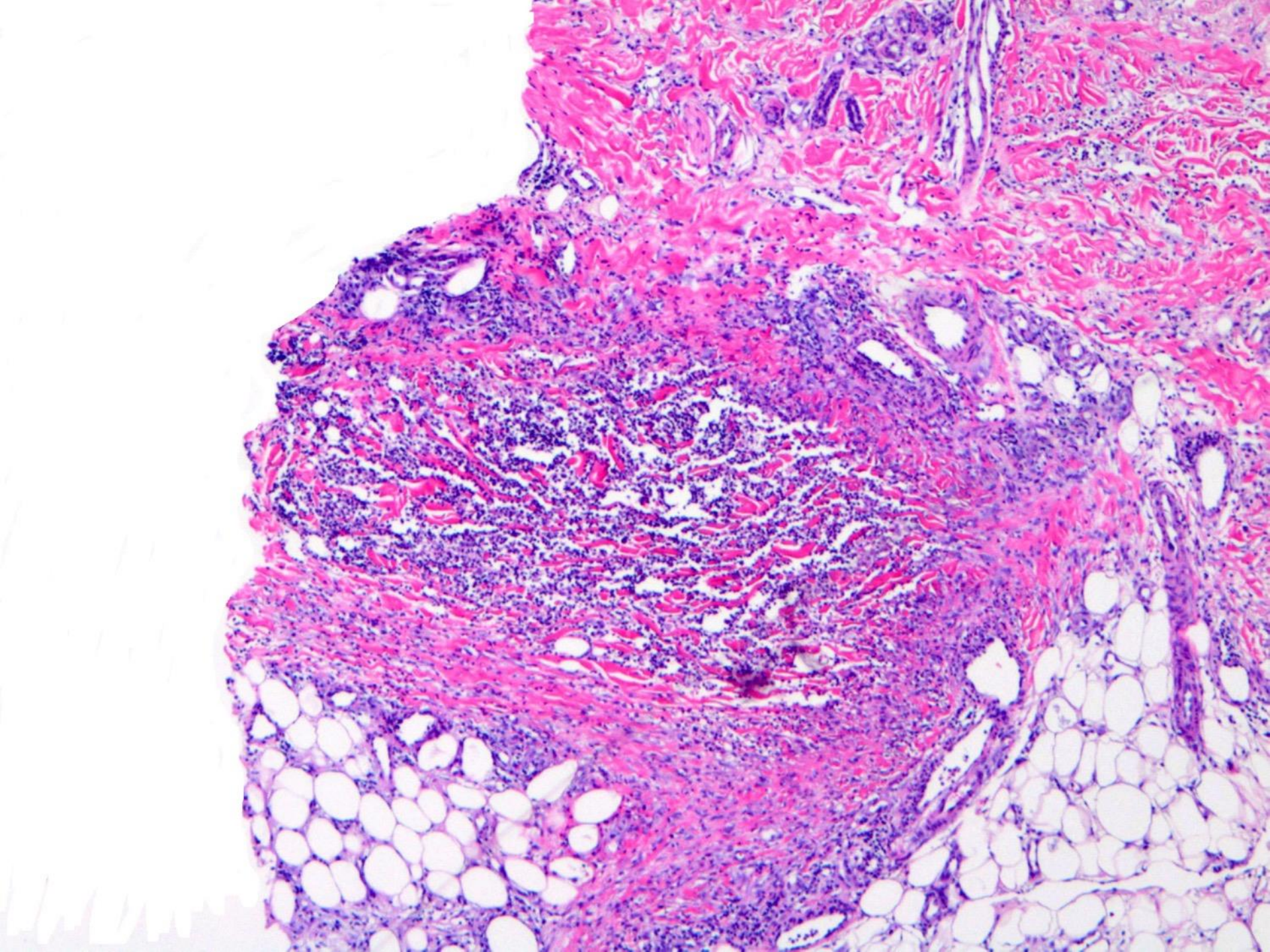


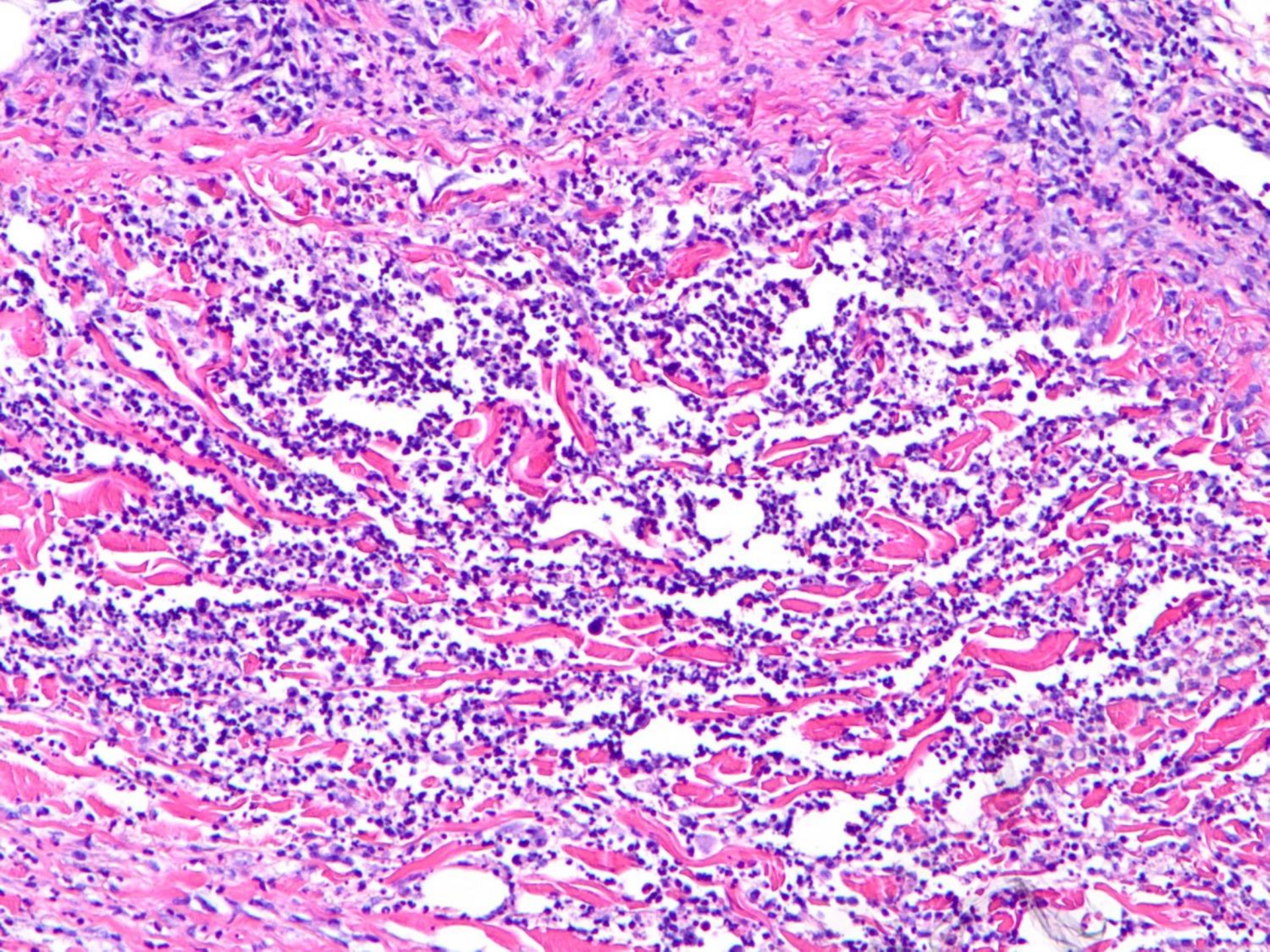












Alpha-1-antitrypsin deficiency
panniculitis, PiZZ type

PiZZ panniculitis: clinical

- Thighs, buttocks most common sites, proximal limbs
- Deep seated nodules often break down ulcerate with oily discharge; painful
- Often follows minor injury
- Systemic signs and symptoms, fever, effusions

PiZZ panniculitis: pathology

- Septal and lobular panniculitis (I + II +?III)
- Necrosis of adipocytes
- Neutrophils in the reticular dermis, splaying between collagen bundles
- Elastolysis, vaculitis

Differential diagnosis

- Pyoderma-infectious panniculitis –get stains
- Traumatic panniculitis
- Factitial panniculitis (polarize, get good clinical history)

Genotyping is important!

- Most severe types require replacement therapy
- Heterozygotes can be managed medically – dapsone/minocycline

Case 5A

- A 30 year-old woman from Ecuador presented with skin lesions in both legs.
- She had a cosmetic procedure



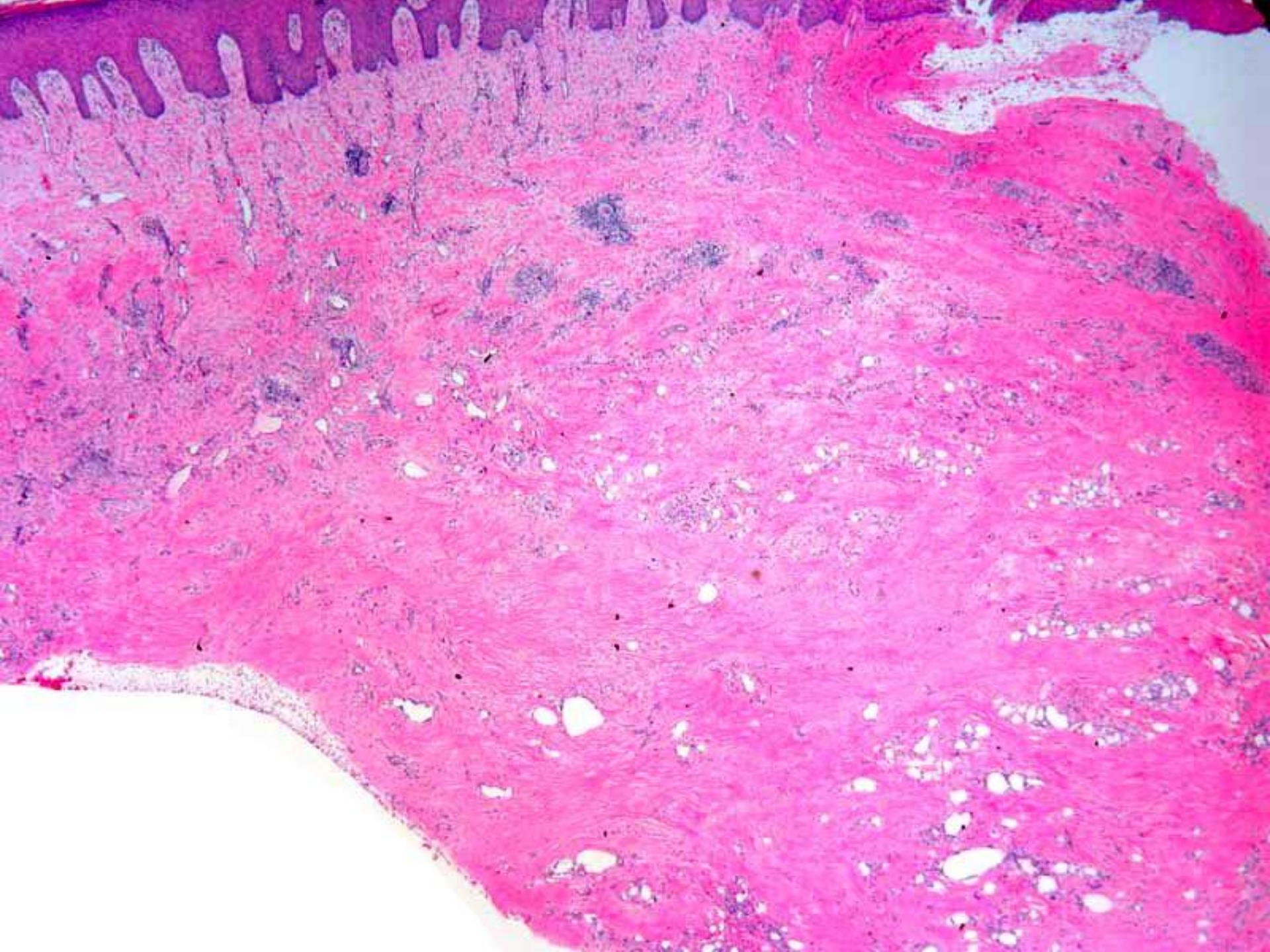


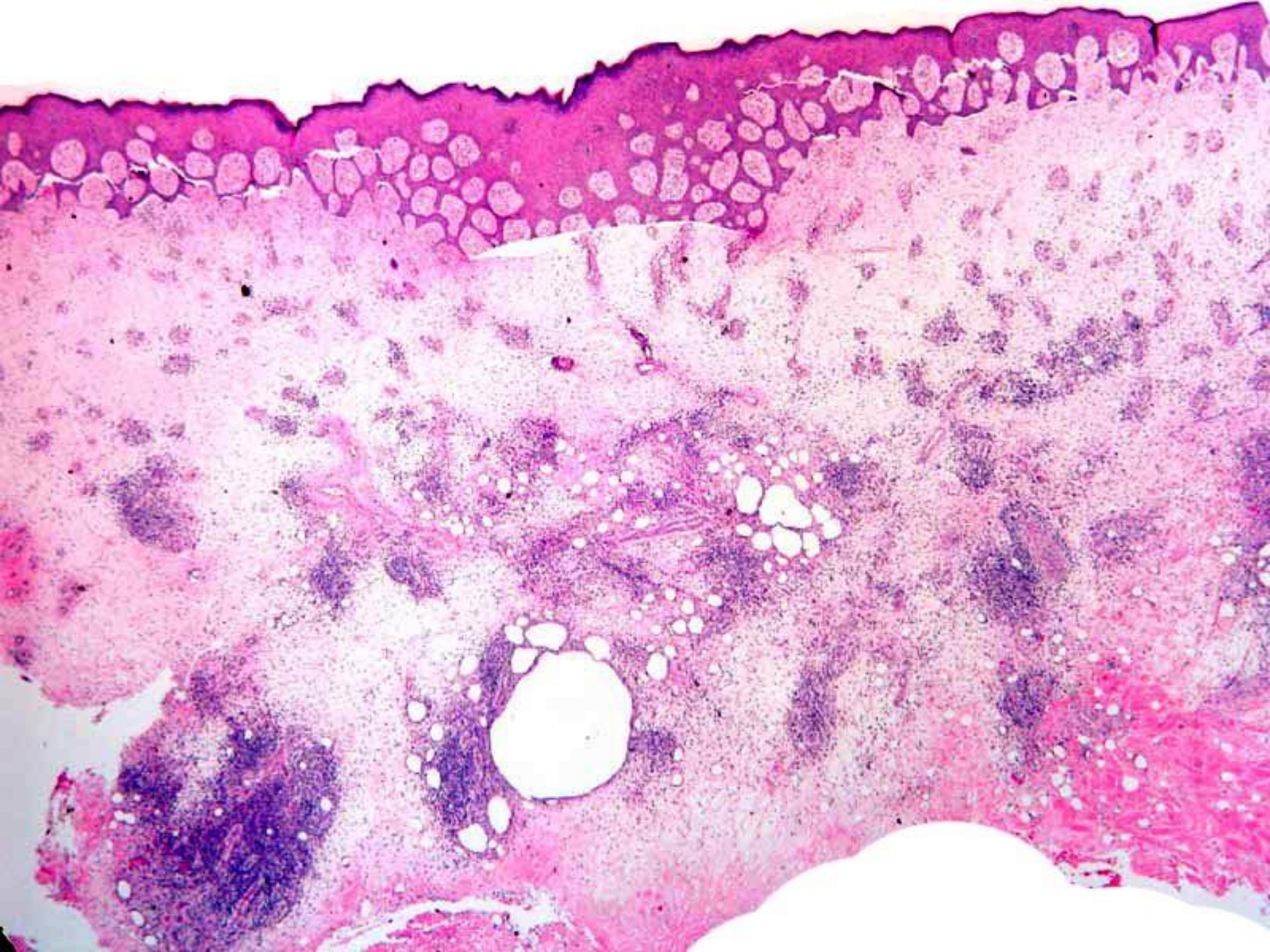


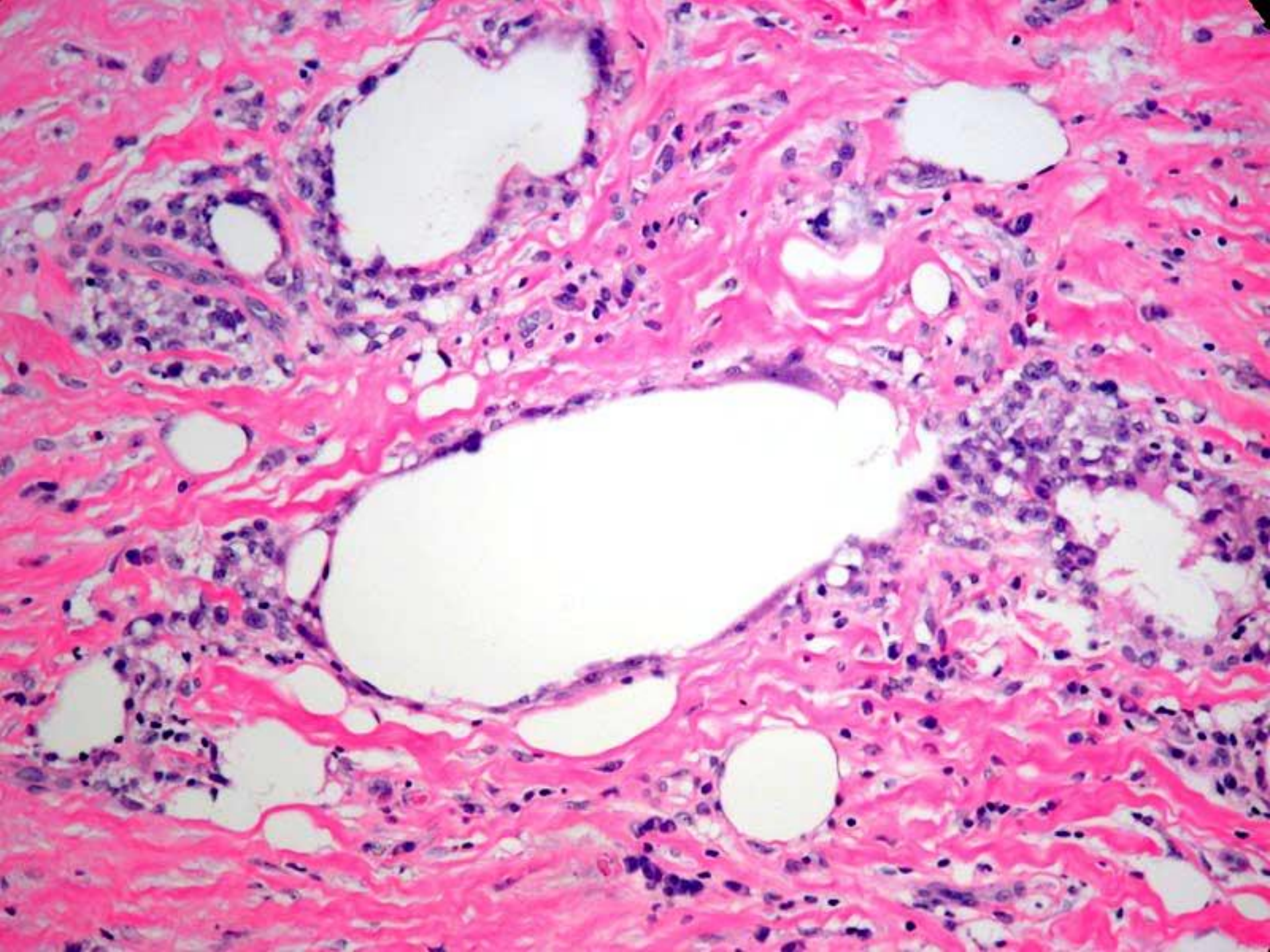


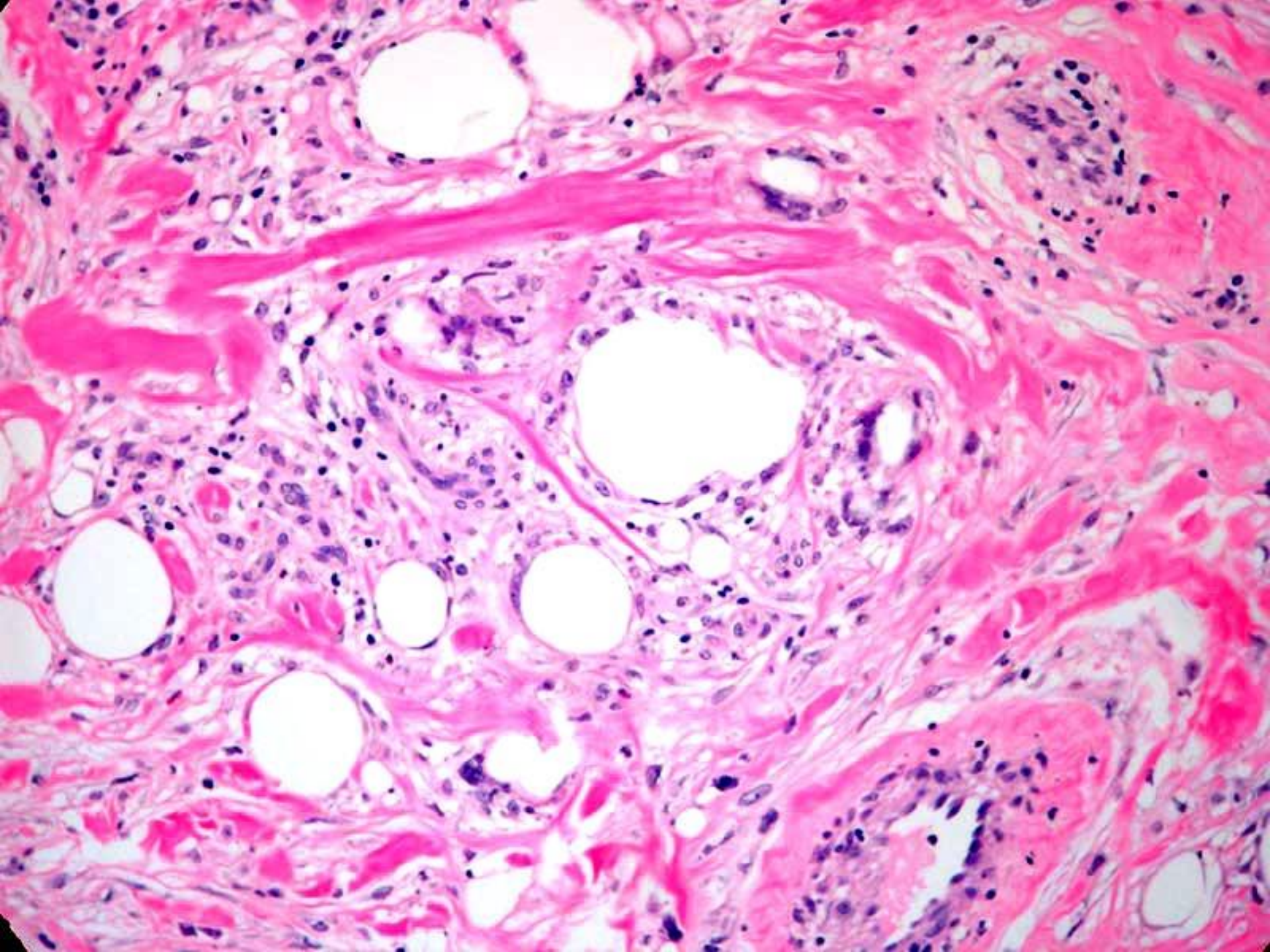


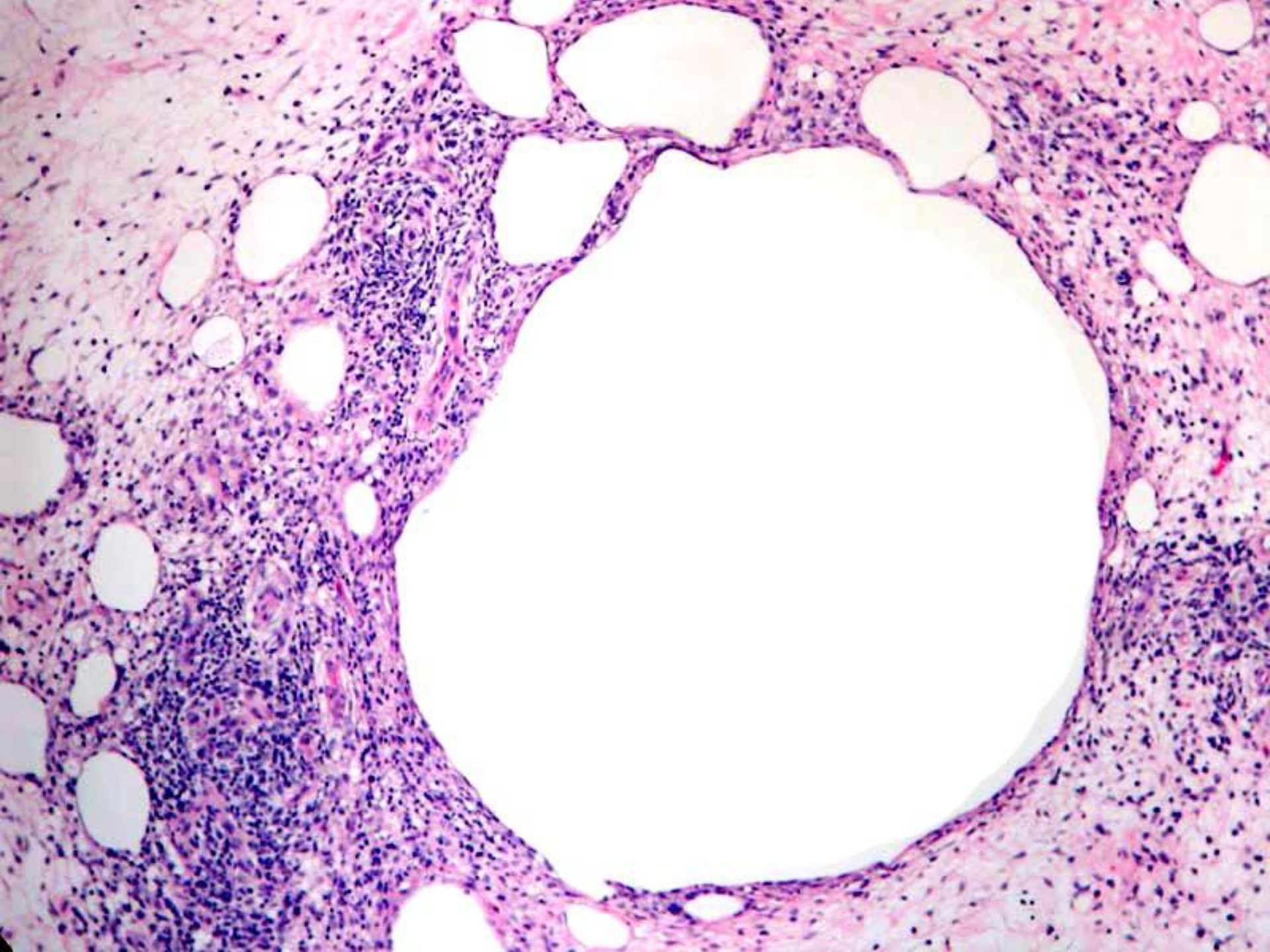












Factitial/"iatrogenic" panniculitis

Pathology

- Foreign body reactions are often deep seated and mimic a panniculitis (II +....)
- Sclerosing lipogranuloma is the chronic phase of the foreign-body reaction
- Macrophages fragment and try to phagocytose paraffin.
- Large drops are surrounded by multinucleated giant cells.
- Stromal fibroblasts produce collagen separate the vacuoles filled with paraffin

Case 5B

- **34 year old female**
- **Recent visit to “cosmetic” clinic**





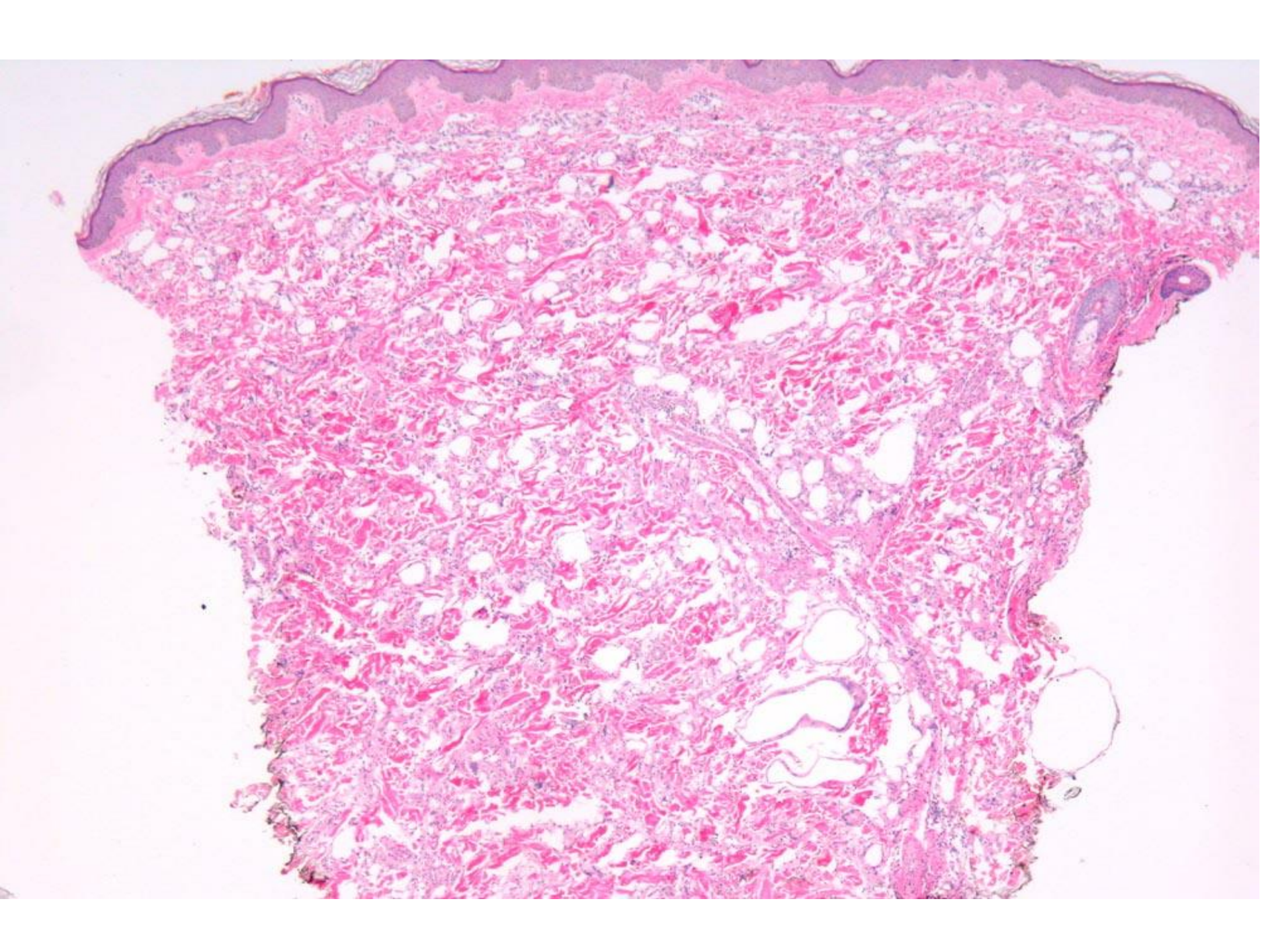
blanching

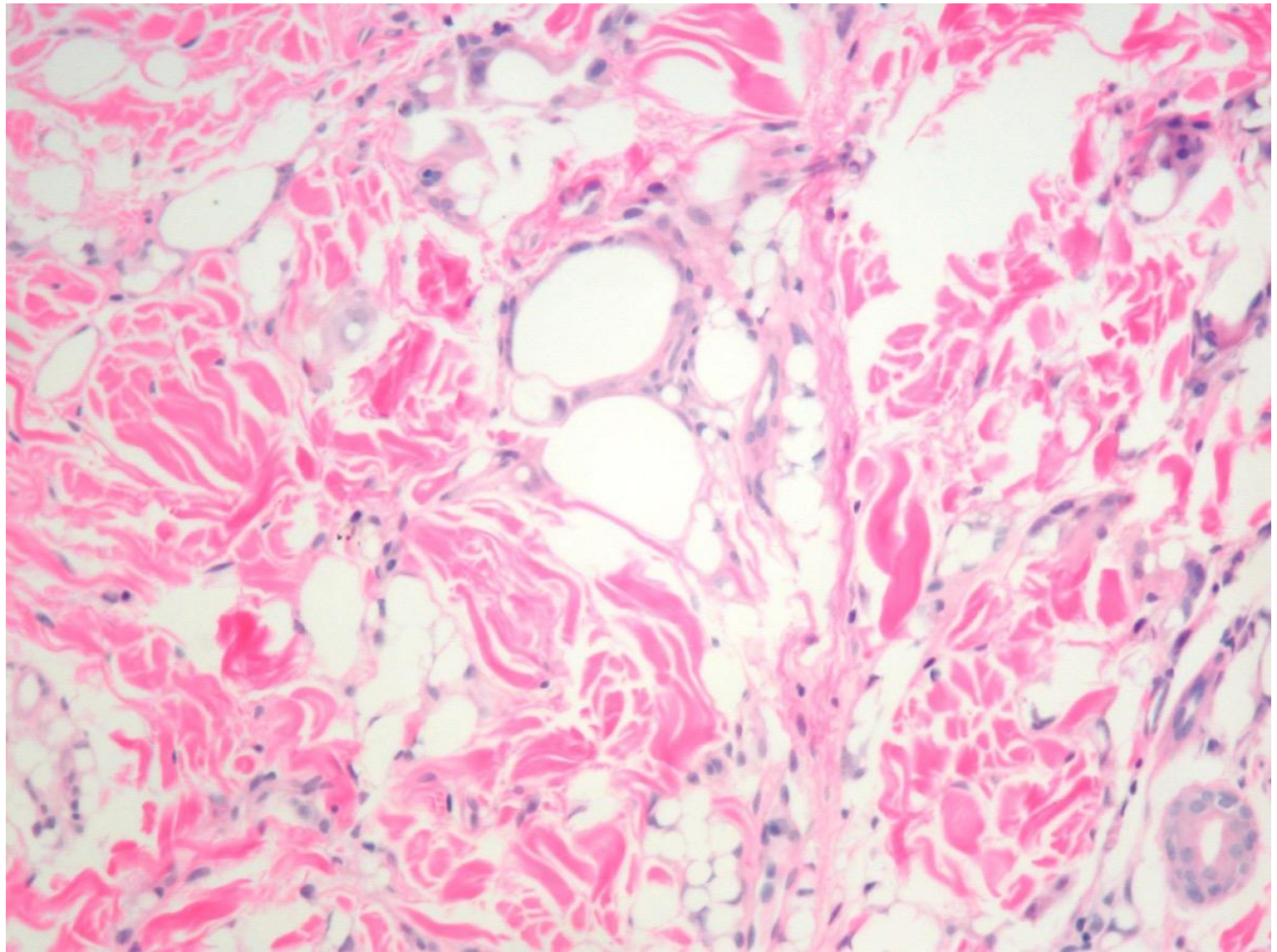


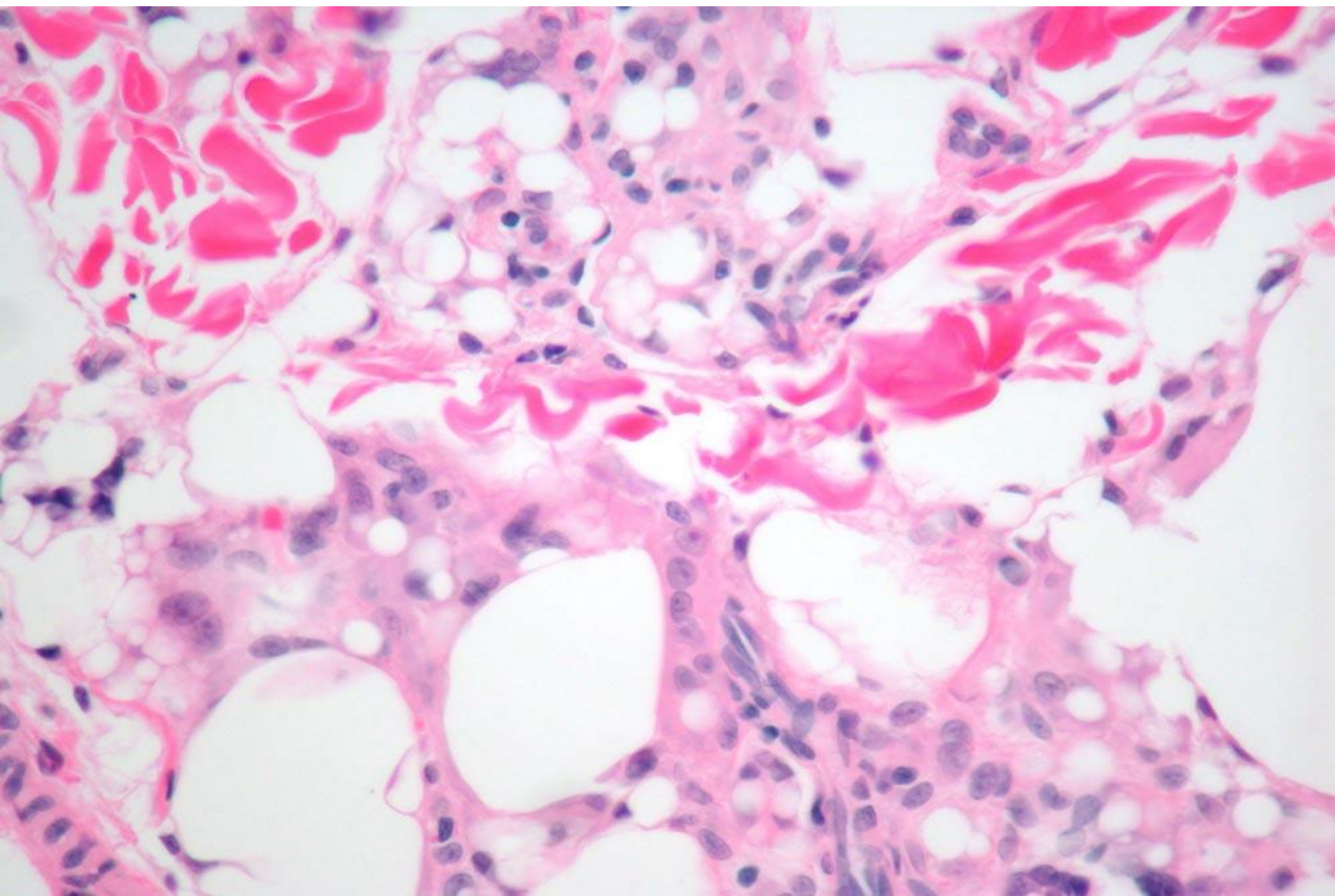
Post 12d of clobetasol, pre-Aldara,
4/2/04

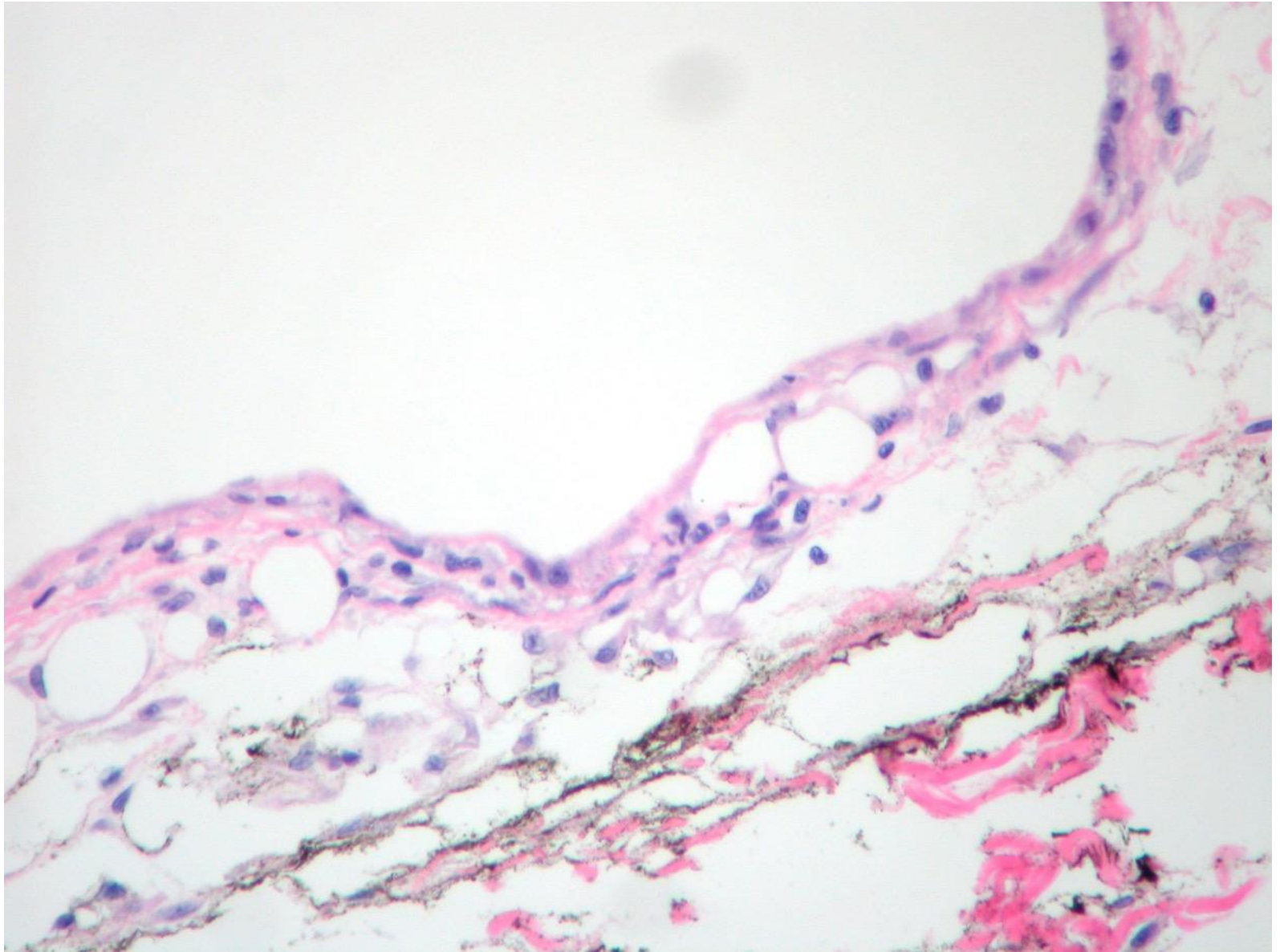
Biopsy site

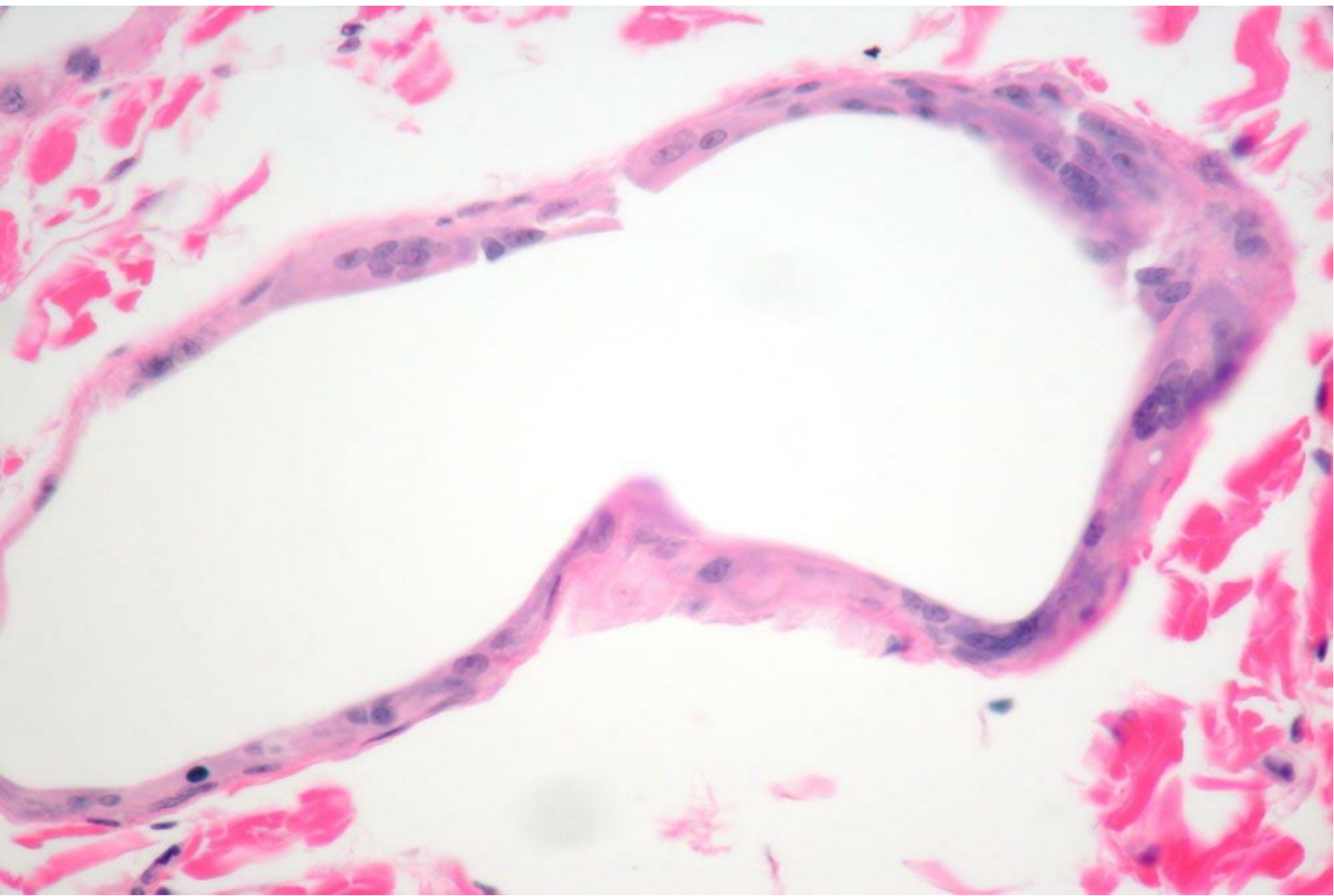












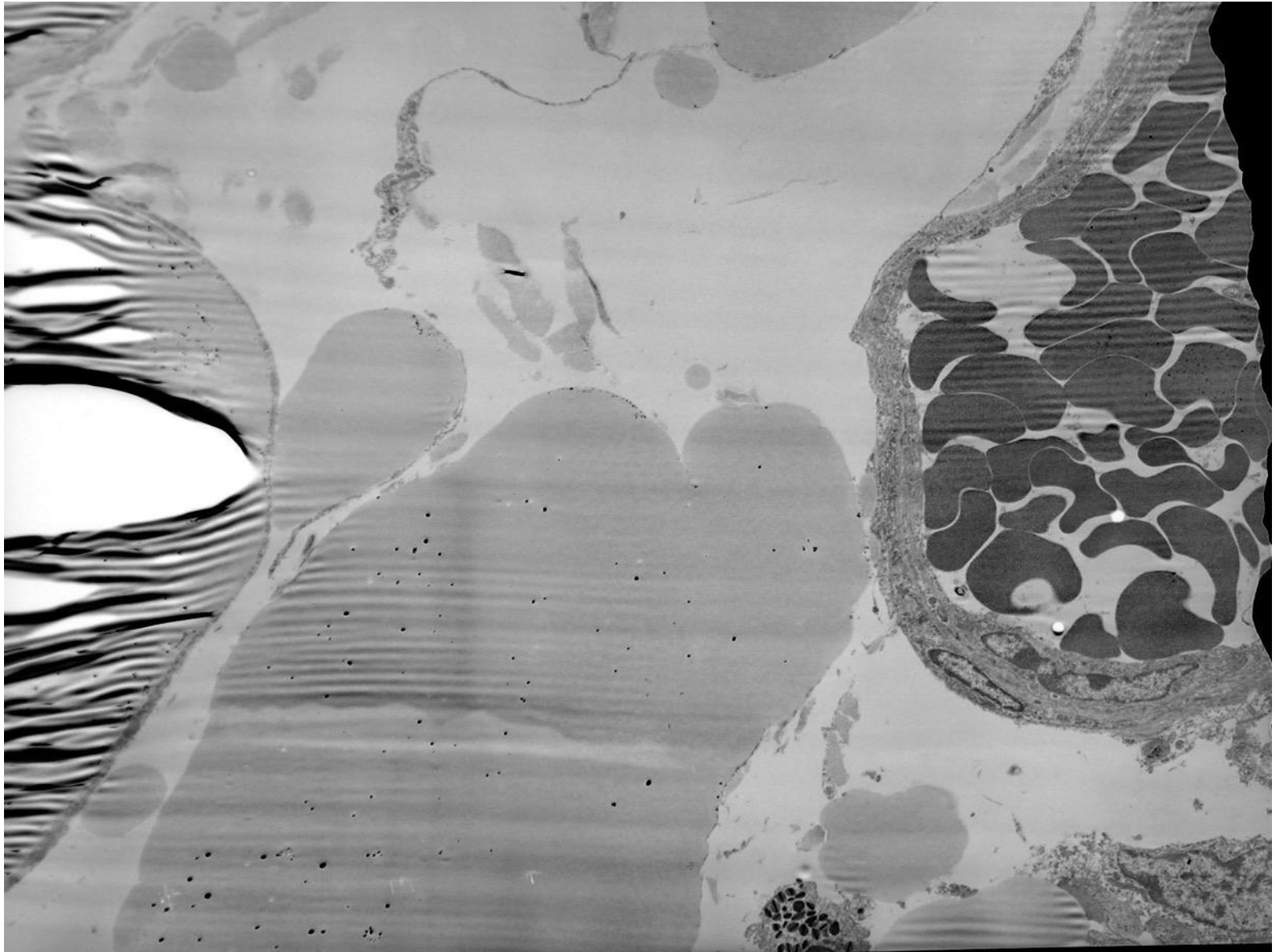
Factitial/"iatrogenic" panniculitis

Clinical presentation

- Indurated plaques, sometimes distant from the primary injection site; foreign body reactions
- Lymphedema
- Sclerodermoid reactions
- Ulceration
- Systemic signs and symptoms; malaise, fever, lymphadenopathy, pneumonitis, embolism
- “Adjuvant disease”

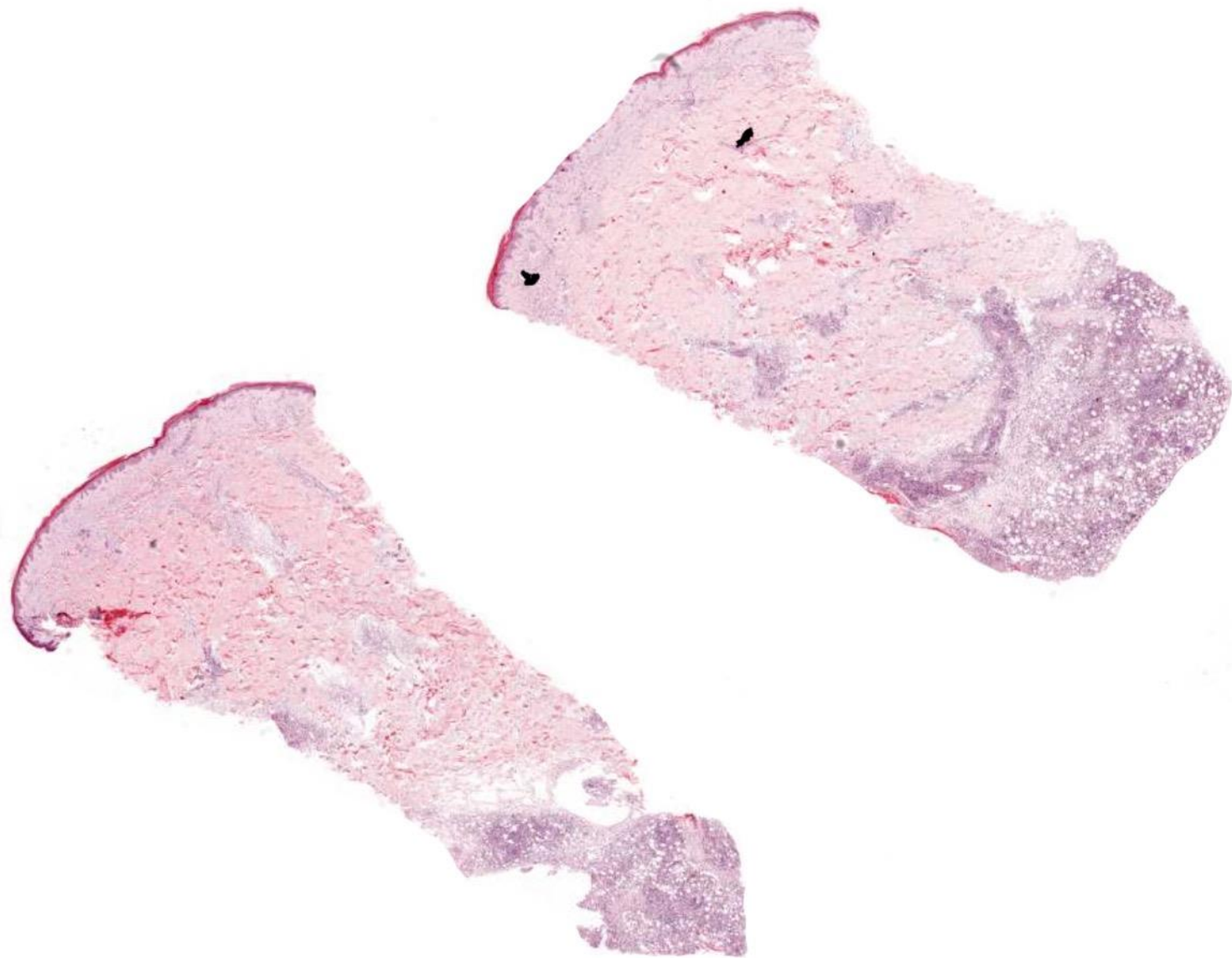
Pathology

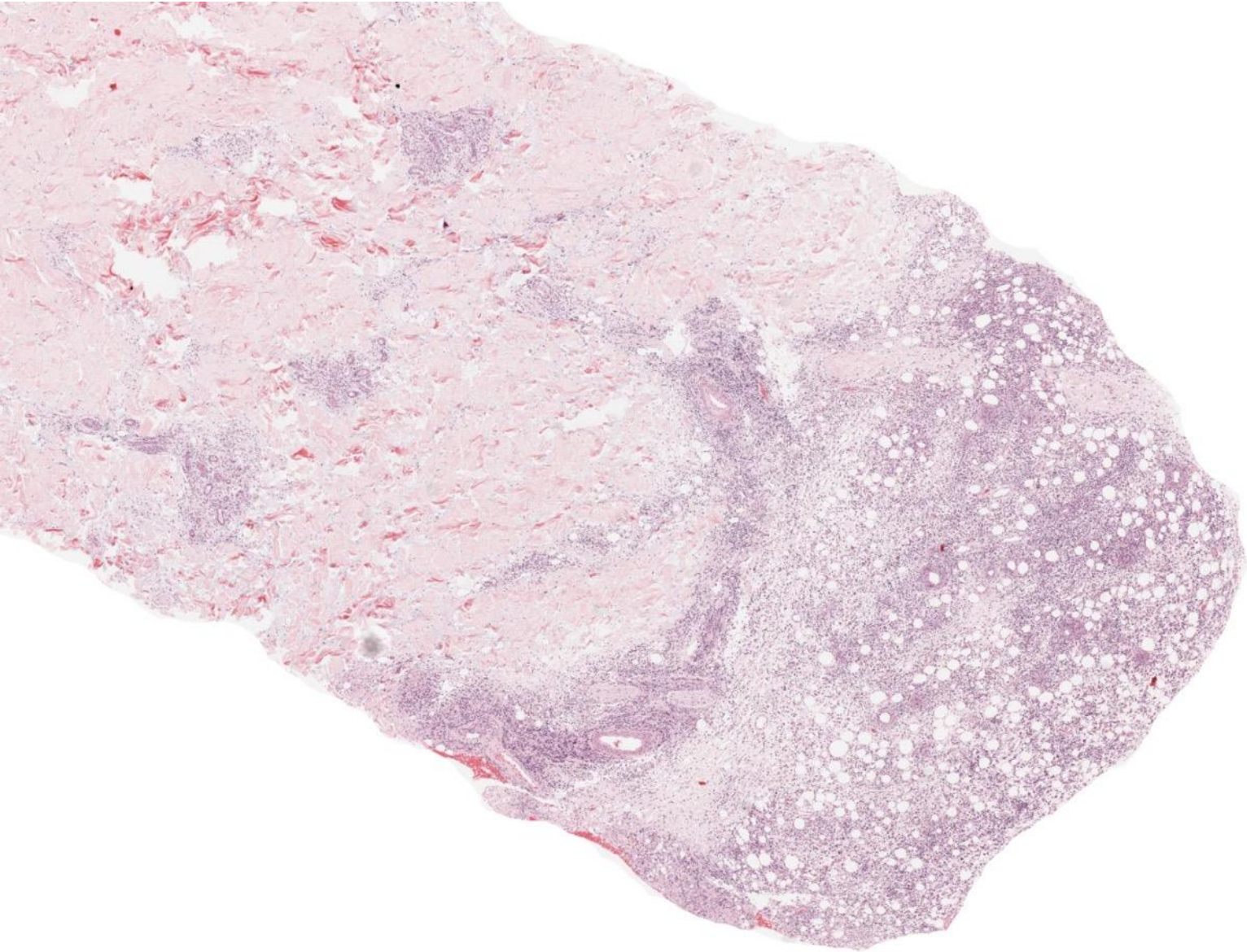
- Multiple small vacuoles simulating fat throughout the dermis; vacuoles of varying size (II +)
- Rare areas of refractility
- Multinucleated giant cells often surrounding the vacuoles (foreign body type)
- Fibroblastic reaction
- Sarcoidal reaction
- Can use EM/EDS if necessary to confirm

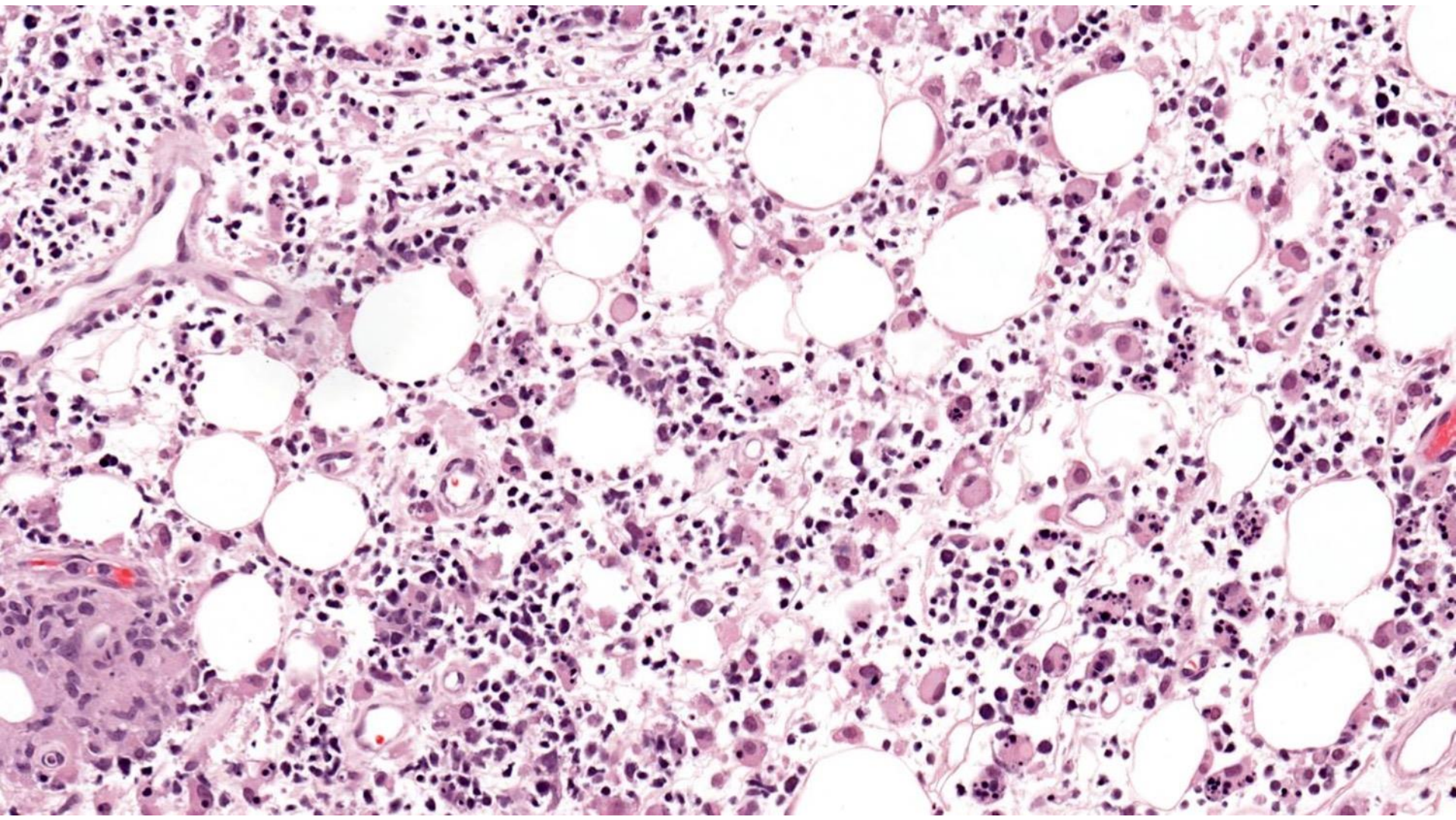


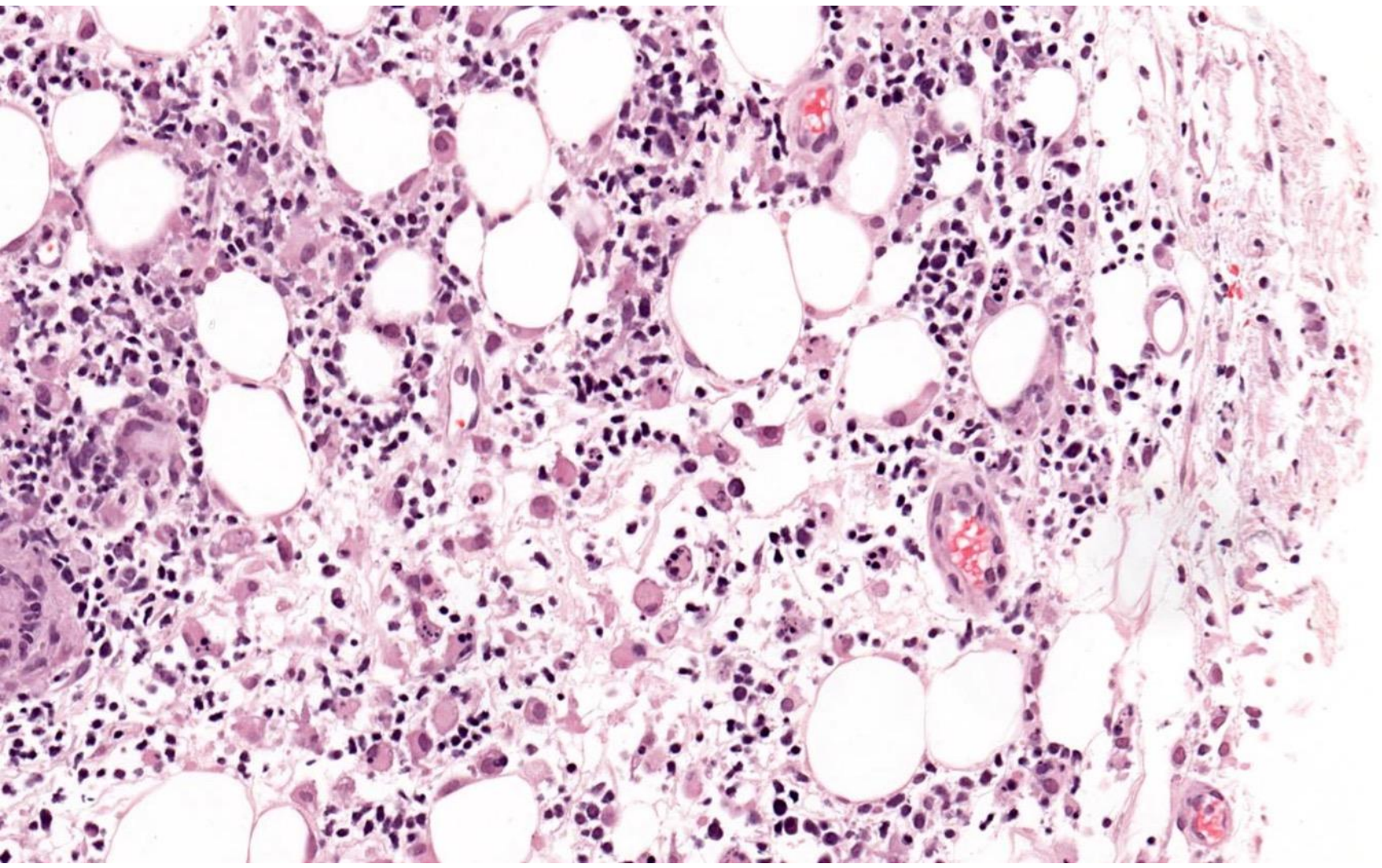
Case 6











Cytophagic histiocytic panniculitis

Clinical features

- Patients with subcutaneous nodules, lower legs, forearms, trunk
- Most with prominent systemic signs and symptoms
 - Fever and malaise
 - Hemorrhagic diathesis
 - Organ failure
- May represent :lymphoma; viral, immune dysregulation or unclassified
- Prognosis: variable

Pathology

- Lobular panniculitis (II)
- Histiocytes and lymphocytes
- Prominent phagocytosis erythrocytes, leukocytes, nuclear debris-bean bag cells
- Atypia only if overt lymphoma is present

Etiology?

- Most likely represents multiple disorders-
benign and malignant

**Histiocytic cytophagic panniculitis Molecular
evidence for a clonal T-cell disorder**

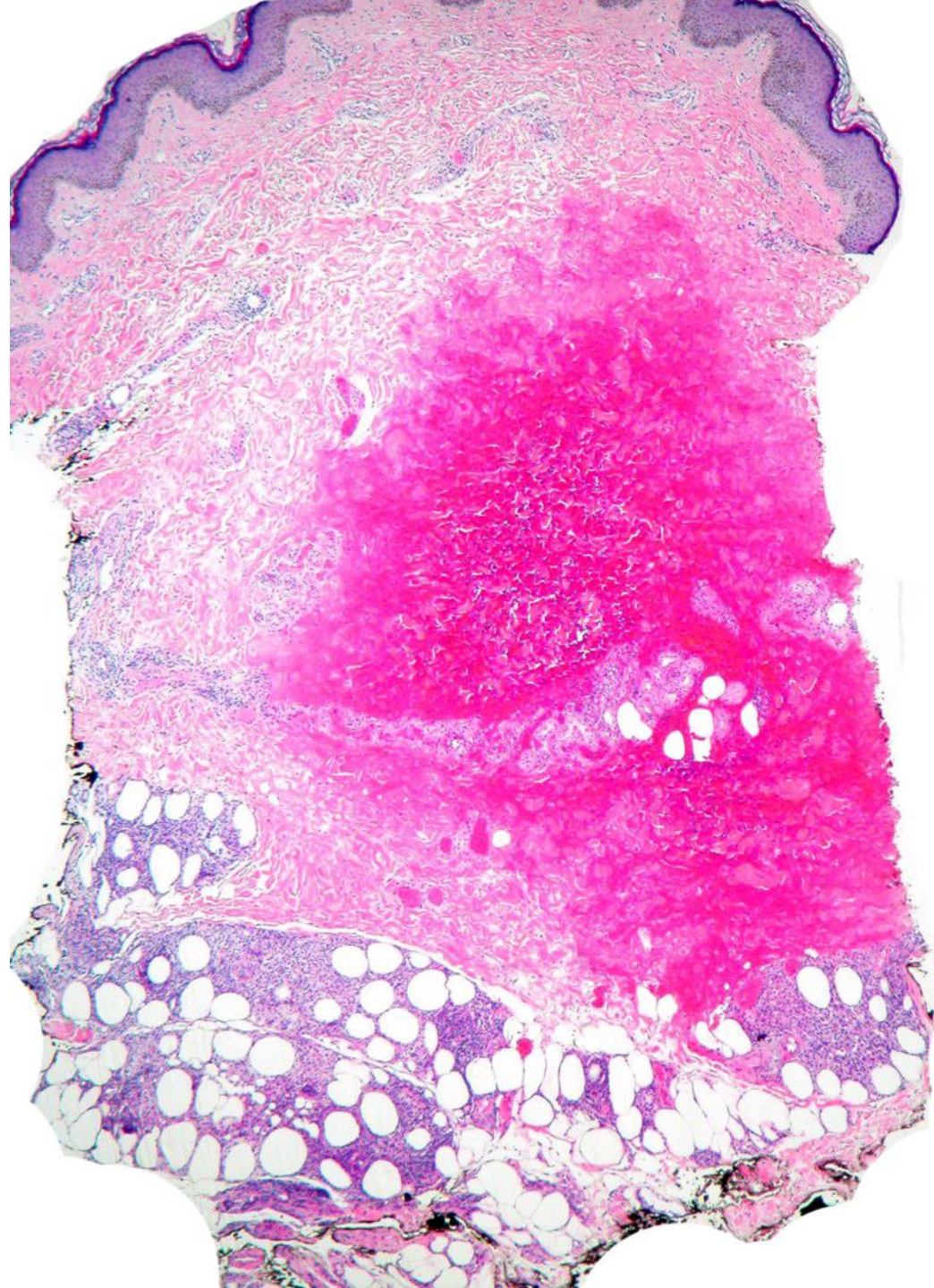
Prodromos Hytiroglou, MD, Robert G. Phelps,
MD, Debra J. Wattenberg, MD, and
James A. Strauchen, MD, *New York, New York*
JAAD 1992

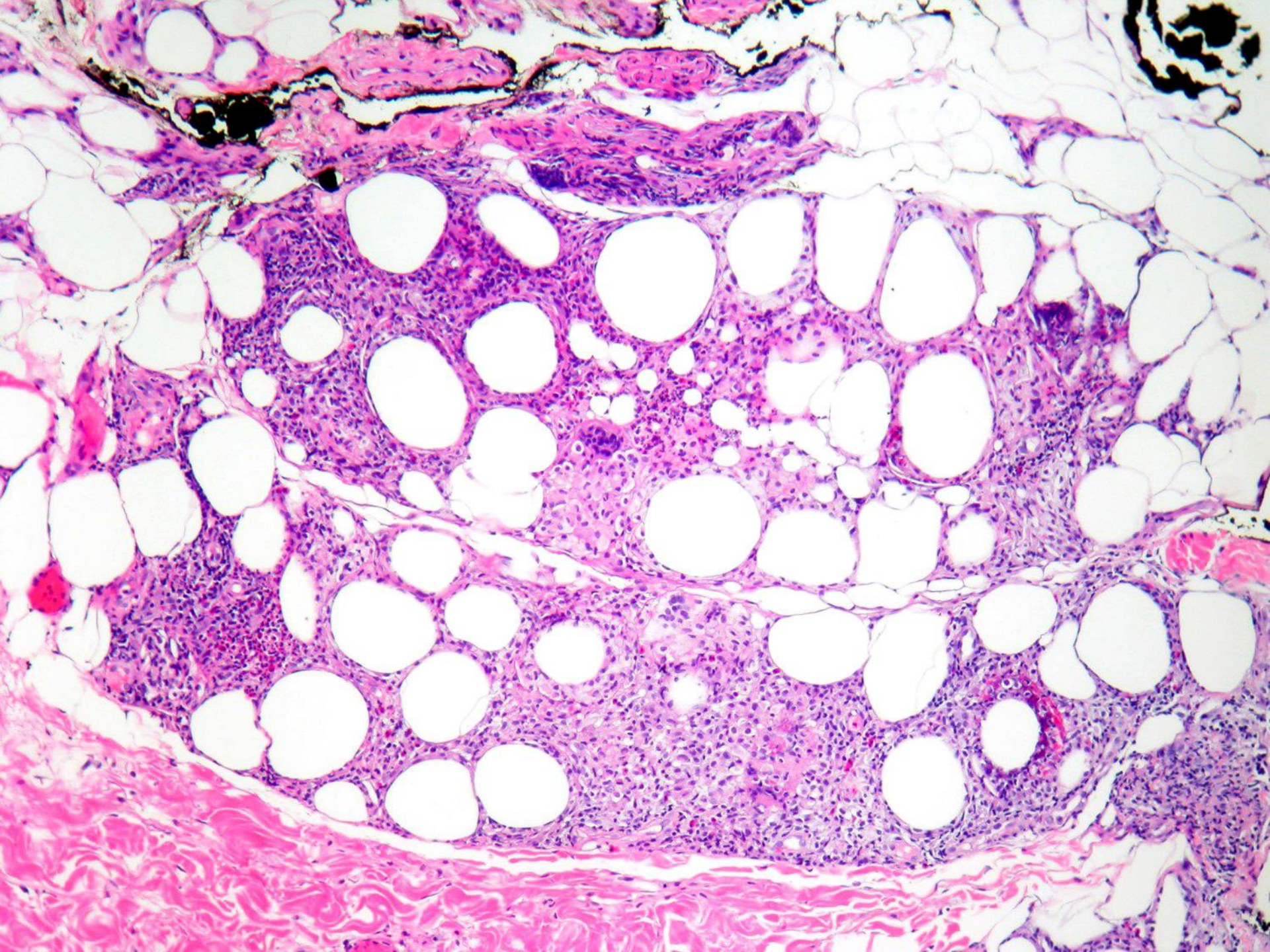
Case 7

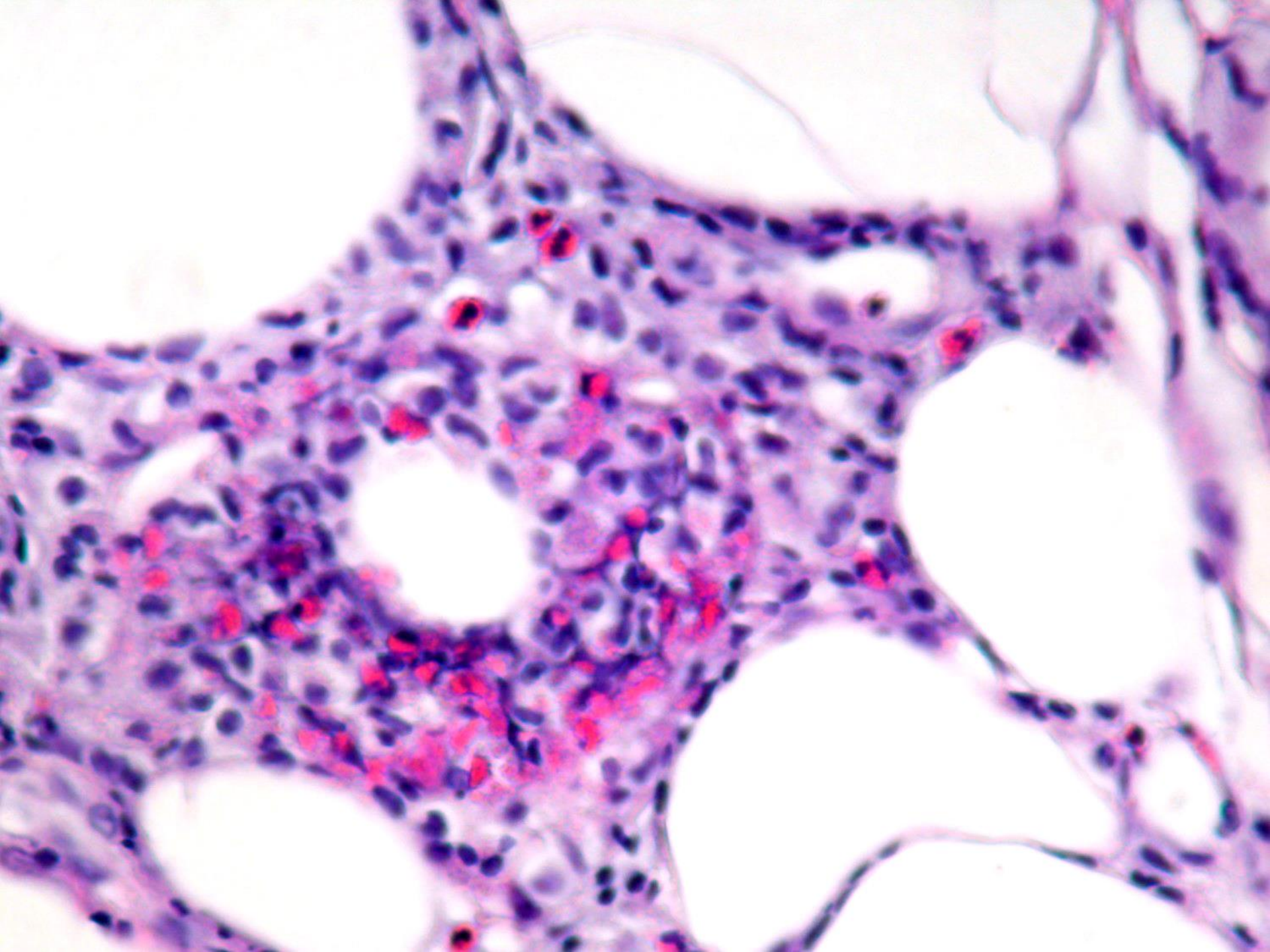
49-year-old African American woman presented to the faculty practice with a chief complaint of redness on her right arm for 4 months.

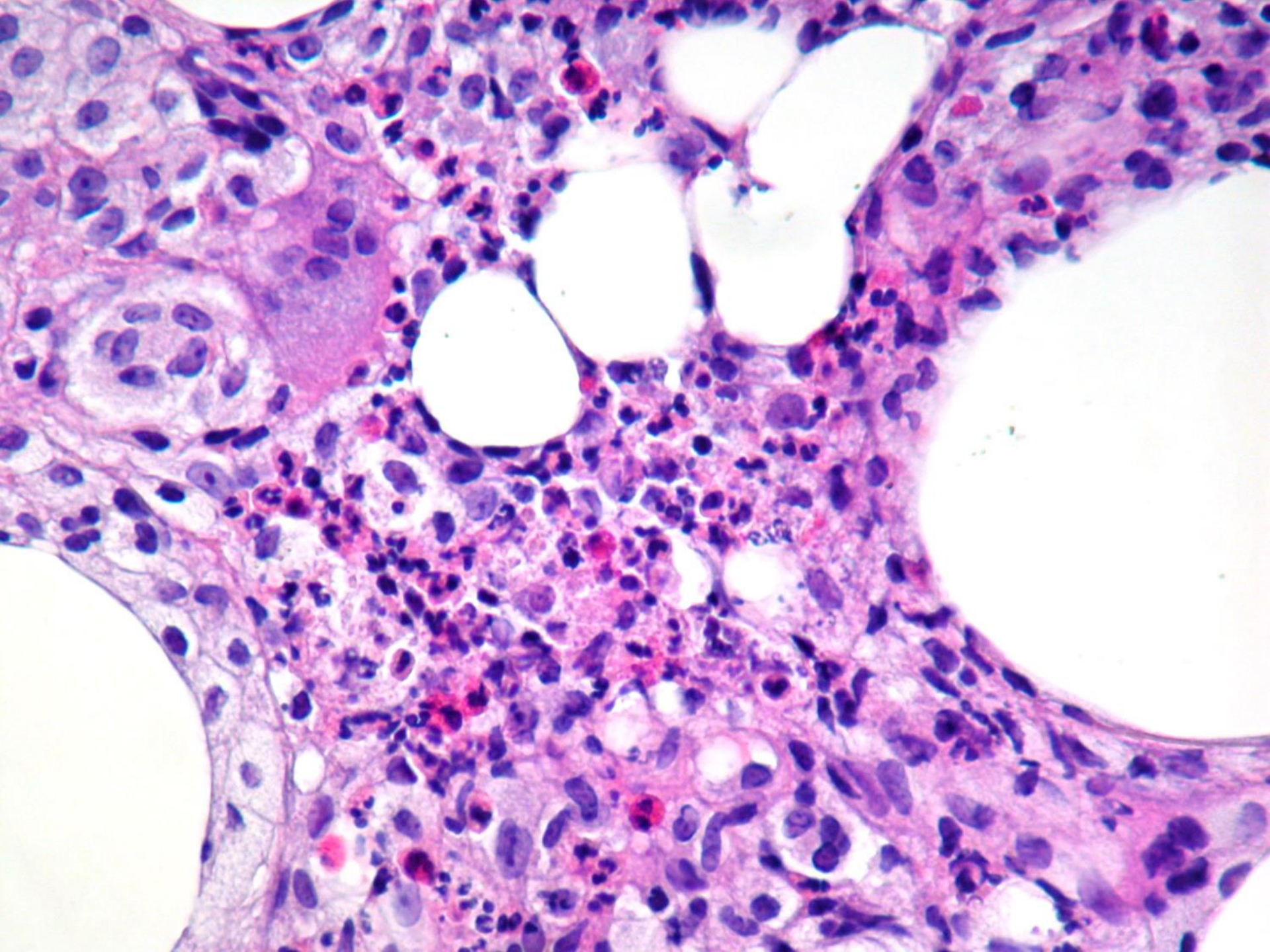












Eosinophilic panniculitis

Clinical features

- Nodules and plaques
- Papules and pustules
- Single or multiple
- Legs, arms, trunk or face

Eosinophilic panniculitis

- Eosinophilic panniculitis is the term used to refer to those cases of septal or lobular panniculitis in which eosinophils predominate in the inflammatory infiltrate (II)
- **Does not represent a specific disease entity, but rather a histopathologic pattern**
- Because it is a nonspecific reactive process, it should be followed by pertinent clinical and laboratory investigations to rule out an associated systemic process

Eosinophilic panniculitis

- Bacterial infections
- Parasite infections with *Gnathostoma* and *Toxocara canis*
- Sarcoidosis
- Leukocytoclastic vasculitis
- Systemic vasculitis
- Wells' syndrome (eosinophilic cellulitis)
- Deep morphea
- Sjögren's syndrome
- Asthma
- Deep insect bites
- Atopic dermatitis

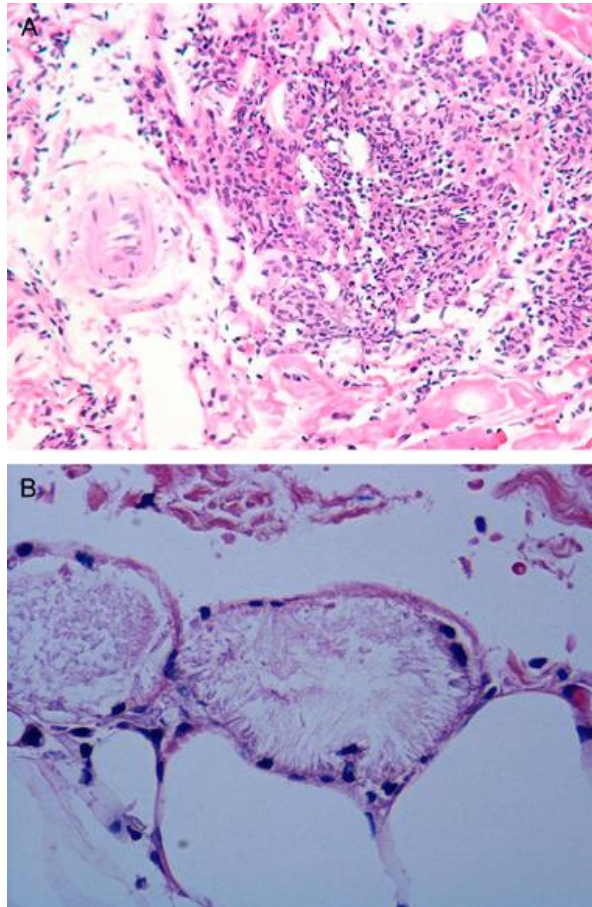
Case 8

Three year old boy with history of
nephrotic syndrome

Poststeroid panniculitis



Poststeroid panniculitis



Poststeroid panniculitis

Clinical

- Rare complication of systemic corticosteroid therapy usually after cessation of systemic steroid therapy
- Multiple subcutaneous nodules often on cheeks, arms, trunk
- Usually heals quickly

Pathology

- Lobular panniculitis
- Foamy histiocytes, neutrophils, giant cells
- Needle shaped clefts within adipocytes
- Crystallized saturated fatty acids-different composition in children
- Differential diagnosis subcutaneous fat necrosis of the newborn, sclerema neonatorum