

Case

- 52 yo F with systemic lupus
- On mycophenolate mofetil and prednisone
- **Presents unresponsive with rash on her right leg only**
- Was well the night before
- Rapidly developed multi-organ failure in ED

Hospital Day 1





Hospital Day 3



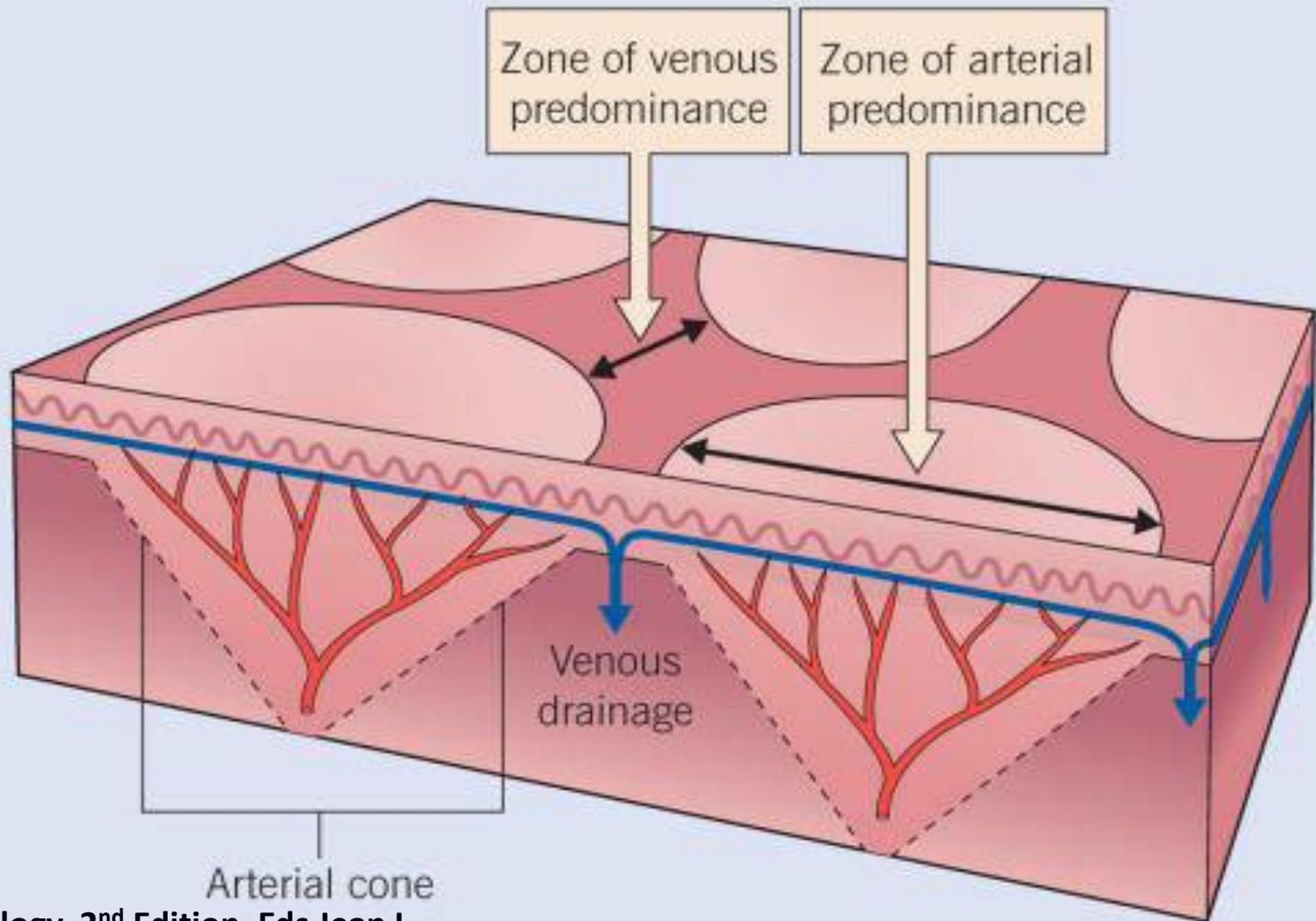


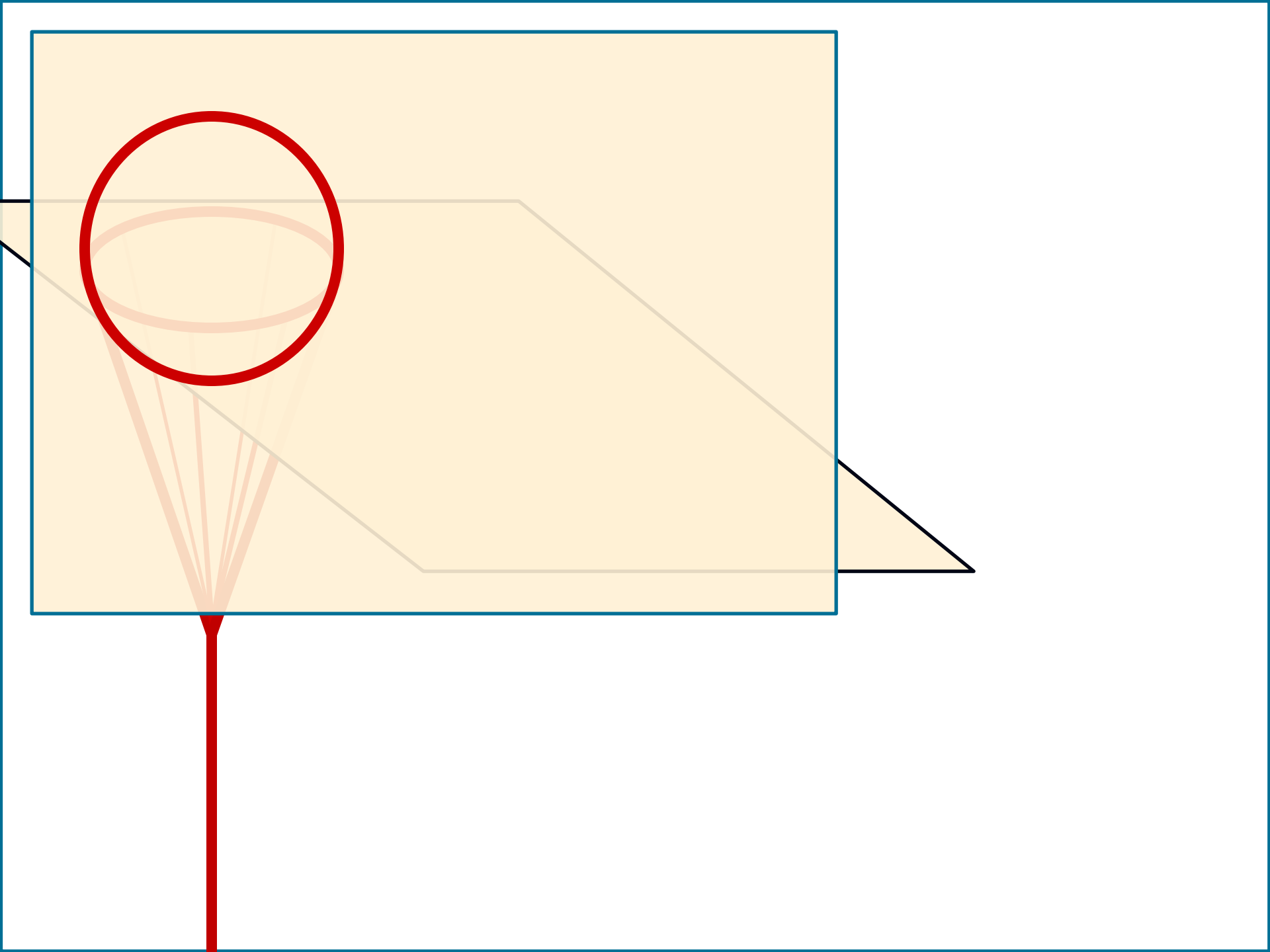


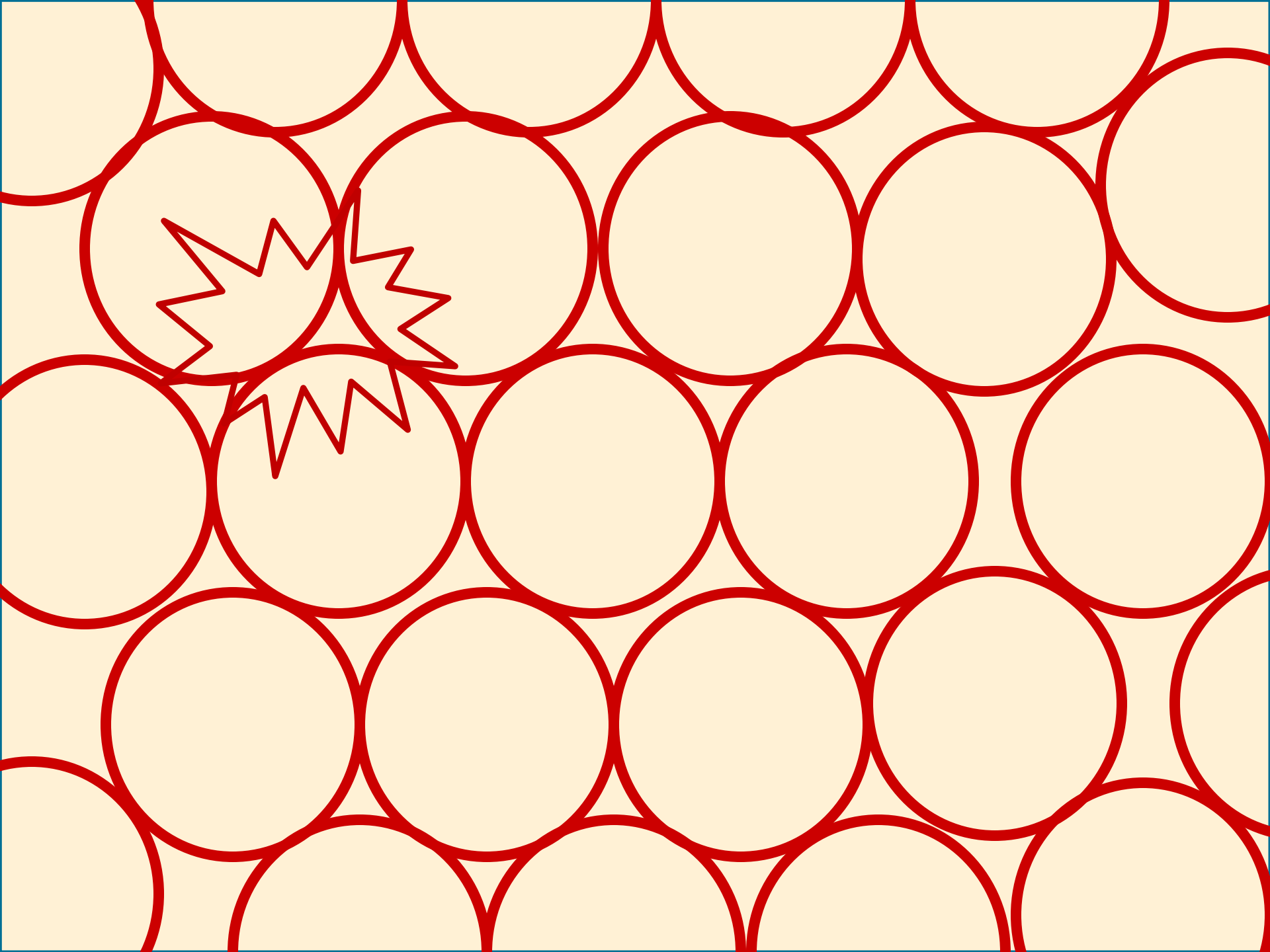
What can morphology tell us about pathophysiology?

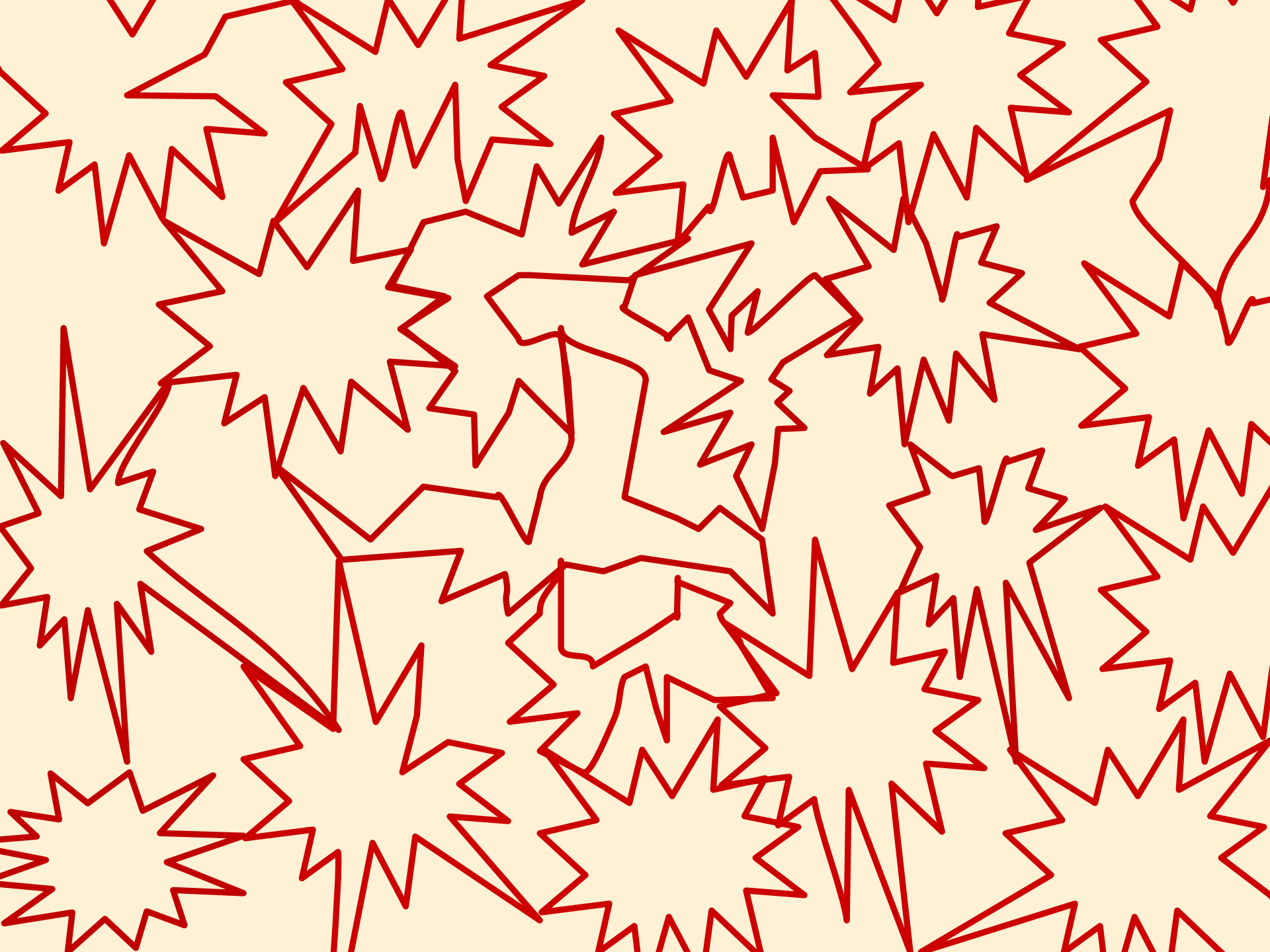


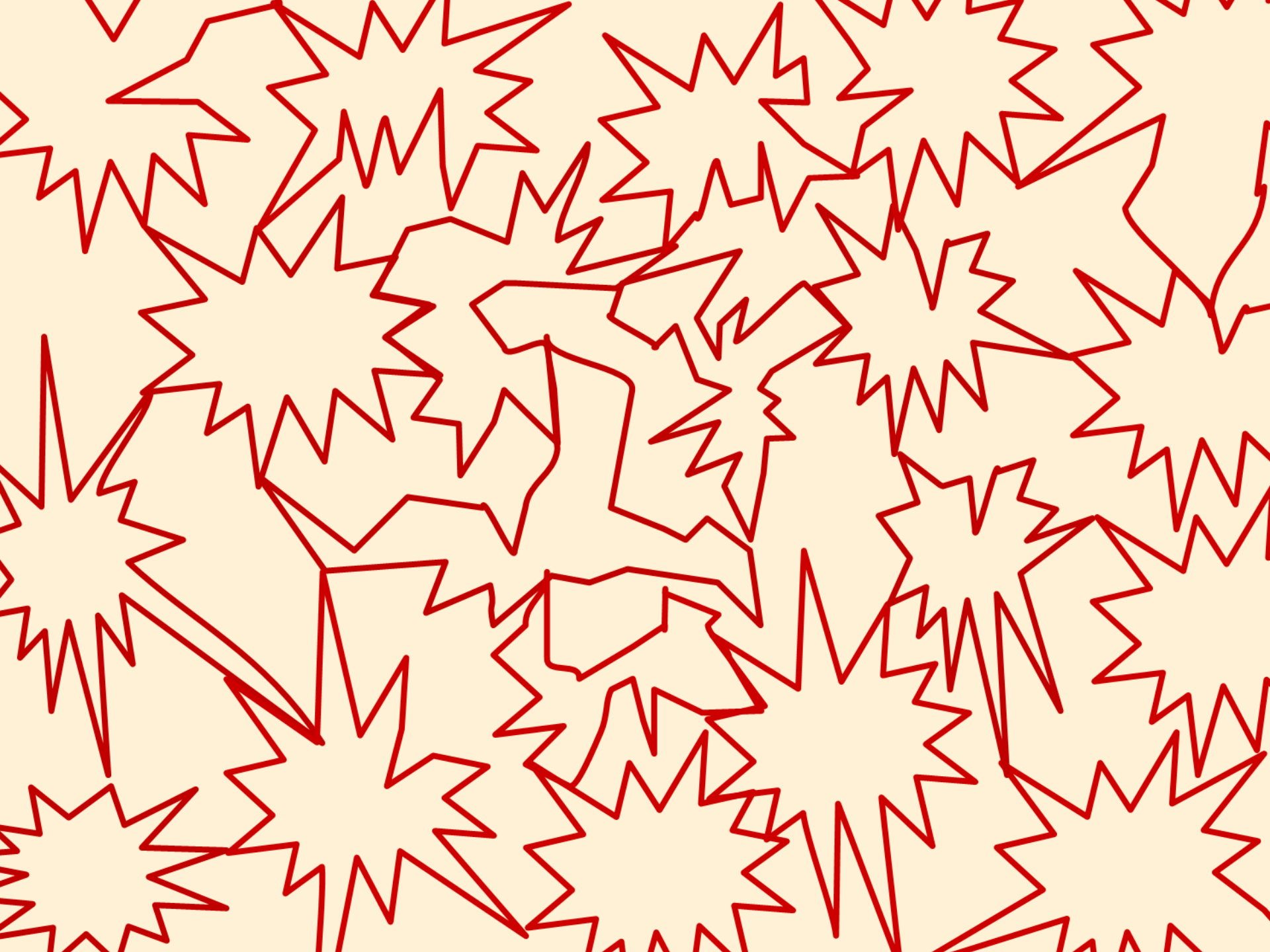
ANATOMICAL BASIS FOR THE DEVELOPMENT OF LIVEDO RETICULARIS

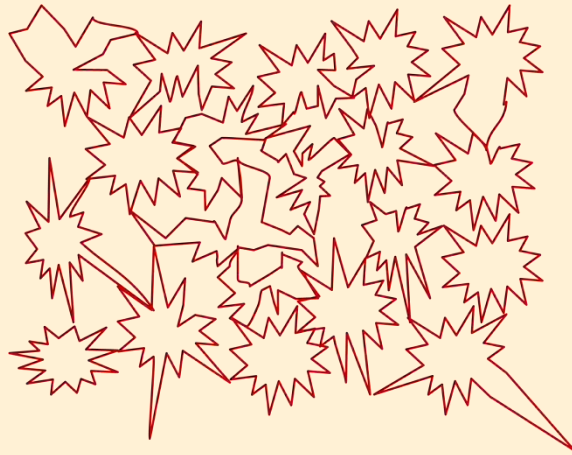
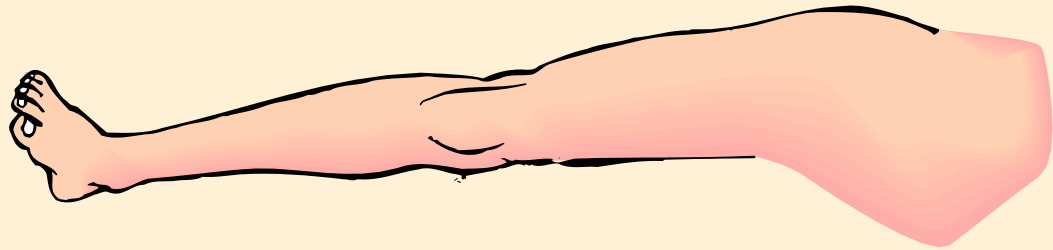


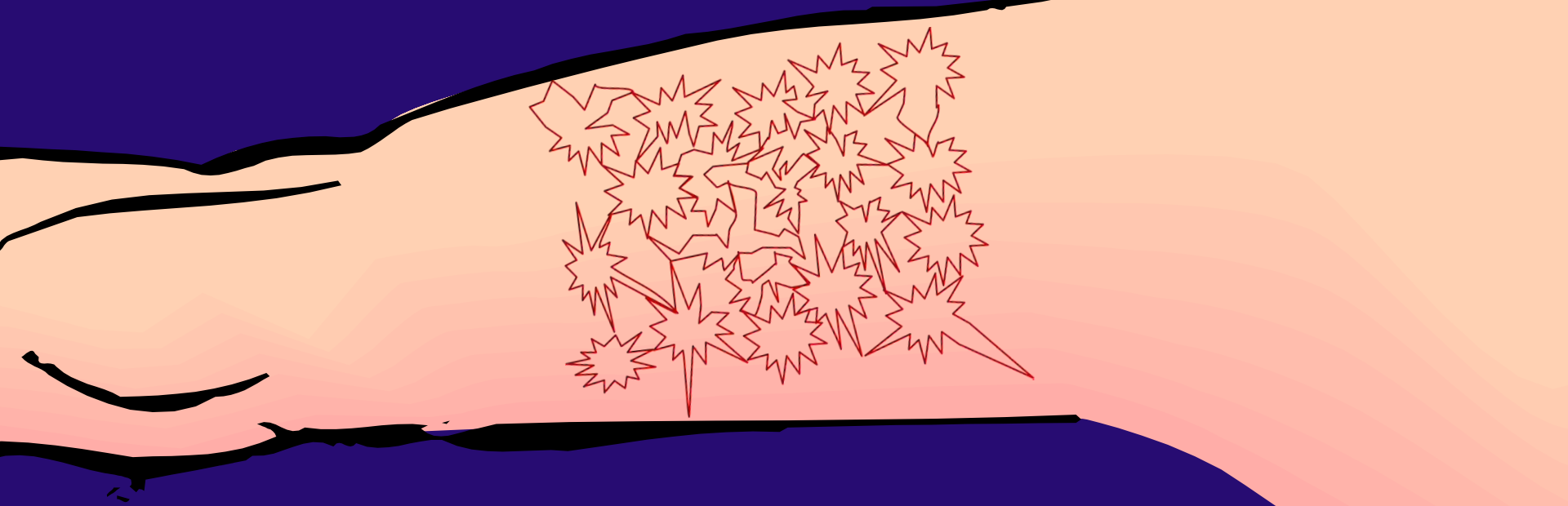










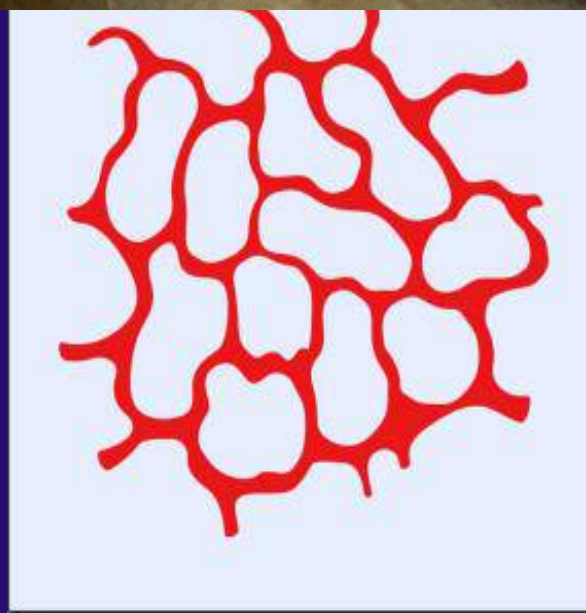
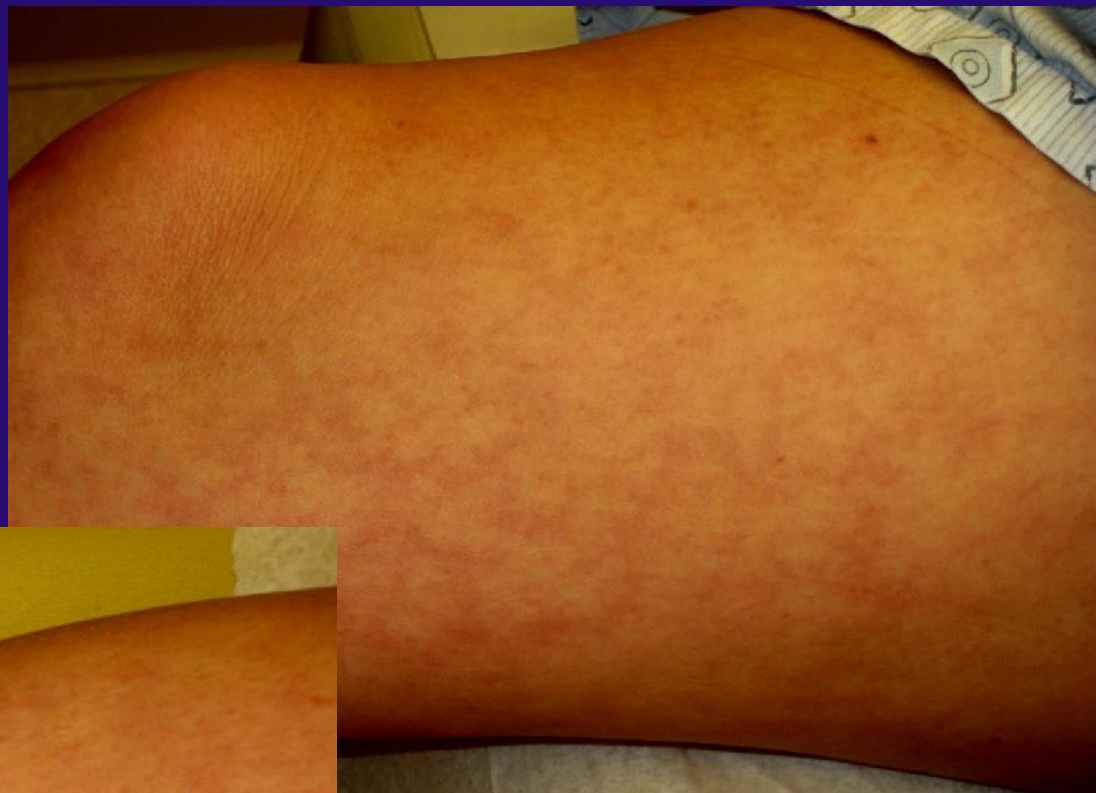


2 potential problems with this system

Problem 1: Livedo Reticularis

- Violaceous erythema
- Outlines 1-3cm stellate patches
- Surface of cones fed by individual perforating arterioles
- From enhanced visibility of zones of venous predominance
 - Increased deoxygenated blood in the venules
 - From engorged veins, constricted arterioles, local hypoxia...

Livedo Reticularis

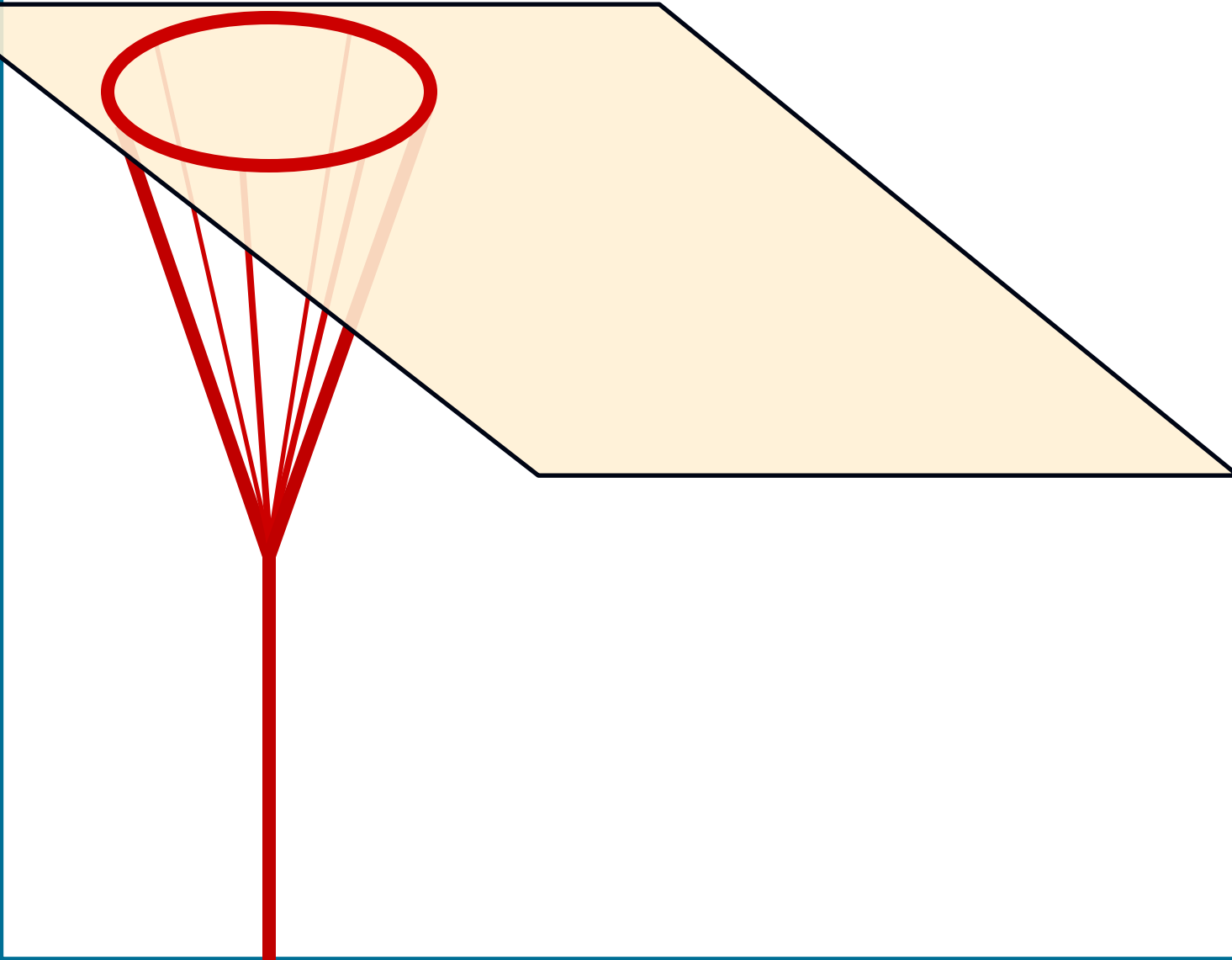


Problem 2:

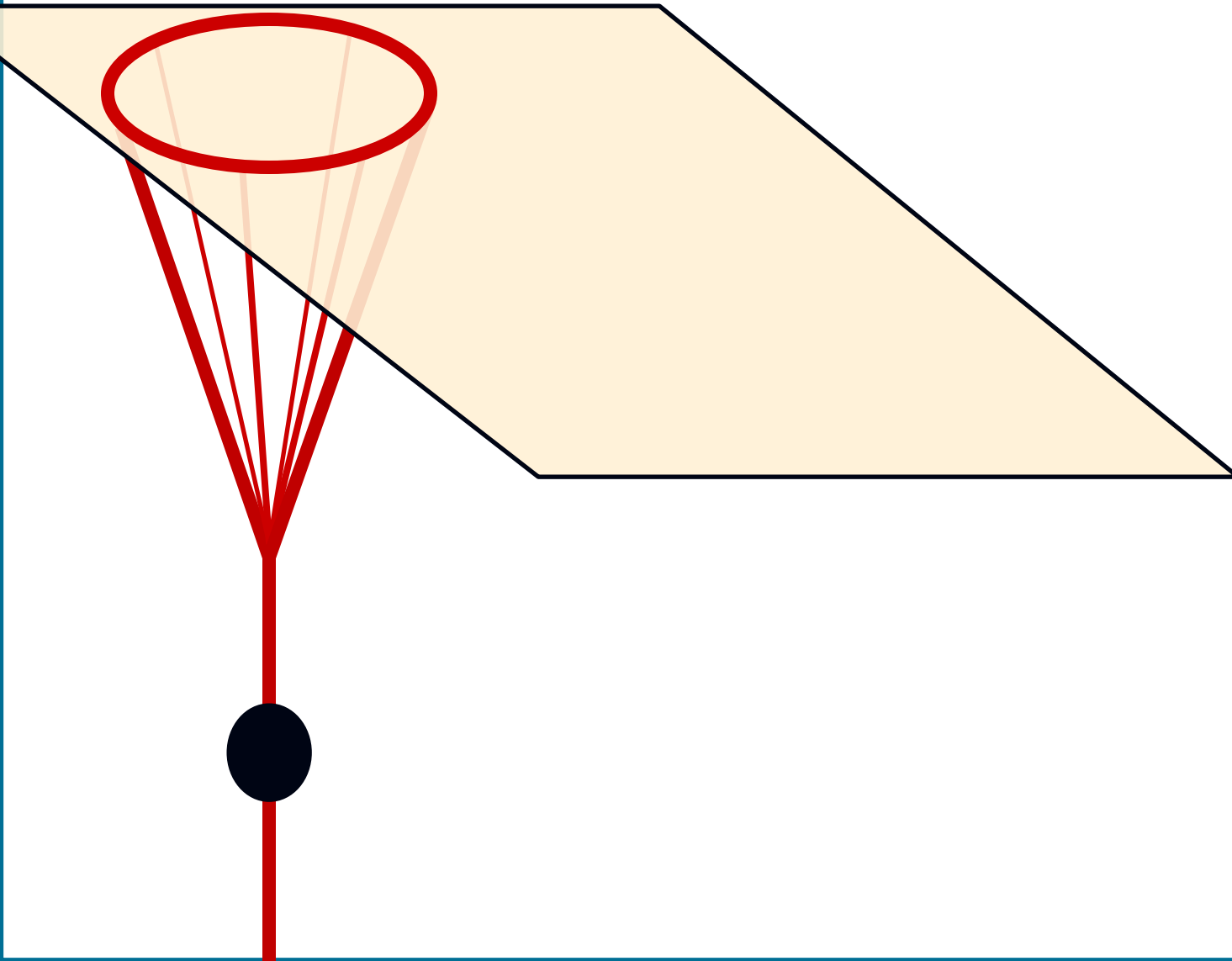
Retiform Purpura

- Purpura of these same stellate patches/plaques
- From occlusion of the perforating arterioles.

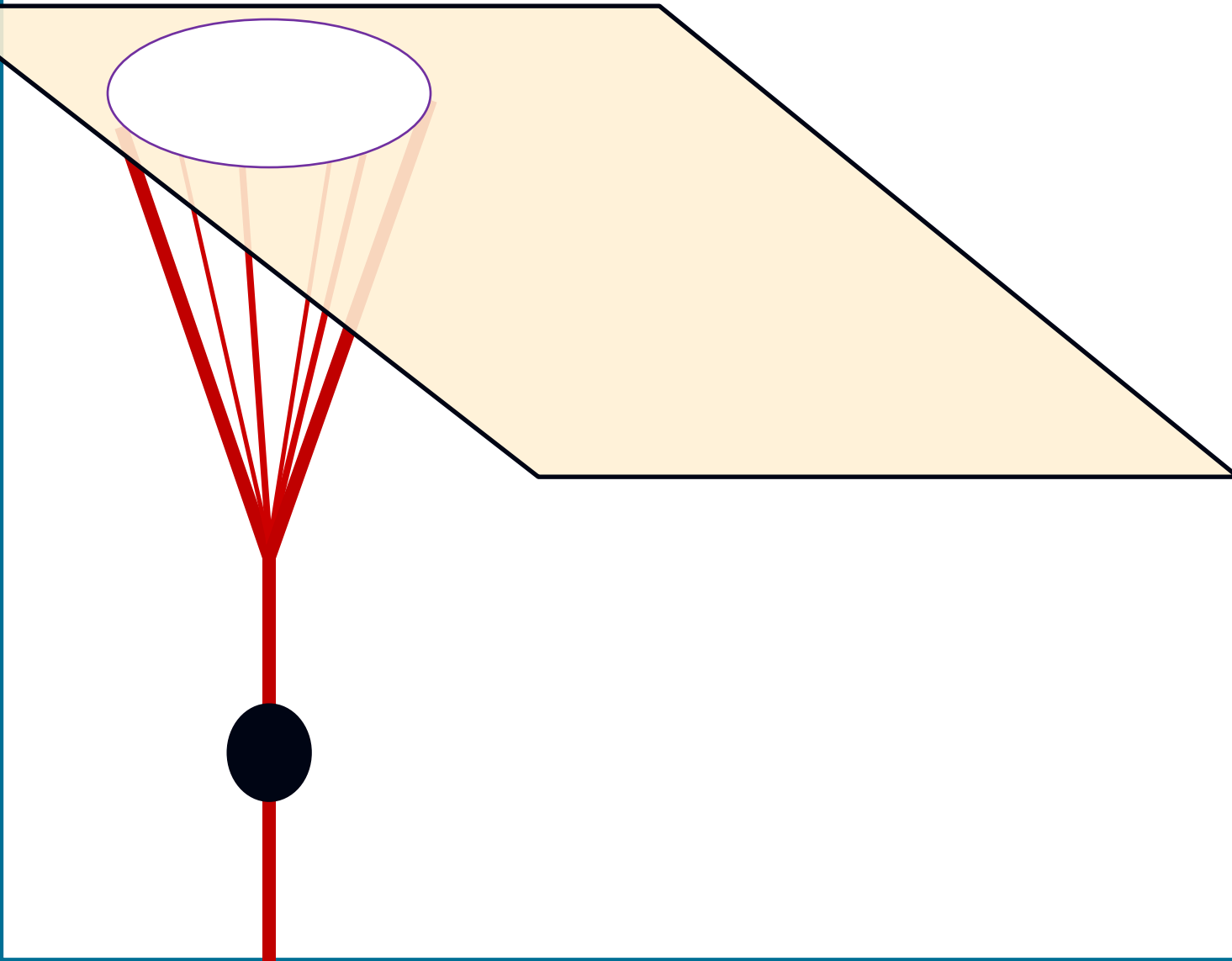
Retiform Purpura



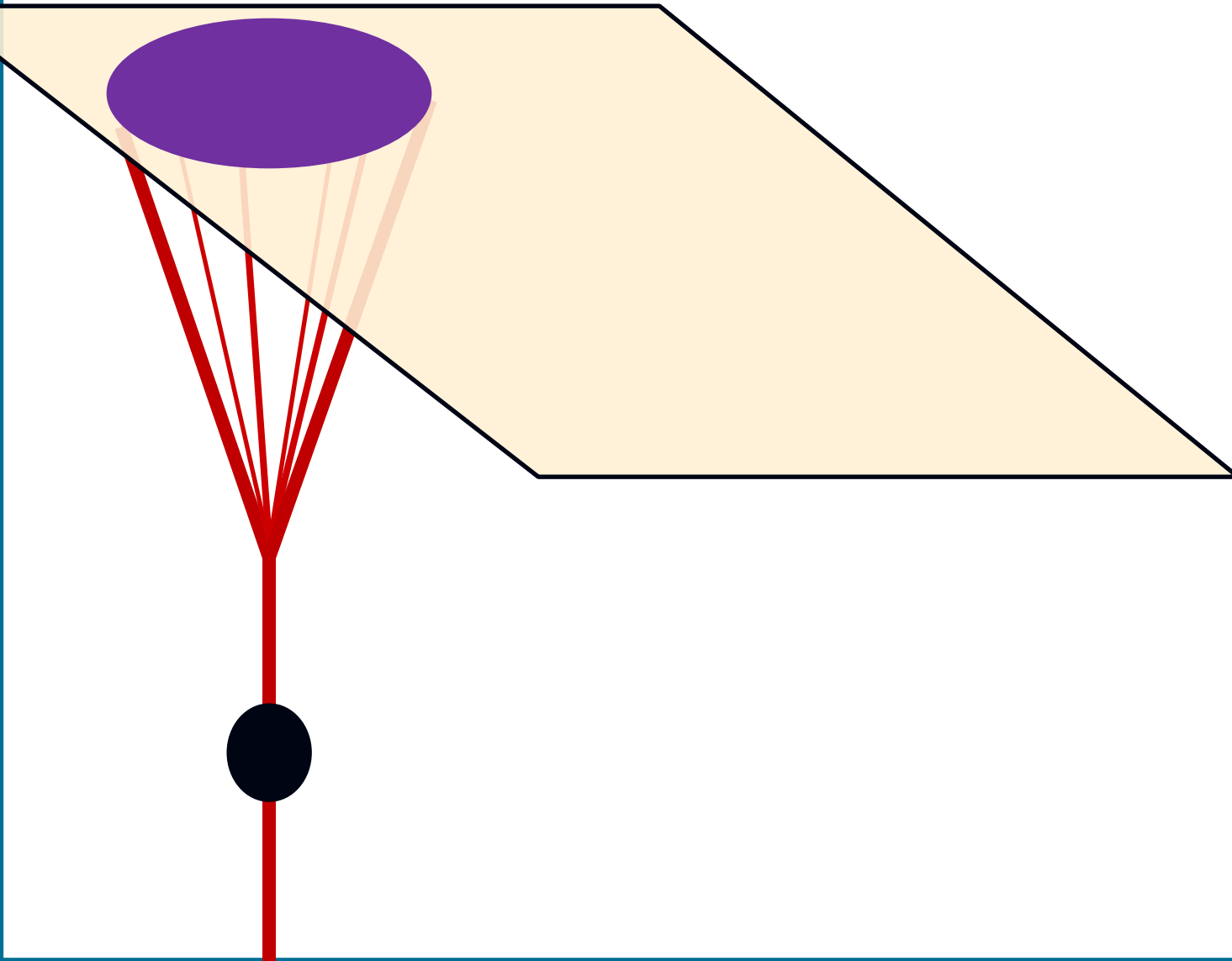
Retiform Purpura



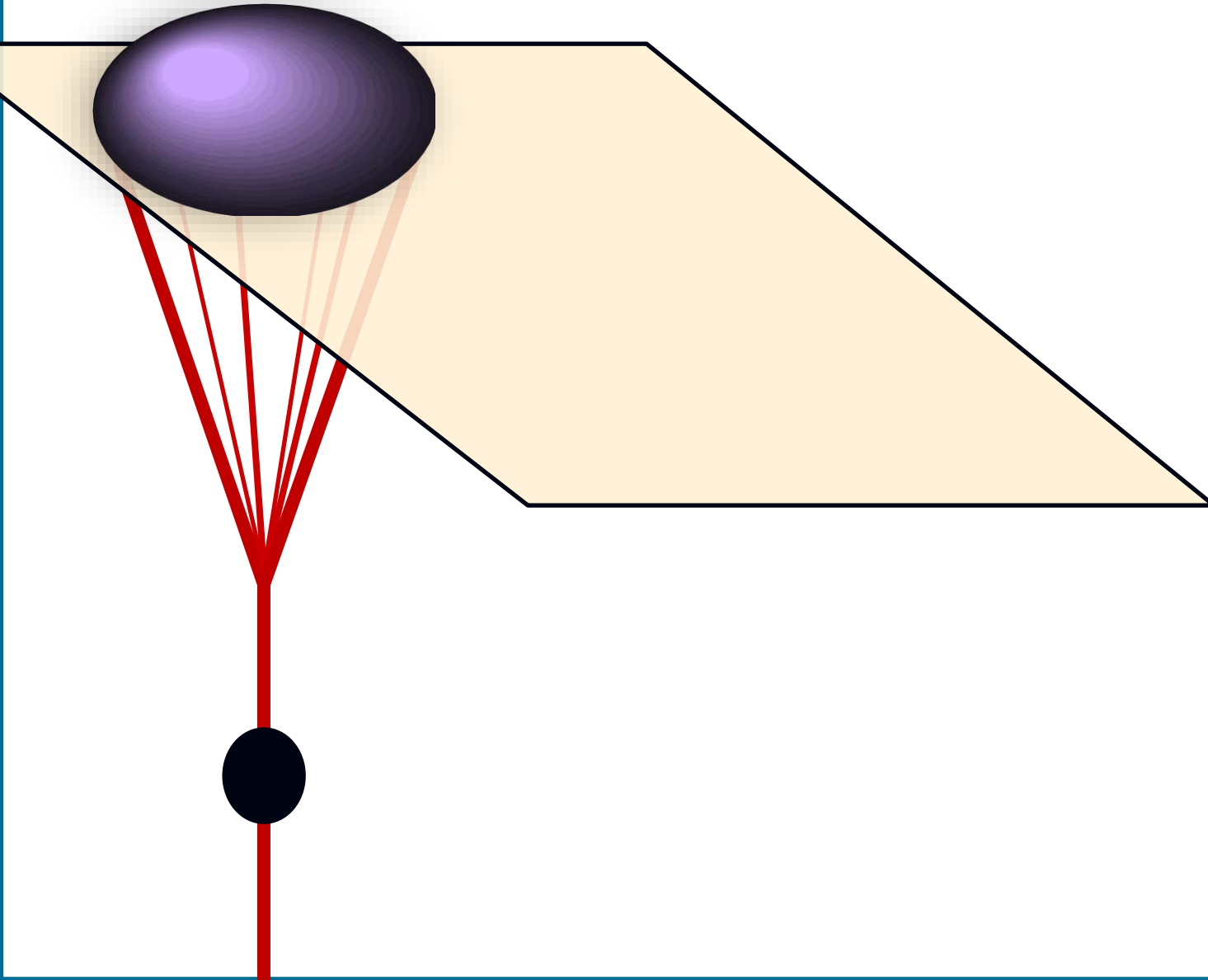
Retiform Purpura



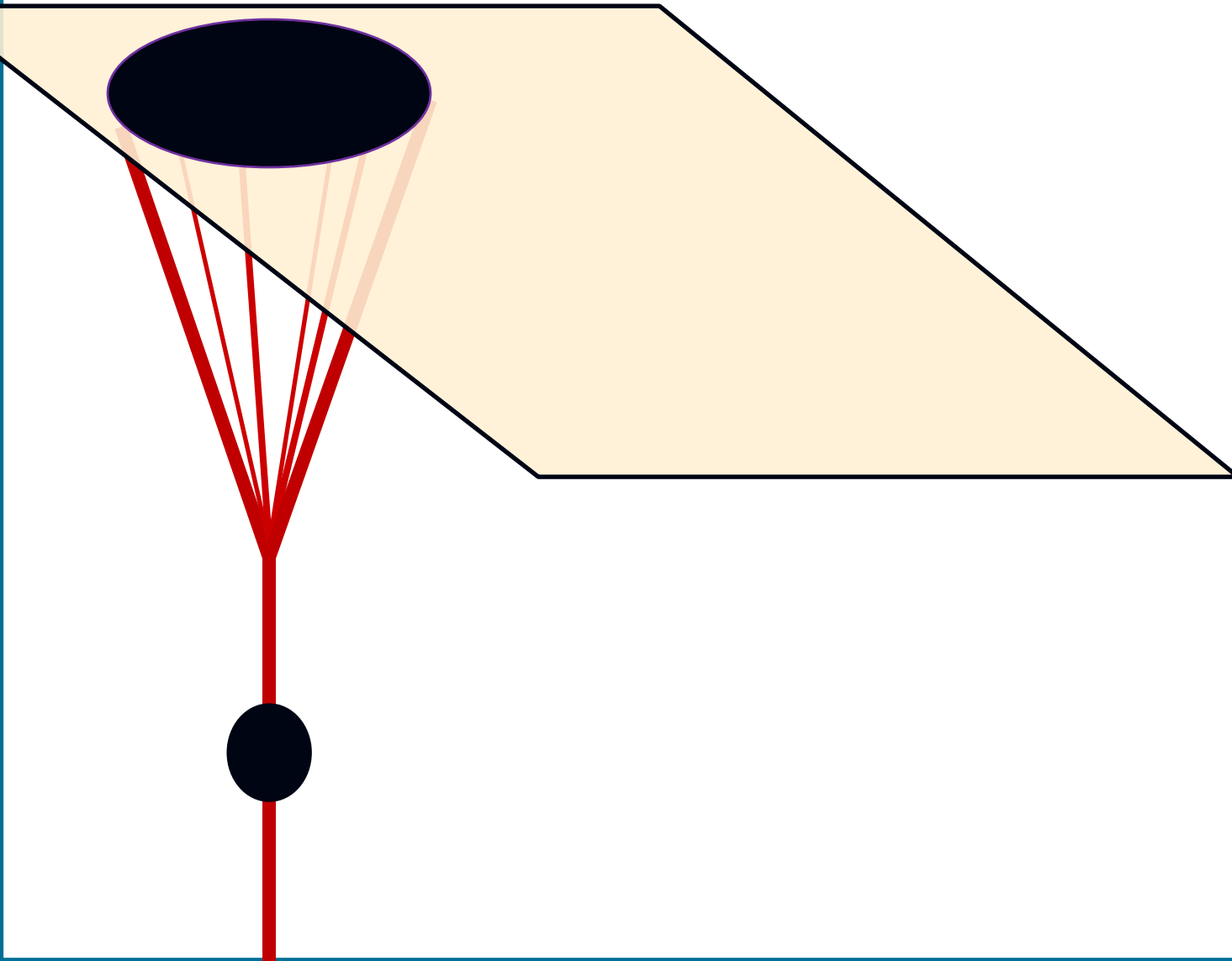
Retiform Purpura

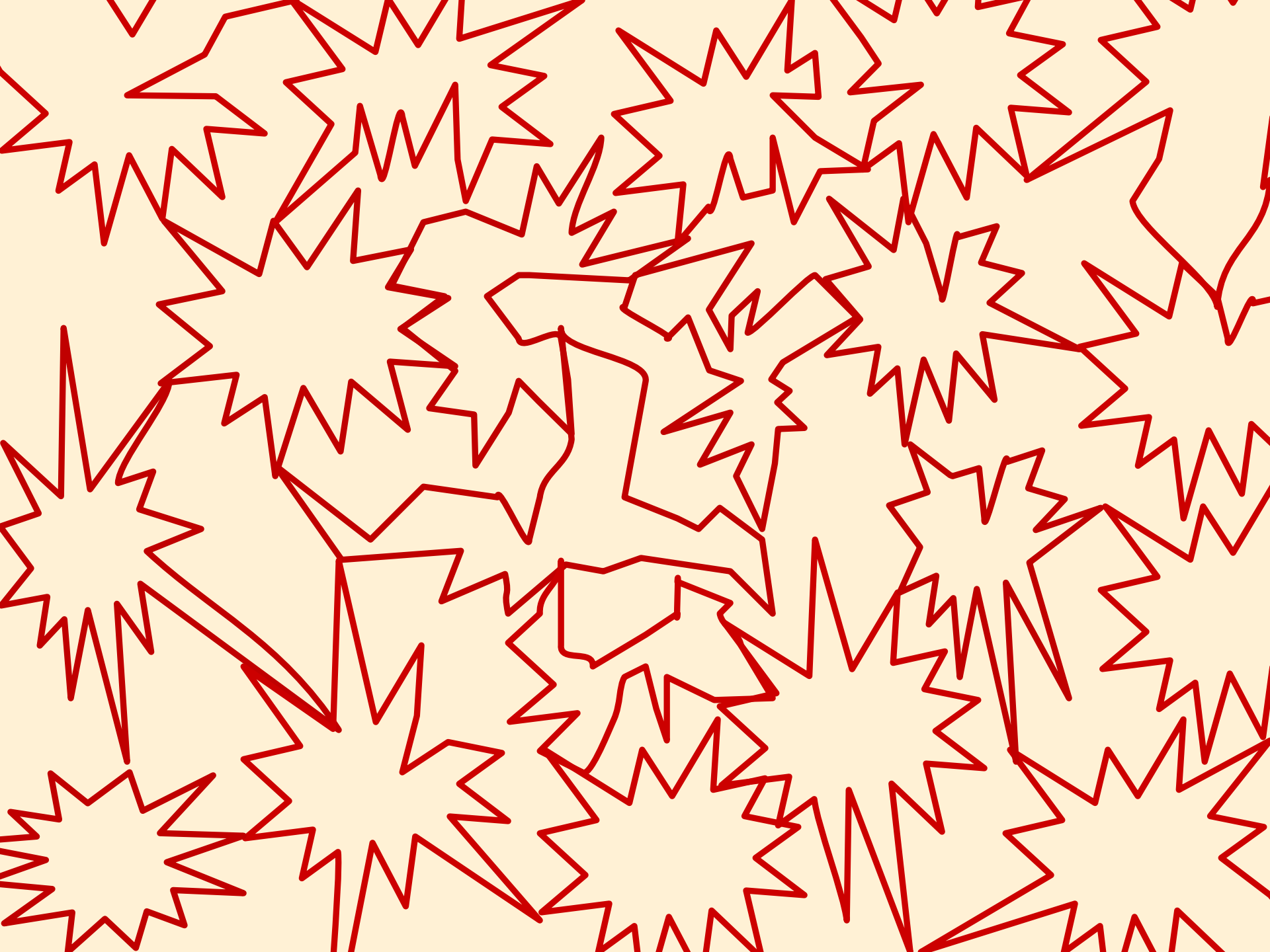


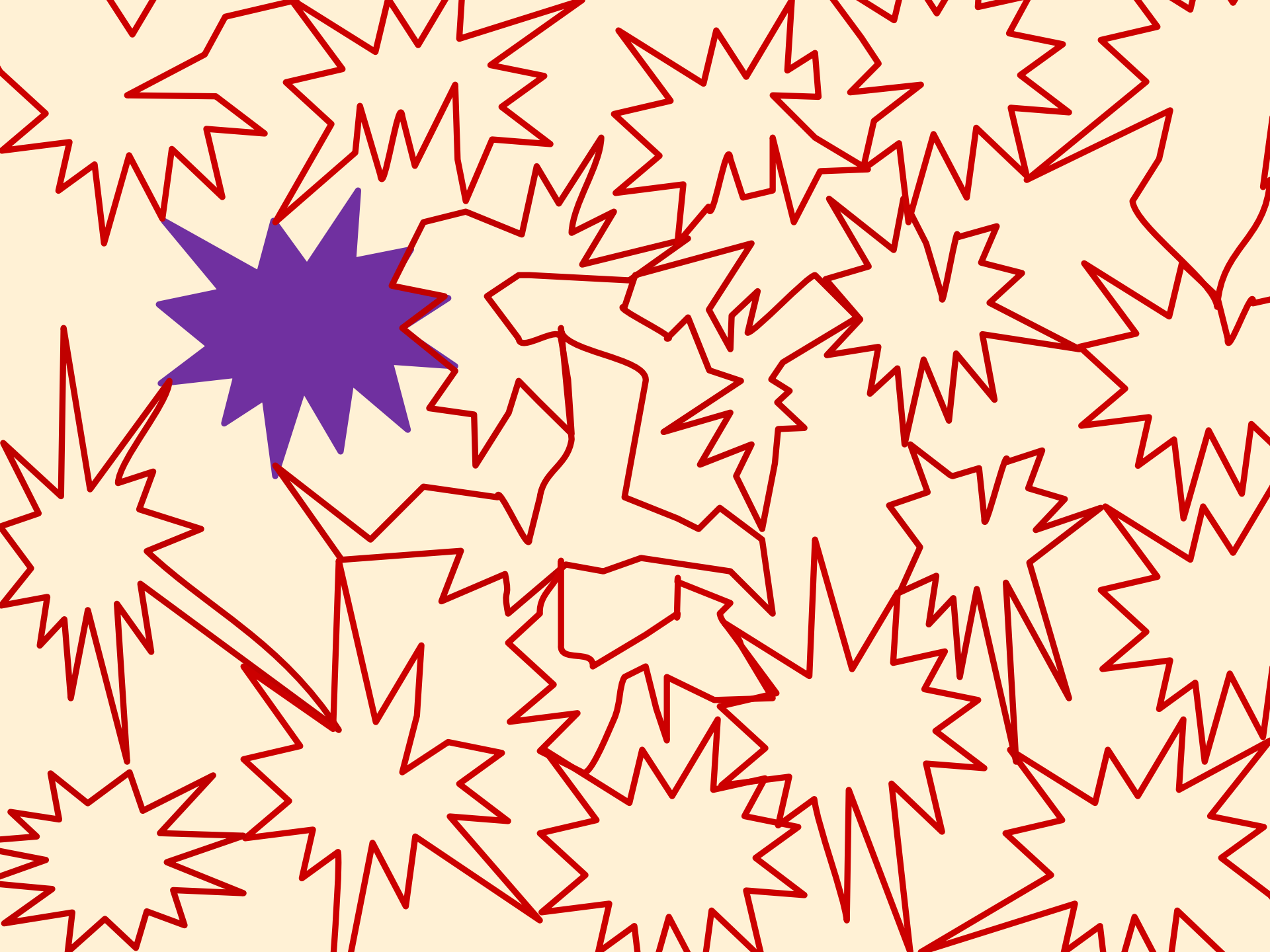
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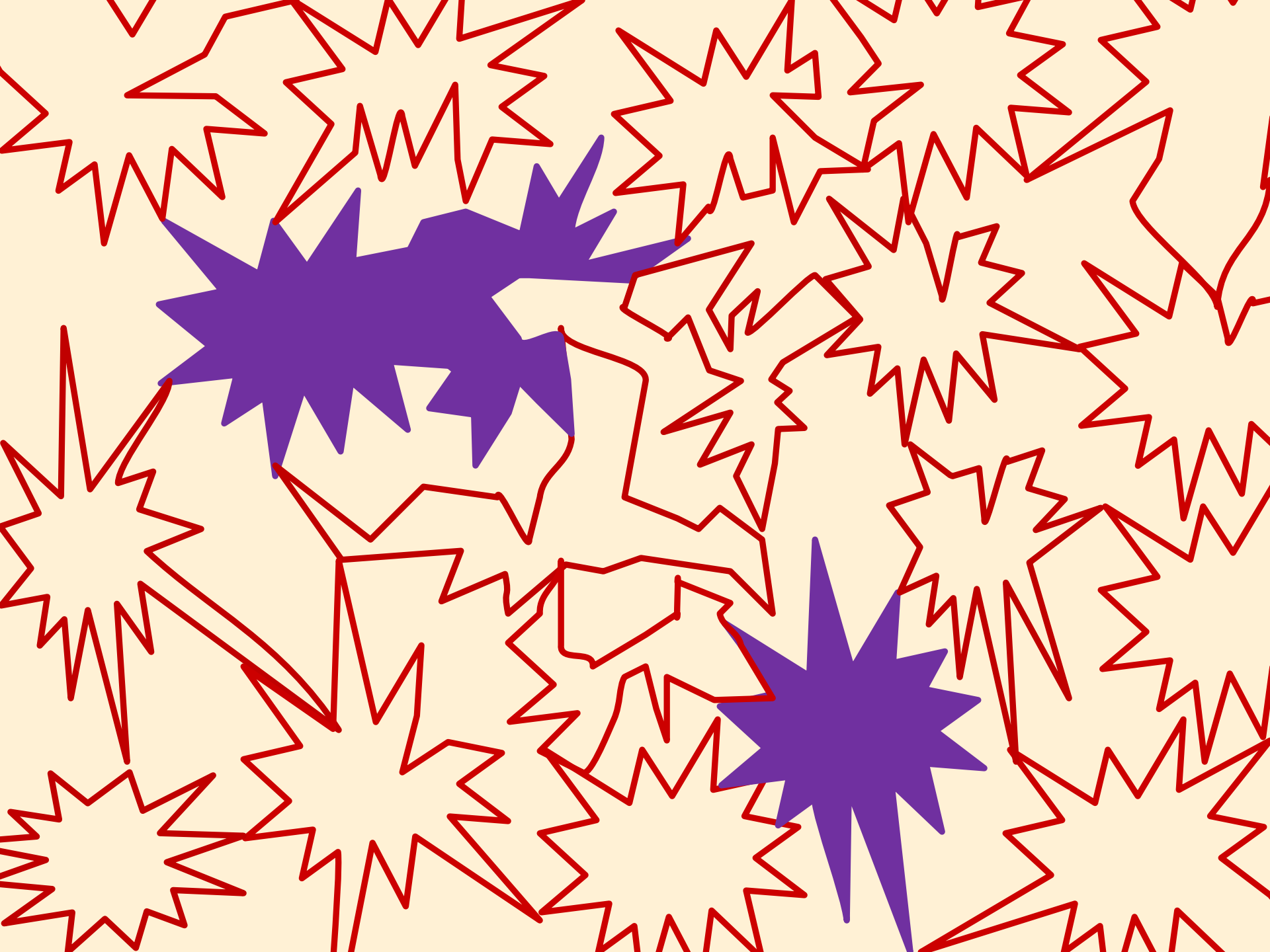


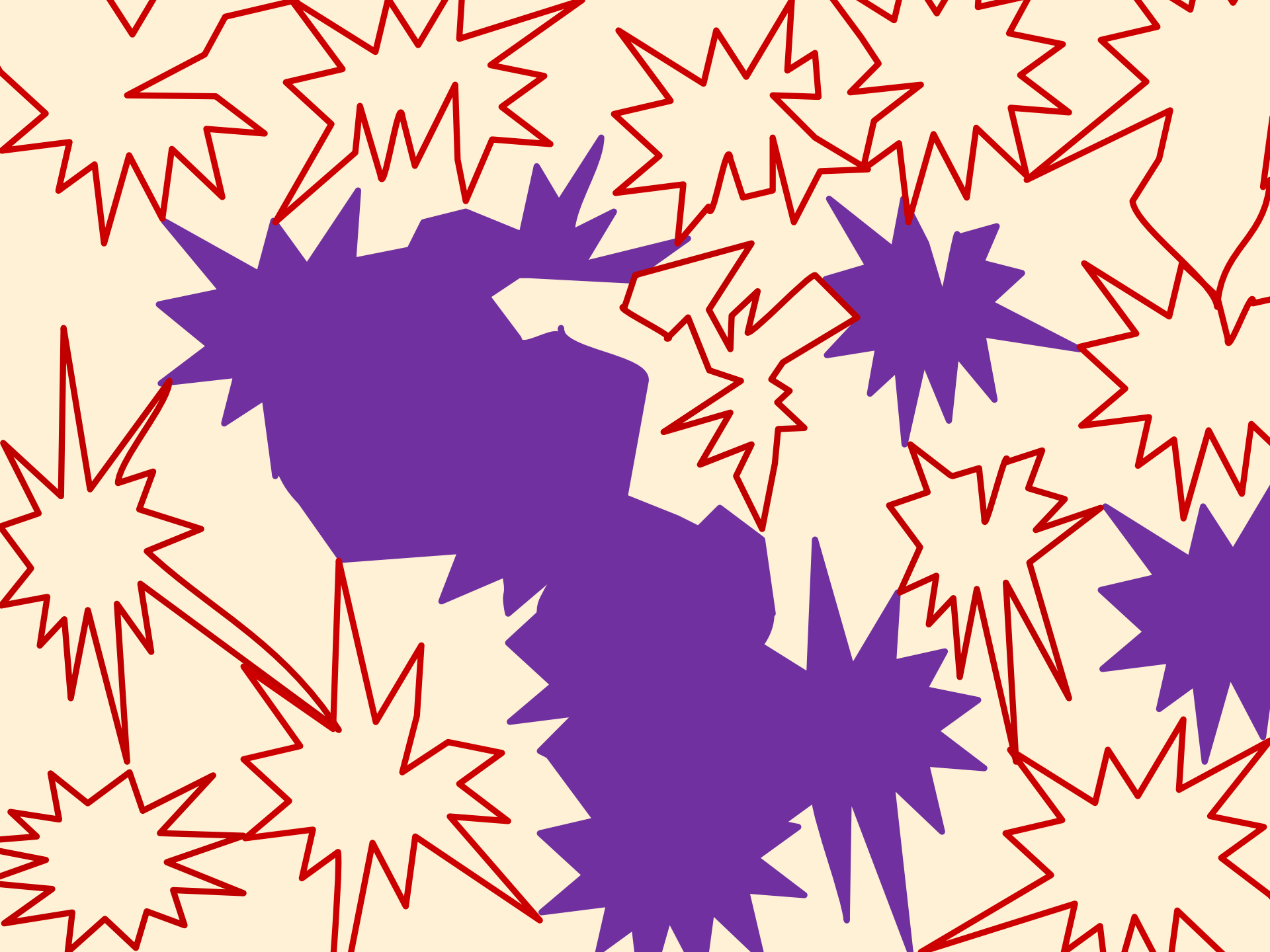
Retiform Purpura











Retiform Purpura

(with necrosis)





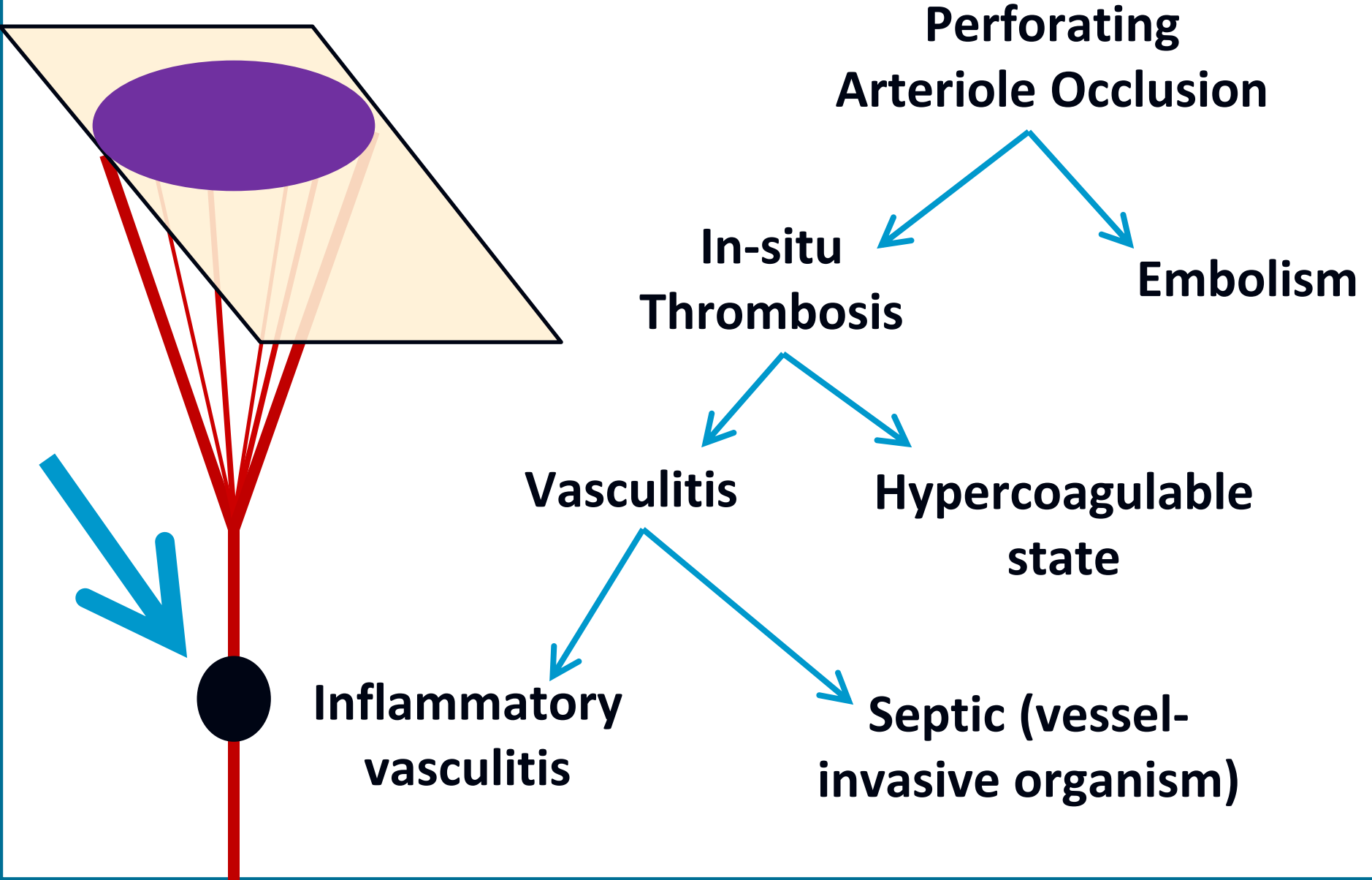




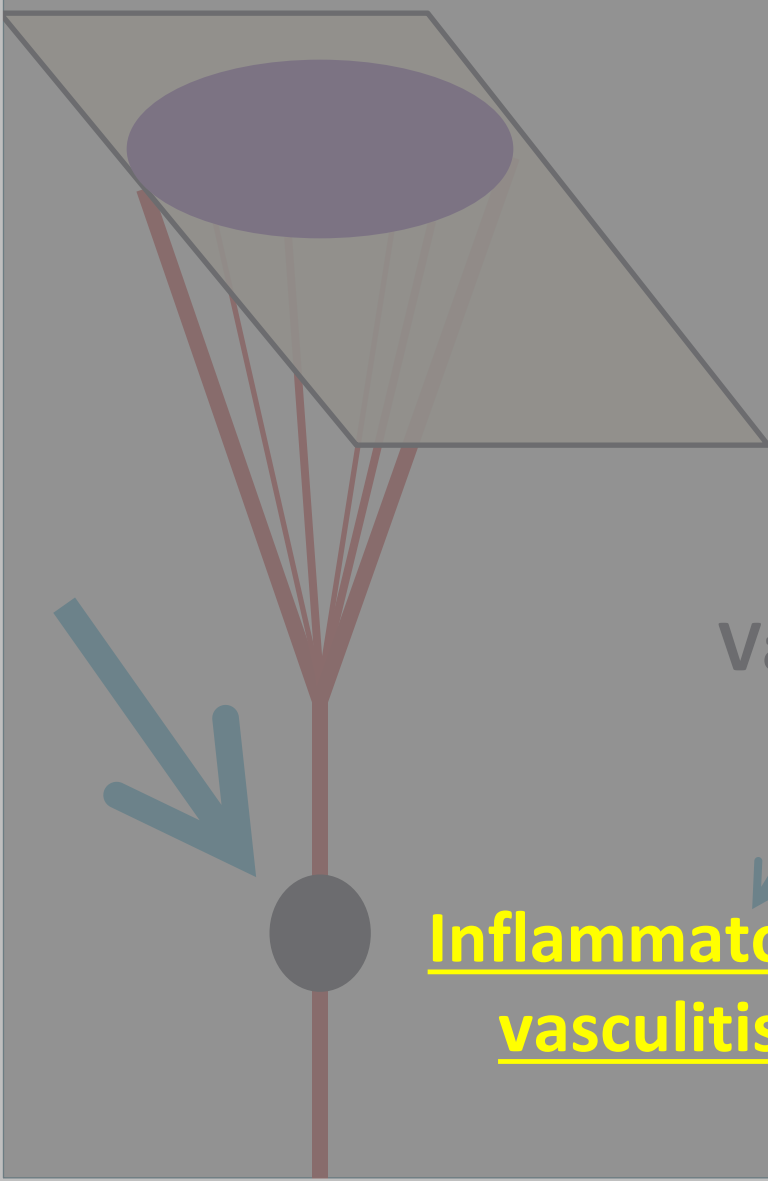
Case Details

- PMH: Systemic lupus, lupus nephritis
- Meds: Mycophenolate mofetil, prednisone
- ED presentation:
 - Vitals: **T104.6, P140s, SBPs 80s**
 - Unresponsive, rash on right leg
- Labs: BASELINES in parentheses after figures
 - **WBC 1.8** (4-9), **HCT 22.7** (24-37), **Plt 76** (150-350)
 - Na 142, K 4.3, Cl 112, HCO₃ 20, **BUN 79, Creatinine 2.7** (1.2)

Retiform Purpura: Differential Diagnosis



Retiform Purpura: Differential Diagnosis



Perforating
Arteriole Occlusion

In-situ
Thrombosis

Embolism

Vasculitis

Hypercoagulable
state

Inflammatory
vasculitis

Septic (vessel-
invasive organism)

Retiform Purpura: Select Differential Diagnosis

Emboli	Cholesterol, Fat, Septic, Calciphylaxis, Amyloidosis, Nitrogen, Atrial myxoma, Ventilator Gas, Hyperoxaluria
Hypercoagulable states	APLAS, Sneddens, Cryos, AT III deficiency, Protein C/S def (especially with meningococemia or coumadin), DVT, DIC, TTP
Inflammatory Vasculitis	PAN, Wegeners, Takayasu's, microscopic polyangitis, Rheumatoid vasculitis, livedoid vasculitis
Septic vasculitis (Angioinvasive pathogens)	Pseudomonas, Serratia, Aeromonas, Klebsiella, Vibrio, Moraxella, Morganella, E.coli, Staph aureus, Candida, Mucor, Aspergillus, Fusarium

Adapted from:

Gibbs MB, English, JC, Zirwas MJ. Livedo Reticularis: An Update. J Am Acad Dermatol 2005; 52: 1009-19

Please note:

(regarding retiform purpura)

- **Nothing on the differential is primary cutaneous**
- **Everything on the differential is bad**

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Catastrophic APLAS ("thrombotic storm")

Thrombotic thrombocytopenic purpura

Systemic infection (Sepsis/DIC, emboli, vascular invasion)

Differential:

Dermatologic Workup and Results

- Day 0:
 - Biopsies by derm and surgery
 - Later that night: Blood cultures stain for **GNR in 4/4 bottles**
- Day 1 post admission: Pathology preliminary results—
 - Neutrophilic inflammation in dermis and adipose with hemorrhage.
 - Deep biopsy has sparse GNR on Gram stain
- Day 2: blood and deep biopsy tissue—
 - ***Serratia marcescens***
- Day 3: Abd CT with contrast shows pan-enterocolitis

Diagnosis

Serratia marcescens sepsis with necrotic
retiform purpura of a seeded limb

More faces of Retiform Purpura

















10/1/14
10/1/14



CASE KEY POINTS

- **Recognize Retiform Purpura:**
 - Well demarcated purpuric patches with jagged edges
 - Violaceous, dusky, white, black
 - Evidence of necrosis (bullae, ulcers, eschars)
- **Early indicator of a systemic, generally malignant process**