

SKIN LESIONS AS A CLUE TO DIAGNOSIS OF SYSTEMIC DISEASE

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MINISTRY OF EDUCATION,
YOUTH AND SPORTS



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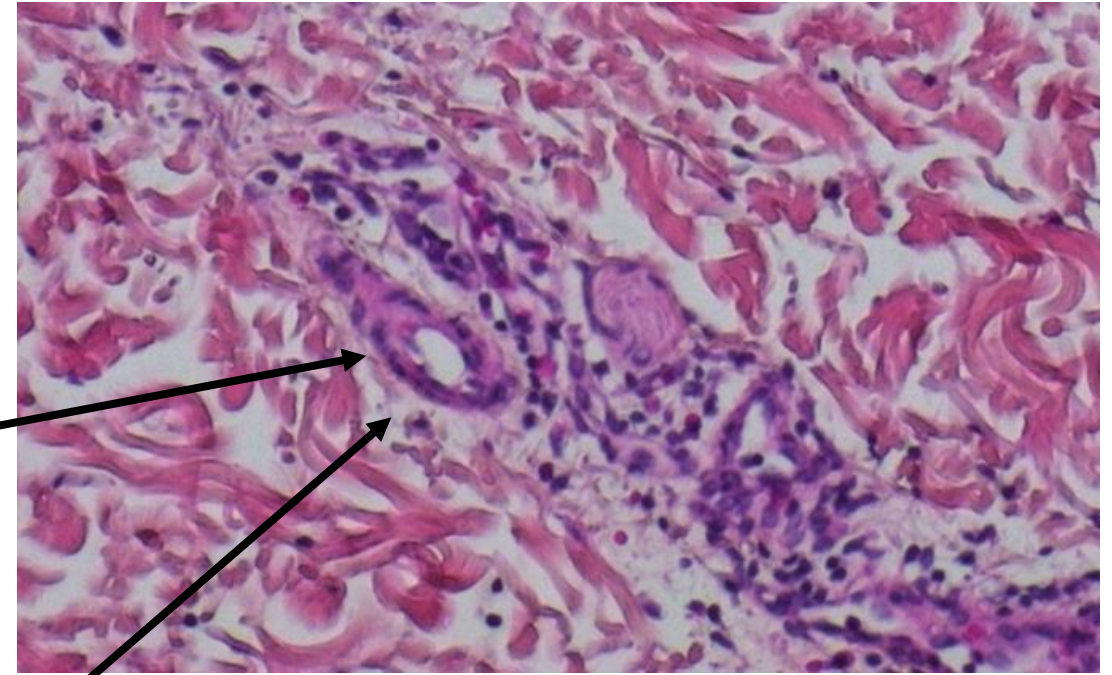
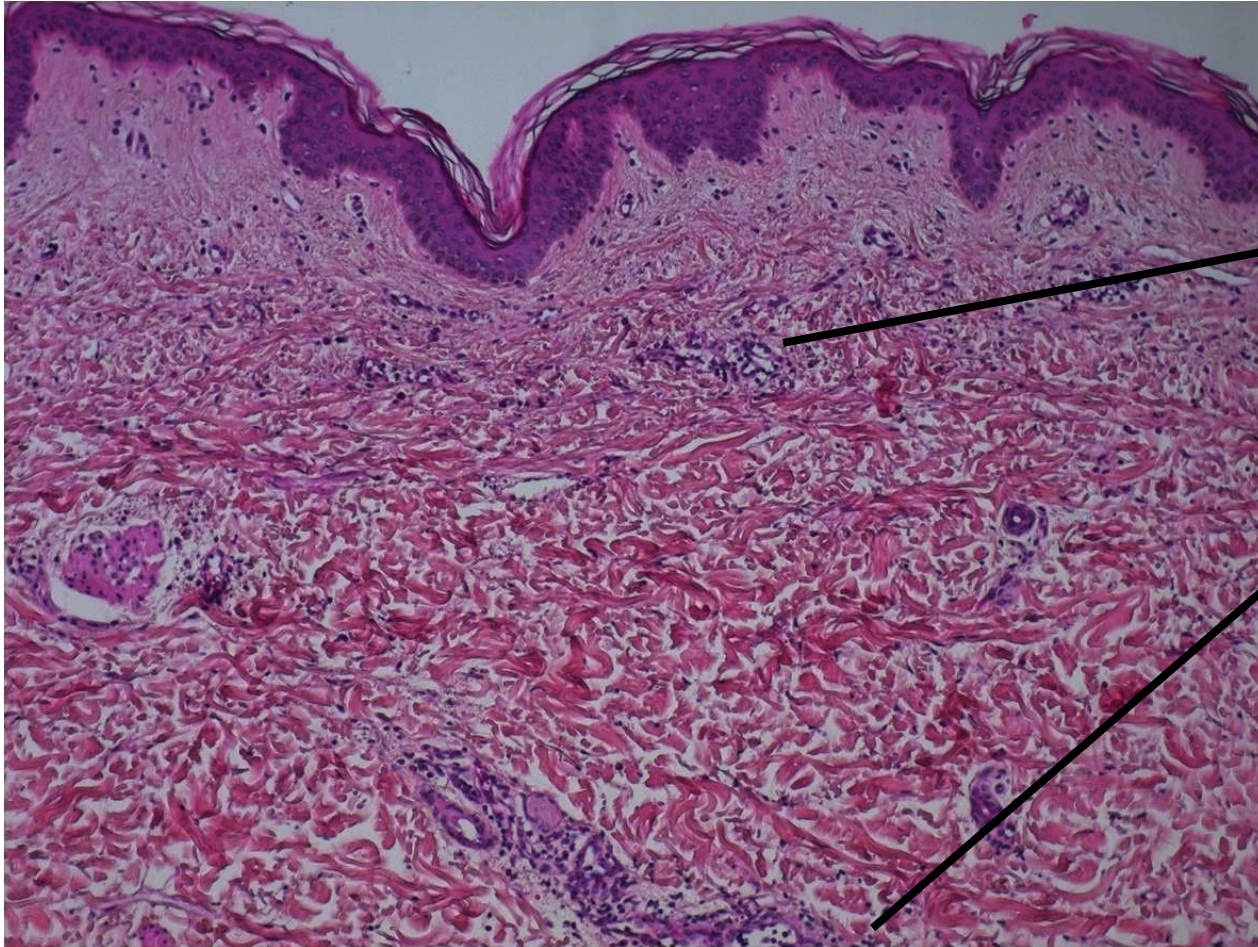
URTICARIA

URTICA – POMPHUS

- 1 lesion lasts less than 24 hours
- Usually is itchy
- Might be associated with
 - angioedema (10% in children)
(pale, painful, lasts 2-3d)
 - other symptoms
(fatigue, oedema and joint or abdominal pain)



HISTOLOGY



Dermal oedema
Perivascular and interstitial infiltrate of
lymphocytes, eosinophils, neutrophils
Minimal epidermal changes

ETIOLOGY – 50% no cause identified

- ALLERGIC – I., II. a III. type
- PSEUDOALLERGIC* - nonIgE, C
(drugs, additives, chinese medicine)
- PHISYCAL – cold, heat, pressure
vibration, UV, water.....
- AUTOIMUNNE *
IgG anti FcεRI/IgE
- IDIOPATIC



LASTS MORE THAN 6weeks

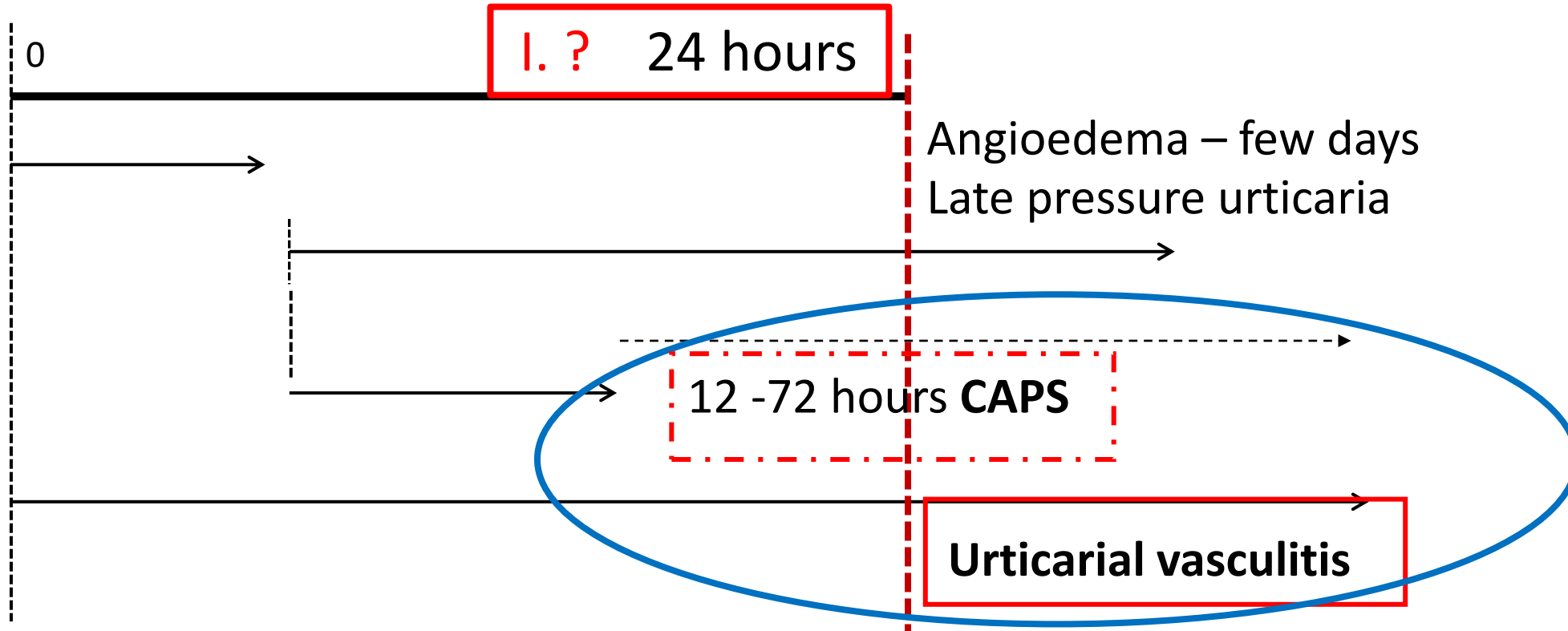
- ACUTE
- CHRONIC min 2xw
0,1 - 3% in children
older or adolescents, ♀ > ♂

WHAT IF IS IT NOT „JUST“
AN URTICARIA?

IMPORTANT CLUES

I. HOW LONG DOES SINGLE LESION LAST

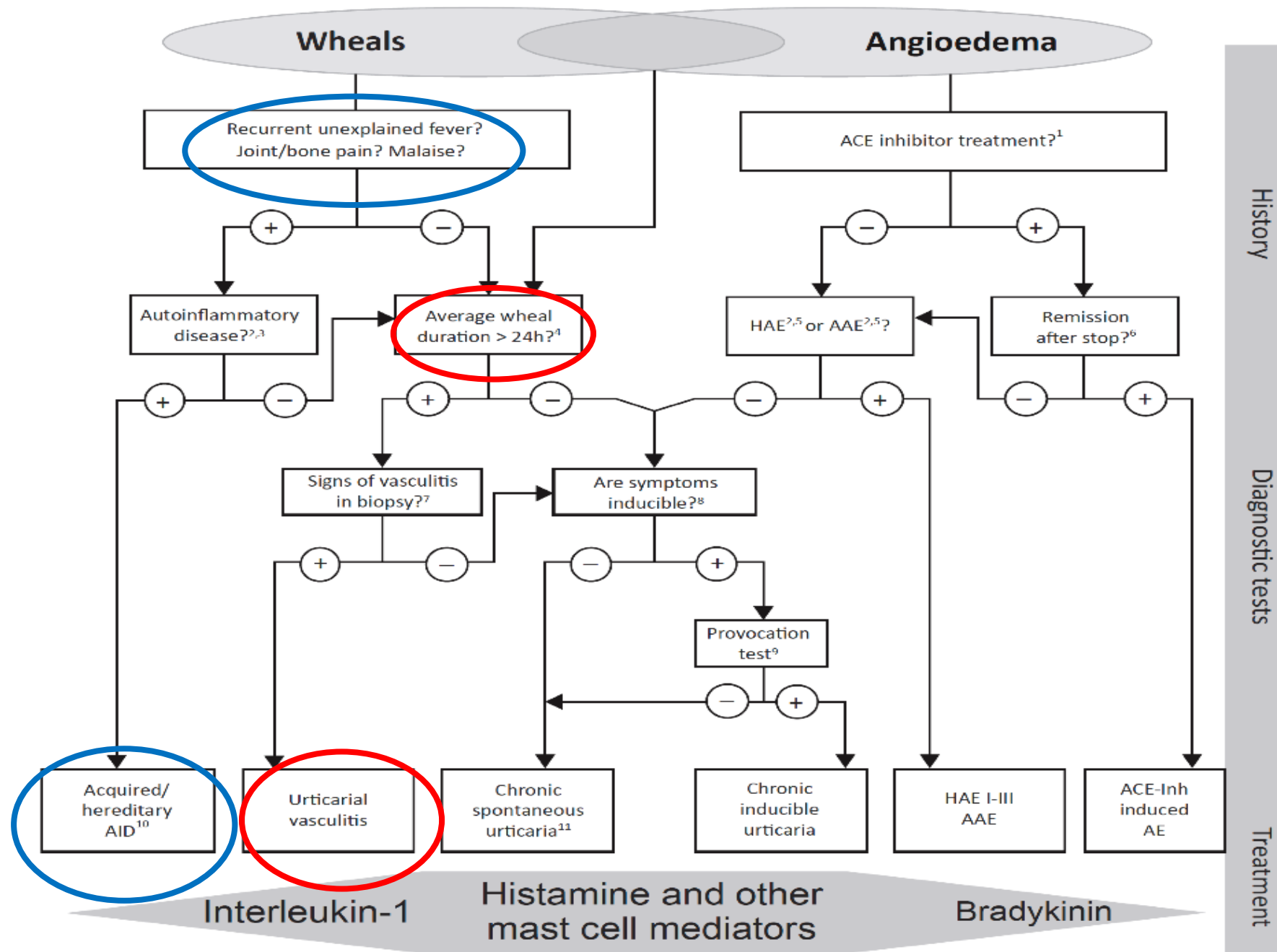
II. PRESENCE OF SYSTEMIC SYMPTOMS



II. ? Systemic symptoms- fever, arthritis

CONNECTIVE TISSUE DISORDERS
MONOCLONAL GAMAPATHY
LEUKEMIA, TUMORS
VIRUS (HBV, CV, EBV)
DRUGS (ACE, PNC, SULPHONAMIDES)

CAPS
HIDS
TRAPS
SJIA
AOSD
Schnitzler syndrome



- residual changes after 24h: hyperpigmentation
purpura
- not itchy, burning
- bad reaction to
antihistamines



HISTOLOGY

Graduate changes

Neutrophilic infiltrate

Leucocytoklasia

Fibrinoid necrosis
Endothelial oedema

URTICARIA

Mastocytes
Eosinophils



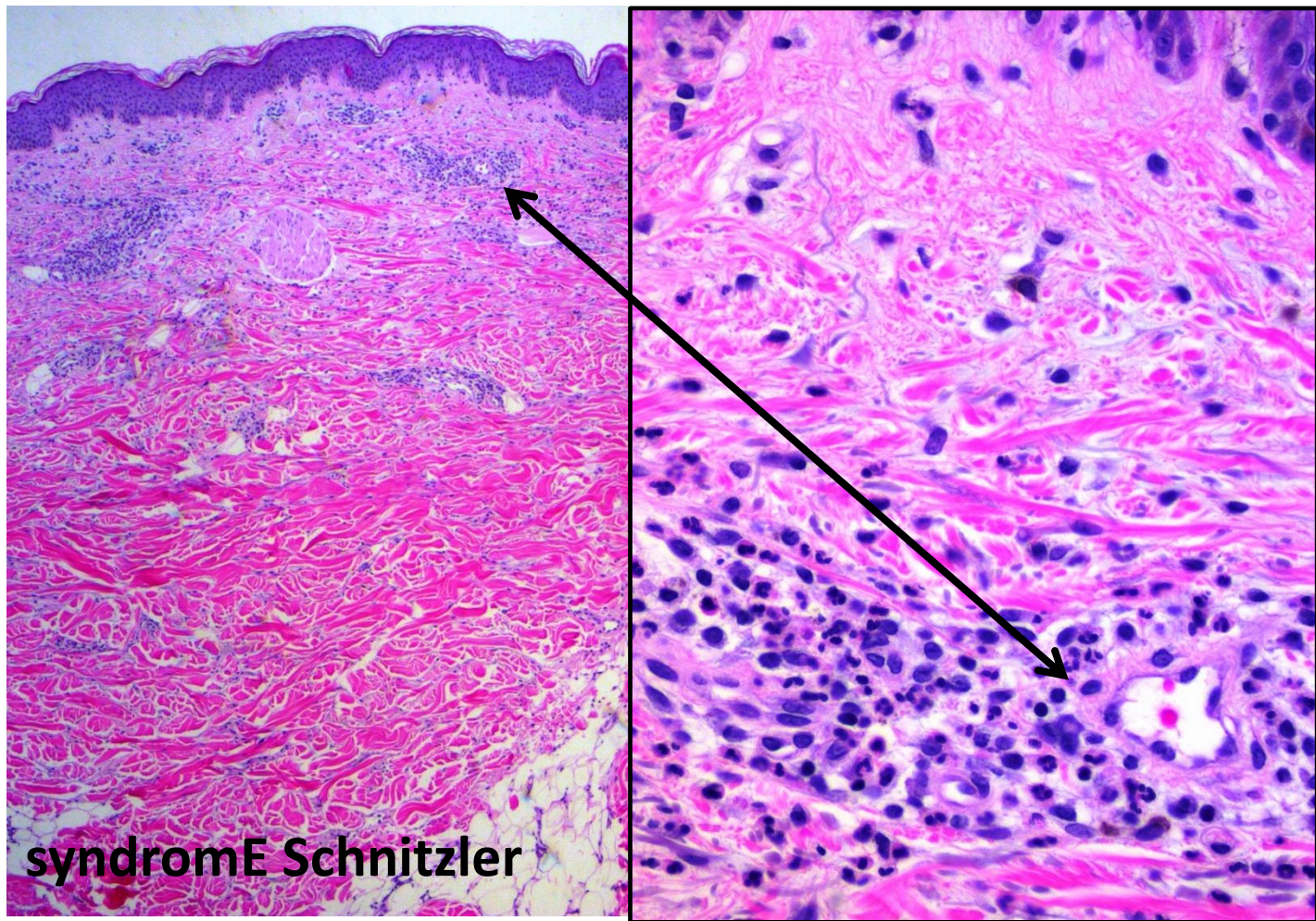
IL-1
Neutrophils

VASCULITIS

IK
Complement
Neutrophils

NEUTROFILIC URTICARIA

...neutrophilic urticaria with systemic inflammation

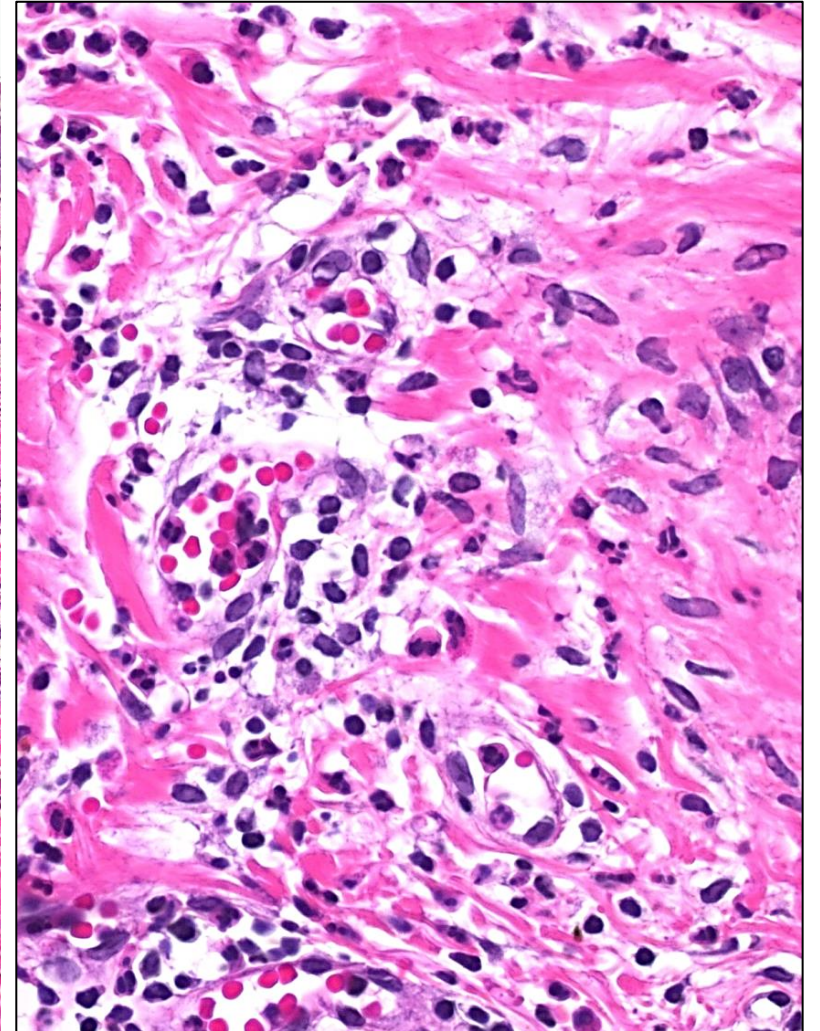
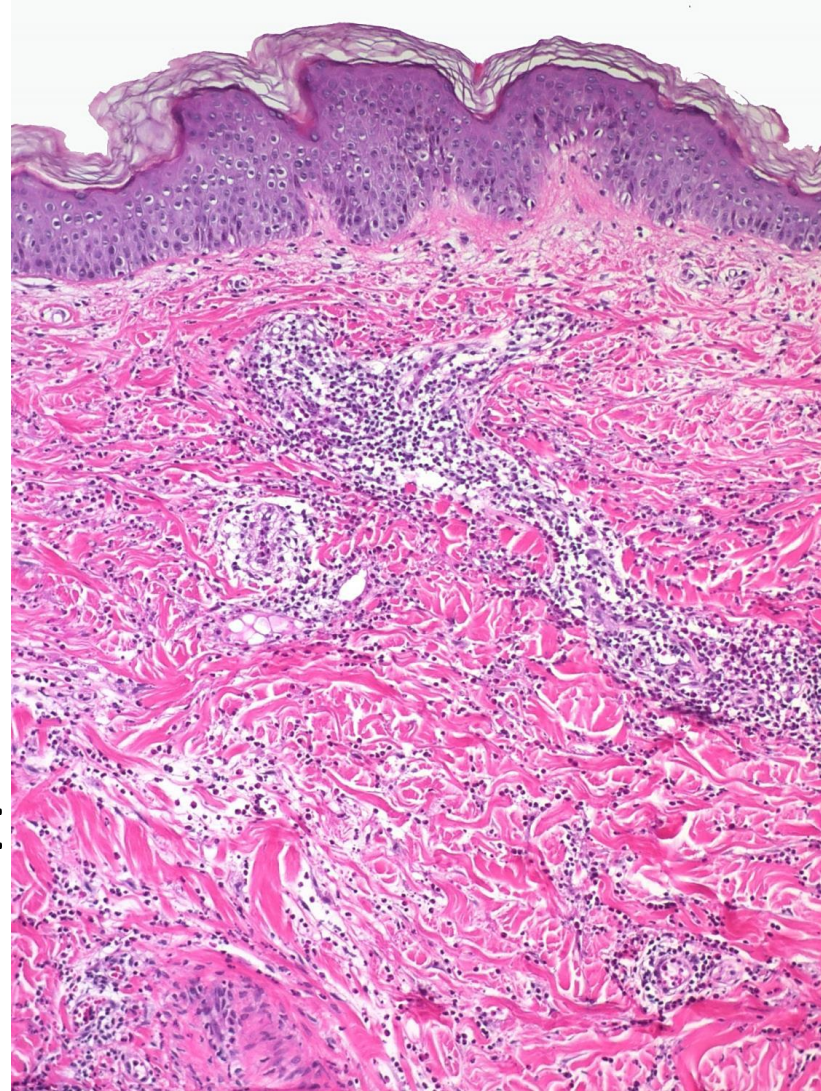


Normal capillary wall without vasculites changes
perivascular mononuclear infiltrates with enutrophils and mild
oedema of superficial corim – sign of clinical wheal

Serum sickness like reaction



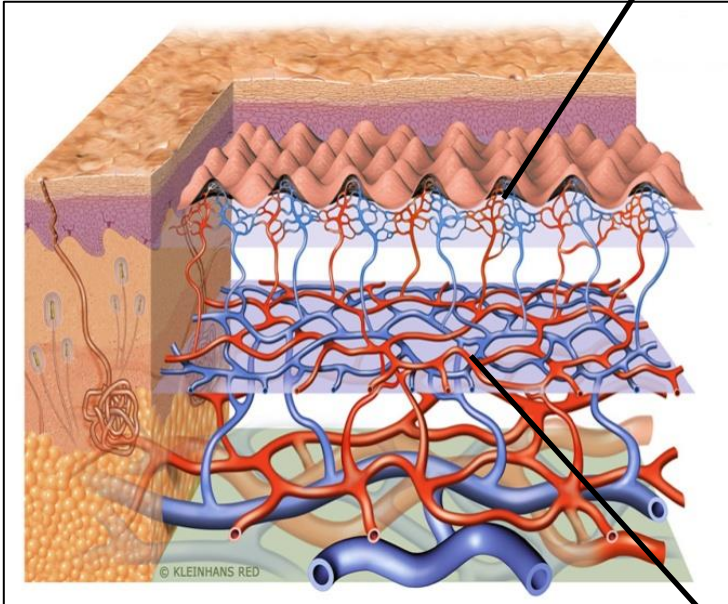
URTICARIA VASCULITIS - 2-10%



COLLAGEN VASCULAR DISEASE
MONOKLONAL GAMAPATHY
LEUKEMIA, TUMORS
VIRUSES
DRUGS
IDIOPATIC

erythrocyte extravasates, neutrophils, leukocytoclasia
+ clinical picture

VASCULITIS



SMALL VESSEL VASCULITIS
(mostly *IK+C+Neu*)

IgA vasculitis (HSP)
Urticaria-vasculitis

MIDDLE VESSEL VASCULITIS

Polyarteriitis nodosa
Kawasaki disease

.....

PALPABLE PURPURA

MACULES, PAPULES, URTICA
VESICLES, PUSTULES

MIXED (SMALL+ MIDDLE) VESSEL VASCULITIS

(s ANCA)

MPA (Microscopic polyangiitis)

GPA (Granulomatosis with polyangiitis)

.....

**NODULES, NECROSIS,
ULCERATION,**

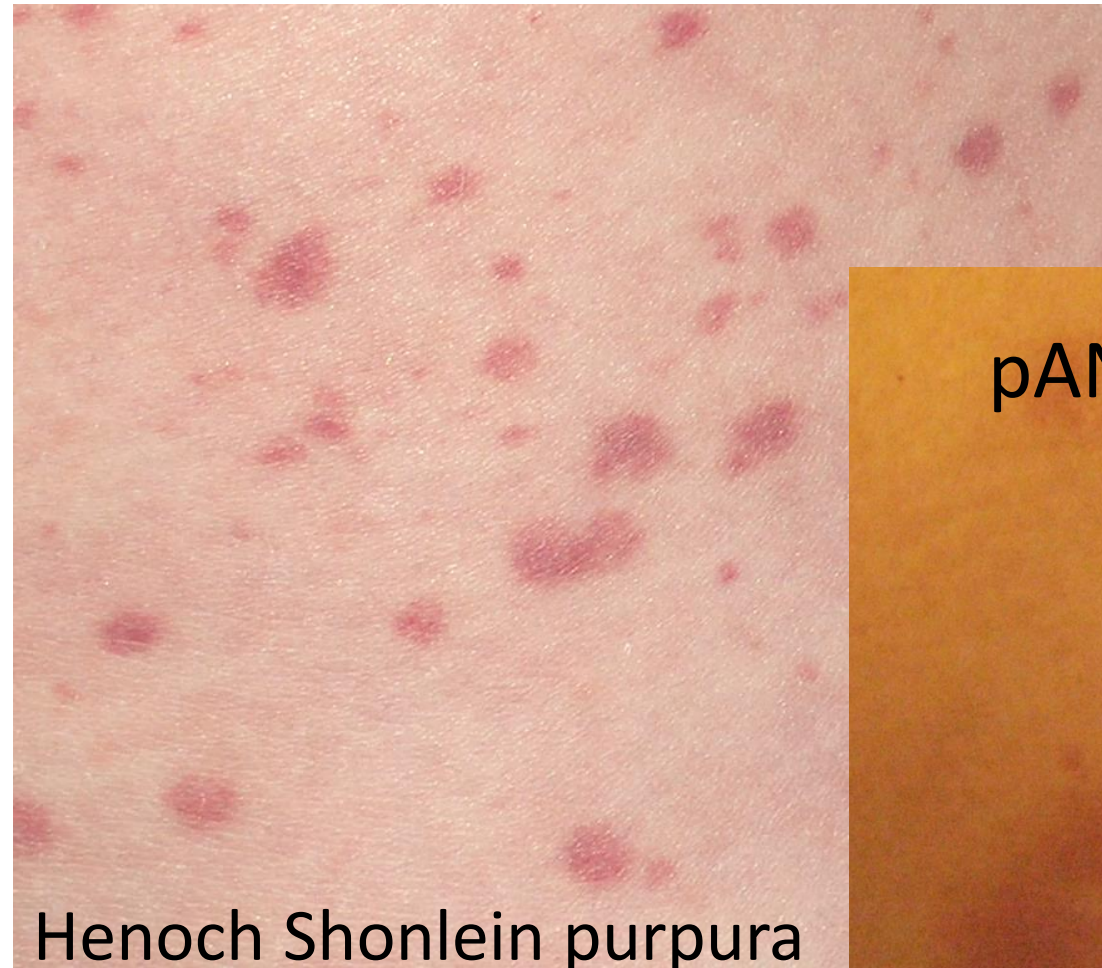
LIVEDO RACEMOSA

pseudo-SWEET SYNDROME

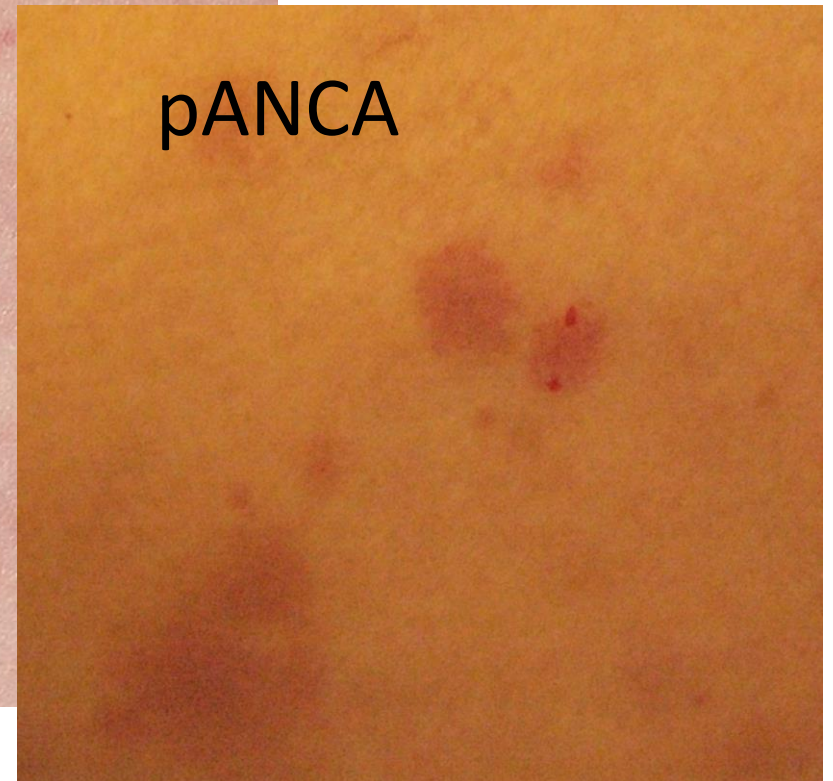
pseudo-PYODERMA

GANGRENOSUM

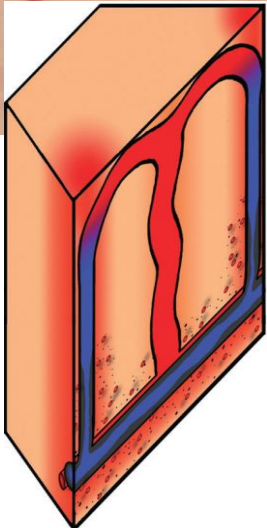
SMALL VESSEL VASCULITIS



Henoch Schonlein purpura



pANCA



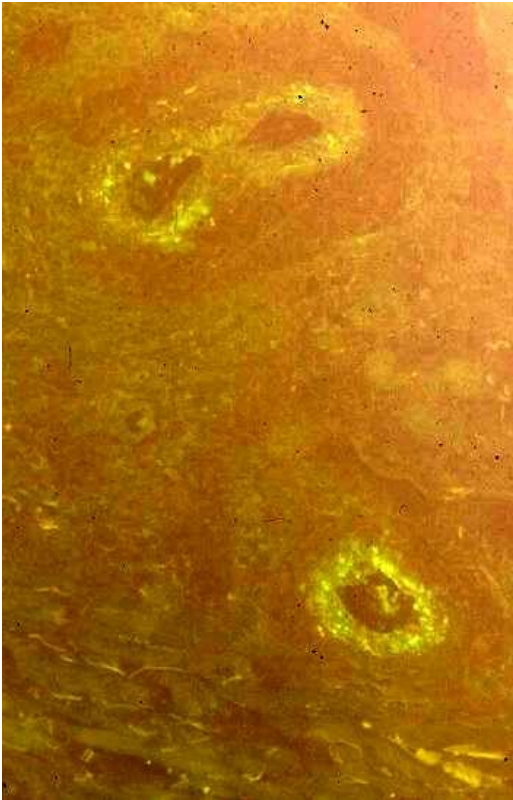
DIRECT IMMUNOFLUORESCENCE HELPFULL IN DIAGNOSIS

Biopsy of LESIONAL skin: lesion should NOT BE OLDER THAN 24 HOURS

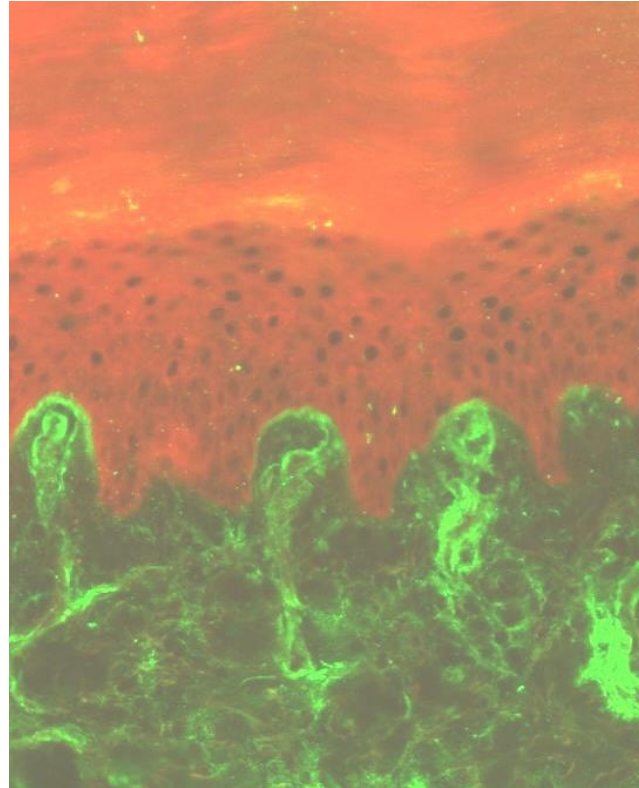
Type of fluorescence: complement C3, Ig M,G, A, fibrinogen

IgA: 90% children / 10% adults

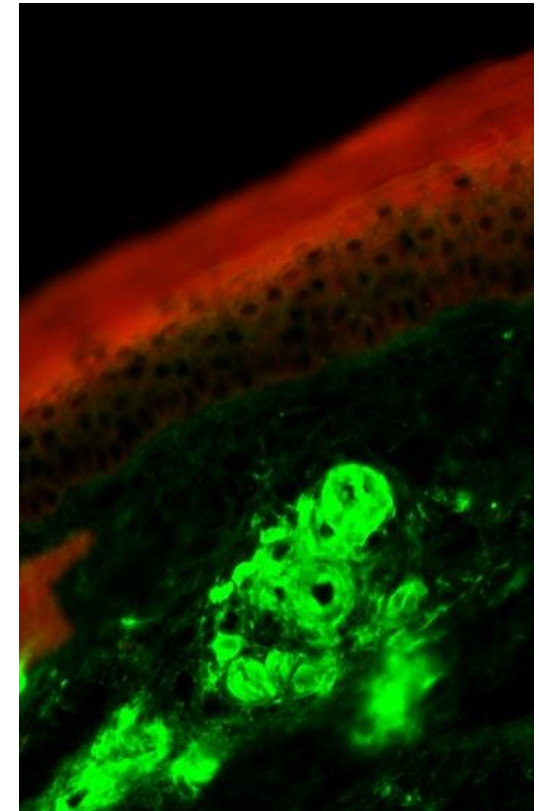
IgG/IgM: 80% adults



C3



Ig A



fibrinogen

CLINICAL CHARACTERISTICS

min 1 systemic sign in 80% of patients

lower limb localization in 93%

IgA: lesions on buttocks

abdominal pain

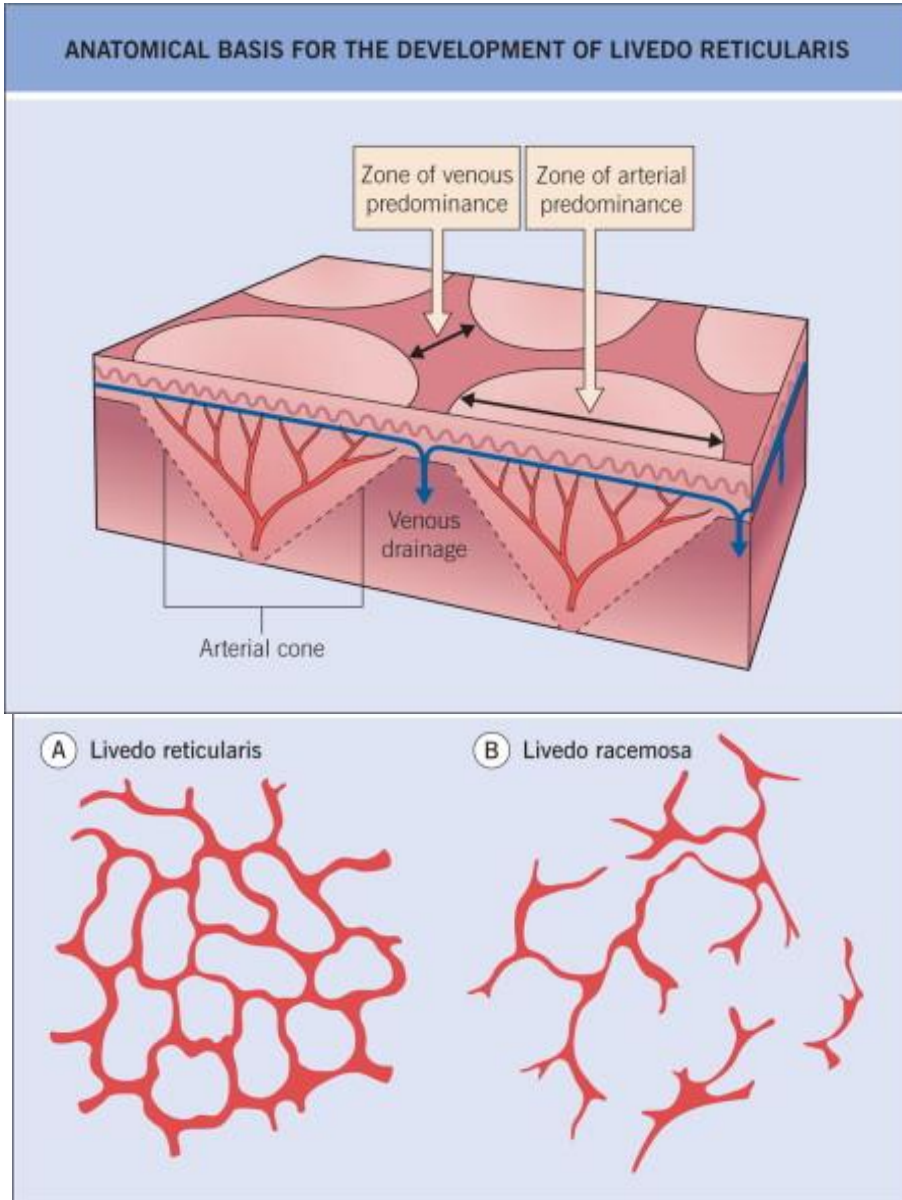
X non-IgA - headache

COURSE: a) one episode spontaneously regressing (60%)

b) recurrent (20%)

c) chronic (20%)

MIDDLE VESSEL VASCULITIS



↑deoxyg Hgb, arteriolospasm
(cold, autonomic dysfunction
– common in neonates)

FUNCTIONAL CHANGE

endotelial dysfunction
hypercoagulation
(vasculitis, collagen vascular
disease, tumors,)



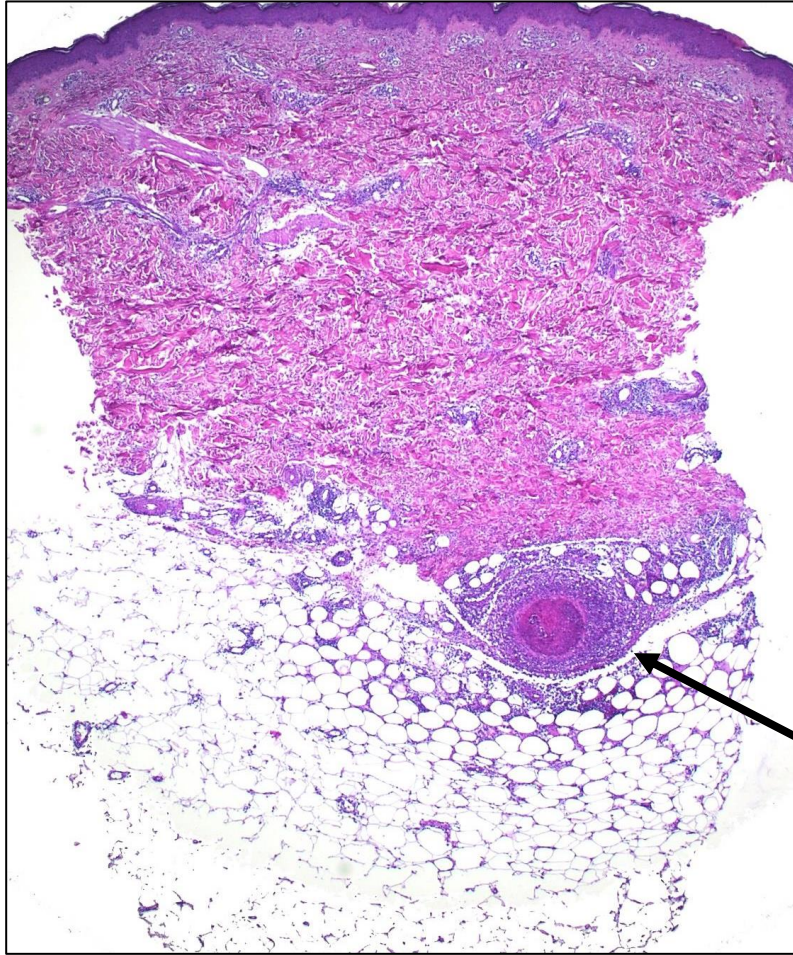
LIVEDO RETICULARIS



LIVEDO RACEMOSA



POLYARTERITIS NODOSA



References

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- <http://urticariaday.org/urticaria-day-2019/urticaria/the-new-urticaria-guideline/>
- Fabbri P. et al. Classification and clinical diagnosis of cutaneous vasculitides. G ITAL DERMATOL VENEREOL 2015;150:169-81
- Sunderkötter C. et al. Eur J Derm 2006, 16:114-24, Carlson JA Histopathol 2010 56:3-23