Comment reconnaître une vascularite

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Conflits d’intérêts

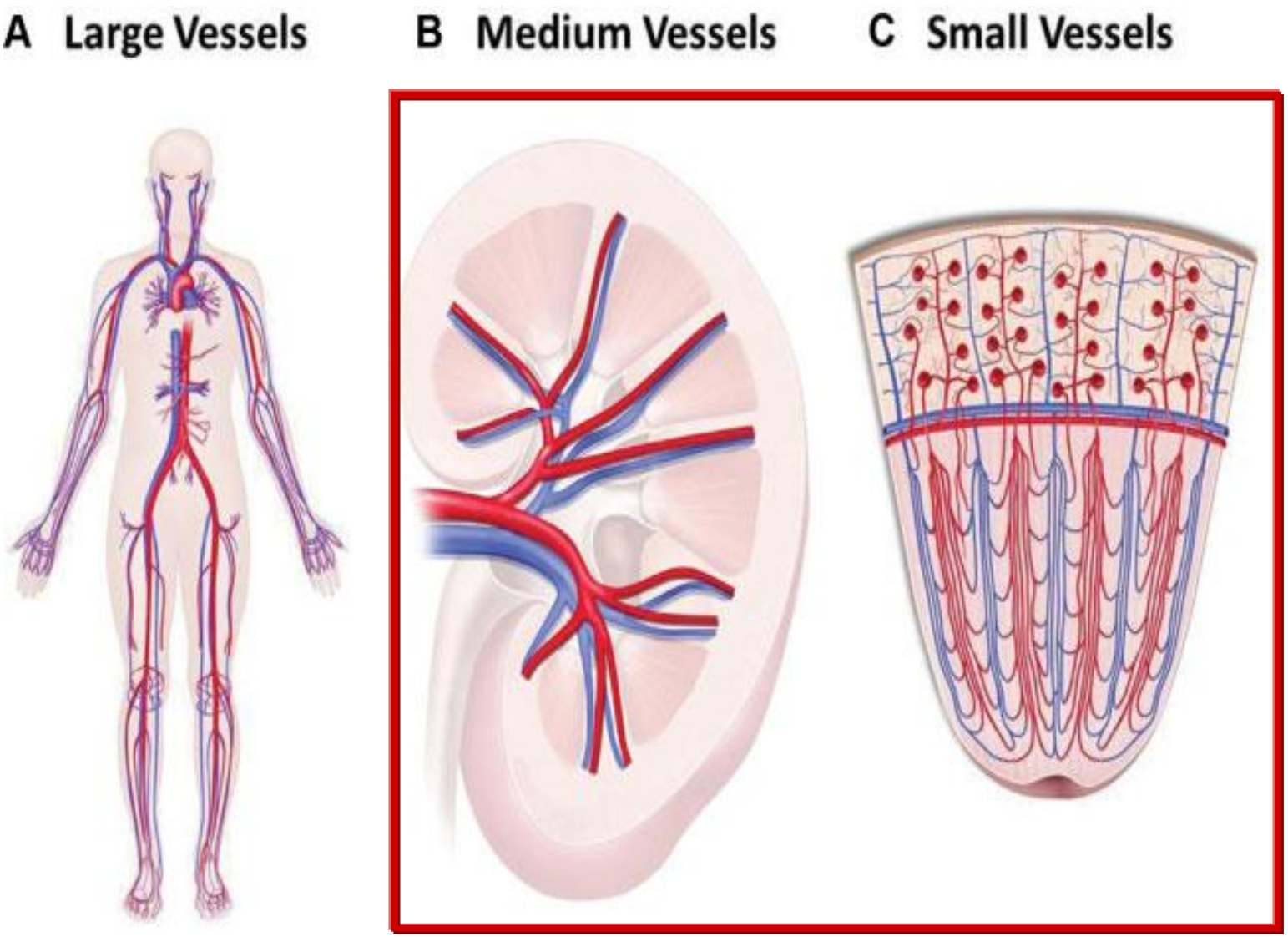
Aucun pour cette présentation

Définition

•Les vascularites systémiques sont caractérisées par une atteinte inflammatoire des vaisseaux sanguins artériels et veineux aboutissant à l'altération de la paroi vasculaire et à la constitution de thromboses.

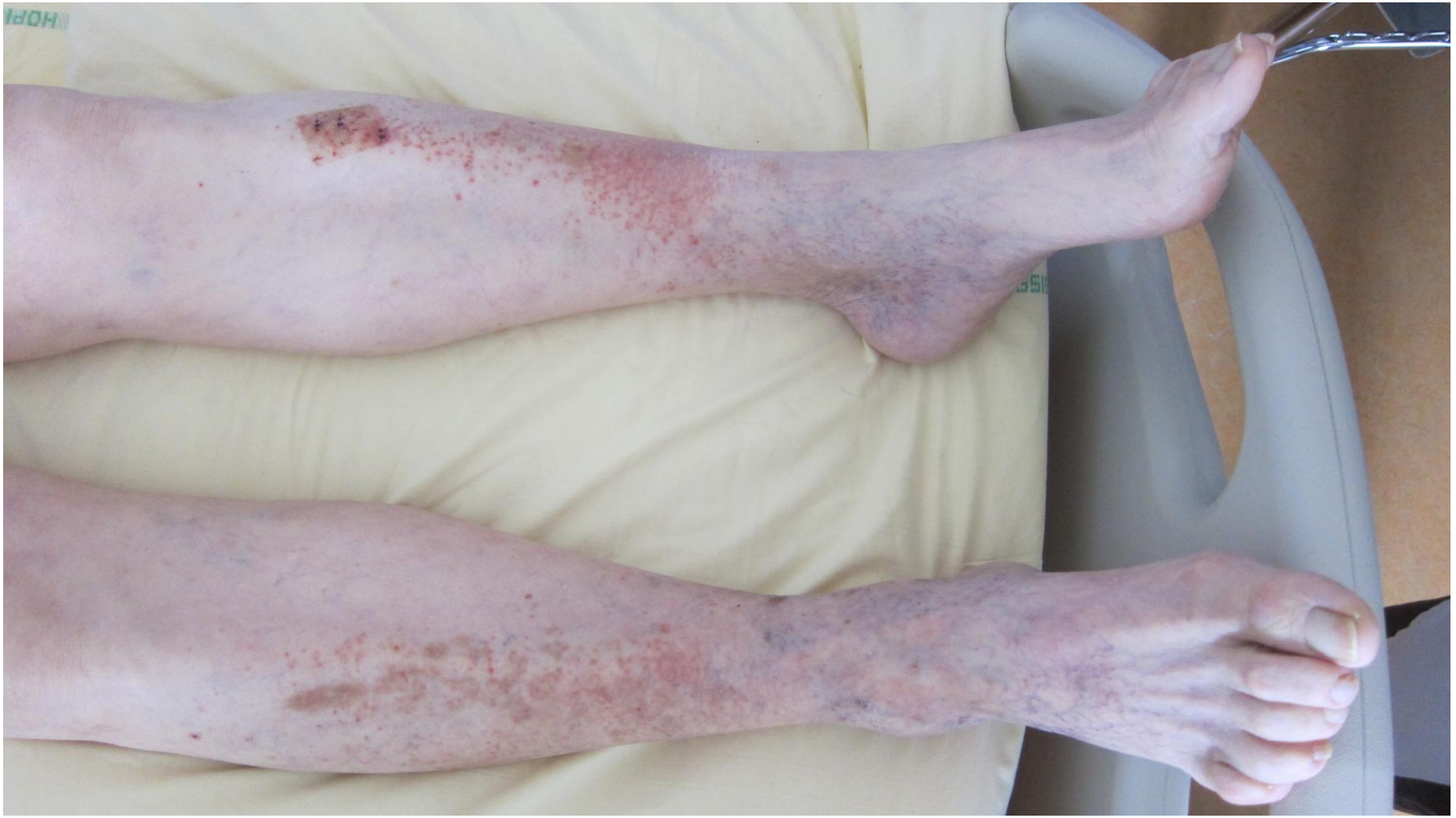
• Le polymorphisme clinique dépend de la taille et de la distribution des vaisseaux atteints.

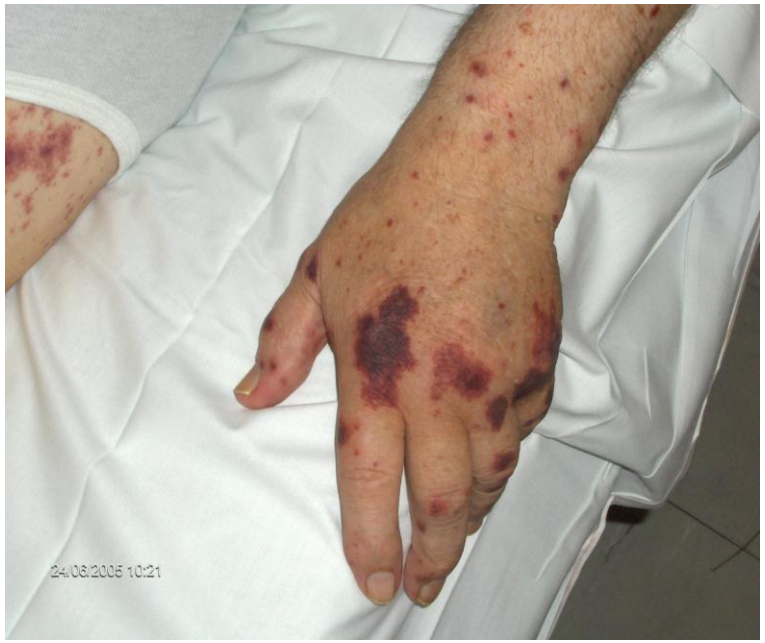
Classification des vascularites



Jennette, Arthritis Rheum, 2012

Purpura vasculaire

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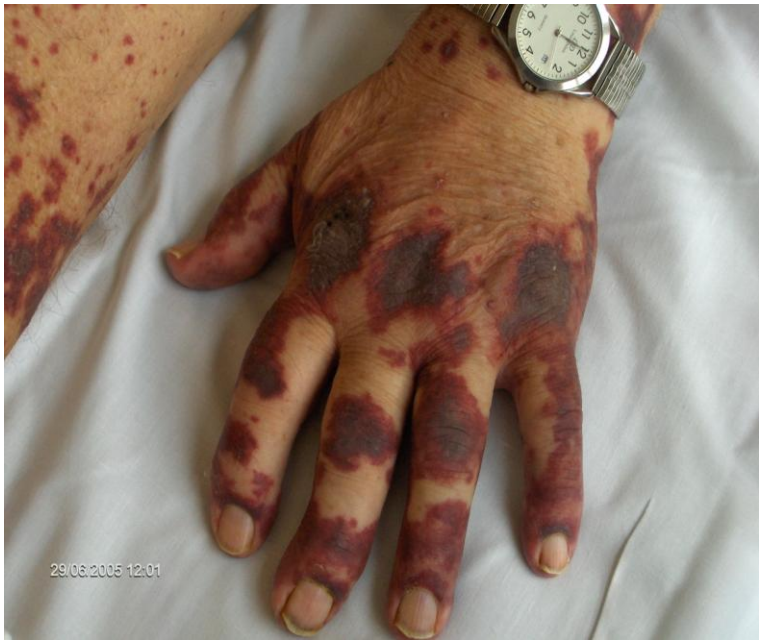
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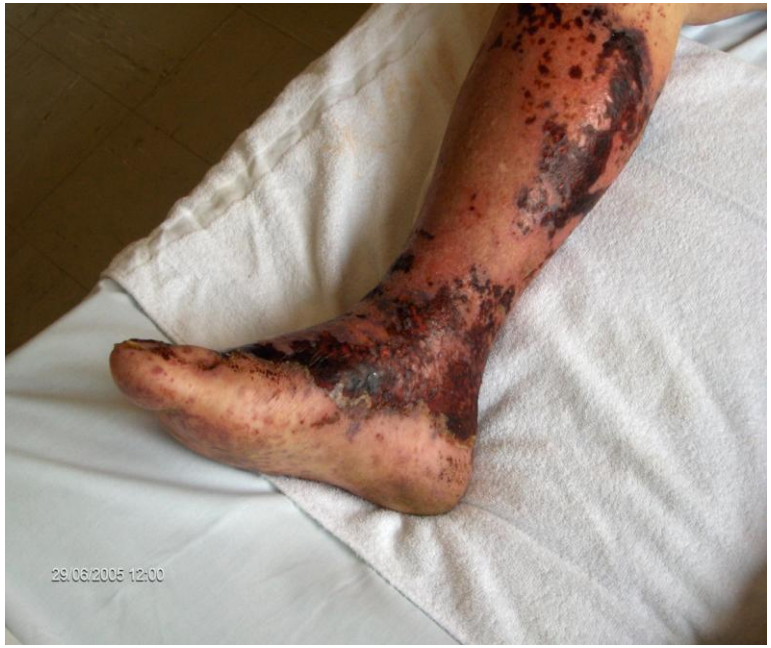
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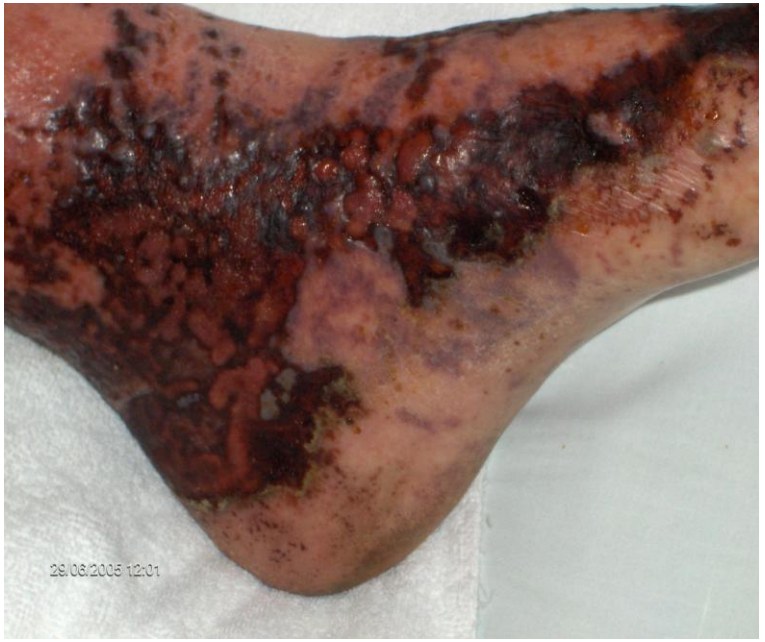
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1990 ACR Classification

Summary of ACR criteria and their limitations [12–18].

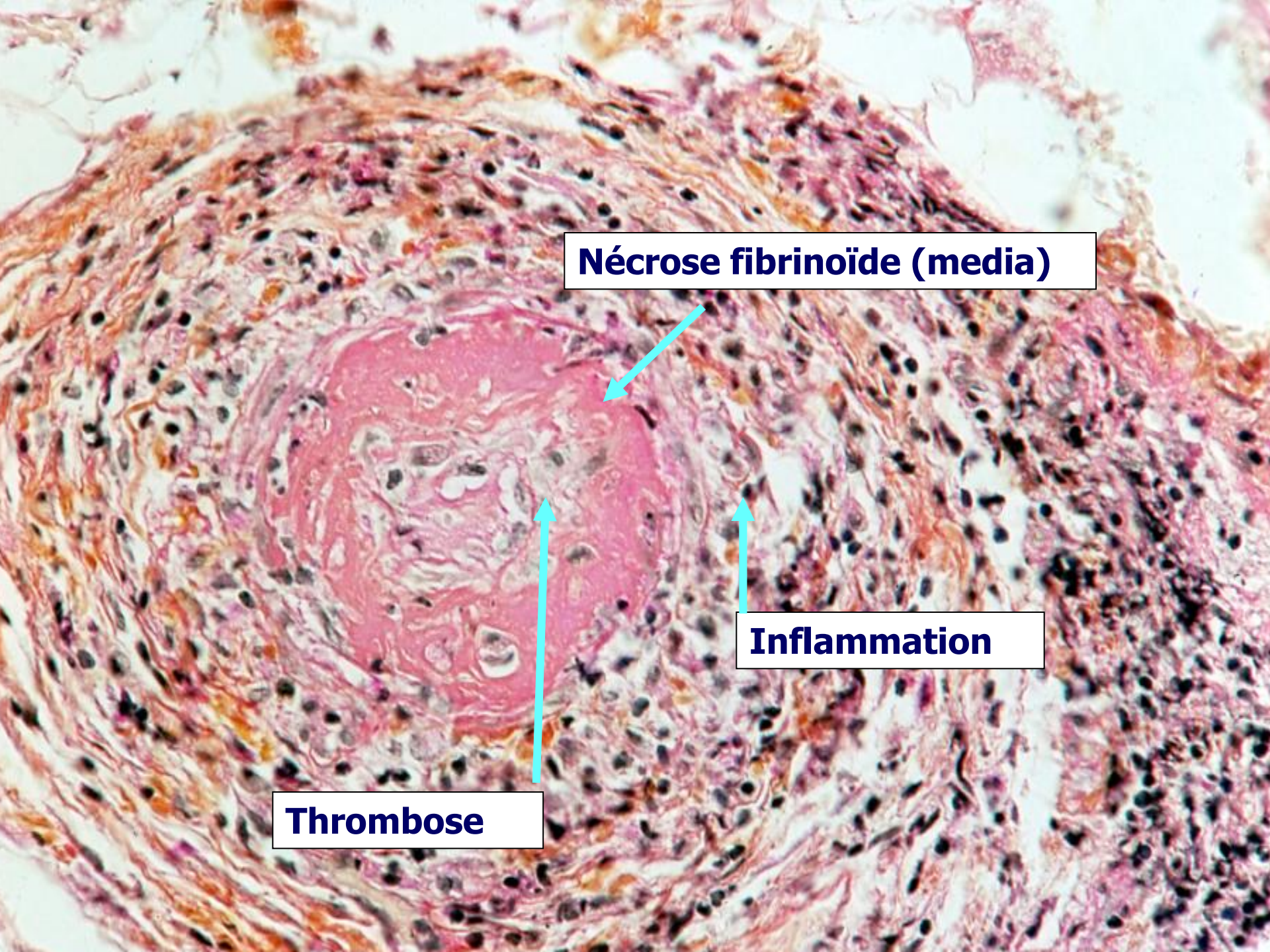
Type of Vasculitis	Sensitivity	Specificity	Limitations
GCA	93.5%	91.2%	Temporal artery biopsy is an important diagnostic tool but is not an obligatory criterion.
TAK	90.5%	97.8%	Newer imaging modalities, such as CT PET maybe useful, but are not included
GPA	88.2%	92%	No clear discrimination between GPA and MPA, or other mimics of GPA. Does not incorporate ANCA test.
EGPA	85%	99.7%	No inclusion of common features such as cardiac manifestations and rash. Does not incorporate ANCA test.
PAN	82.2%	86.6%	No absolute requirement for arteriography, or biopsy findings. No clear discrimination between PAN and MPA.
IgAV	87.1%	87.7%	Do not distinguish between IgAV from allergic reactions, or infectious related purpura. Common features; arthritis and nephritis are excluded. Age set as important criteria, but almost 30% of patients were above the age of 20.
Hypersensitivity vasculitis	71%	83.9%	Difficult to distinguish from IgAV
Microscopic polyangiitis			Not recognized by ACR

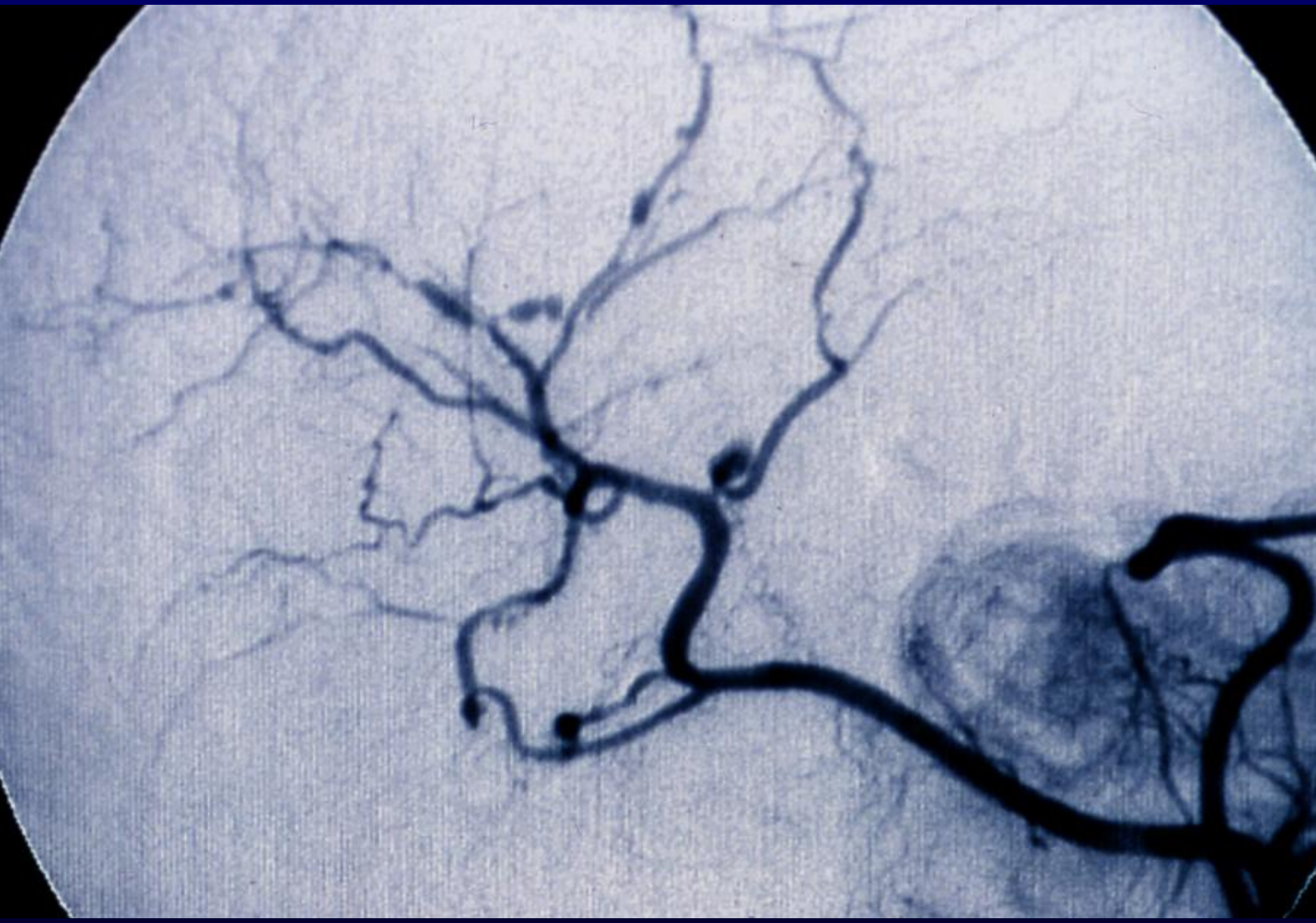
Pas maladie de Behcet, pas ANCA, IgA, cryo, anti-GBM

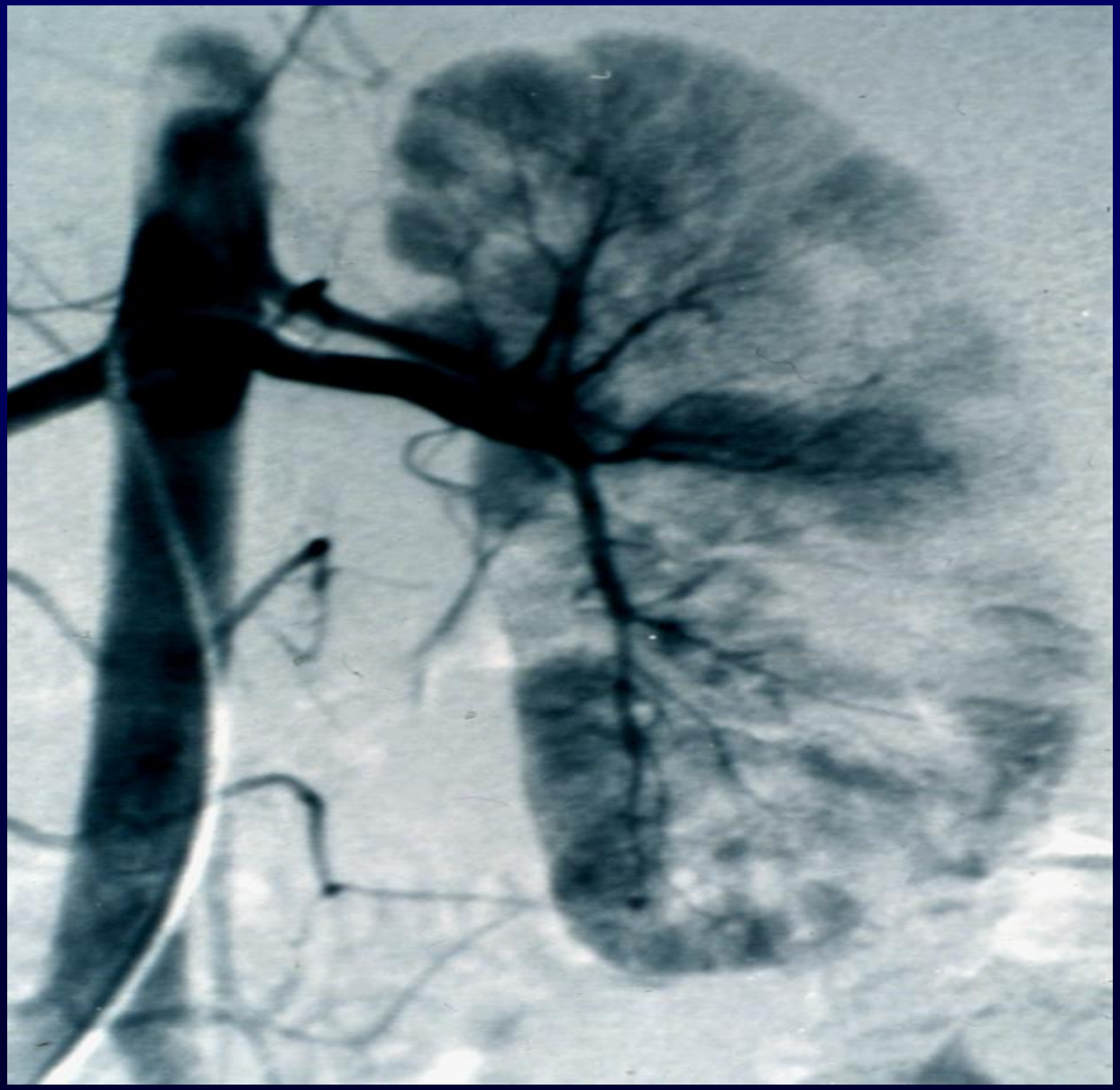
PAN : 1990 ACR CRITERIA

- Weight loss < 4kg
- Livedo reticularis
- Testicular pain or tenderness
- Mono- or polyneuropathy
- Diastolic BP > 90 mm Hg
- Elevated BUN or creatininemia
- Hepatitis B virus
- Biopsy of small or medium-sized artery containing PMN

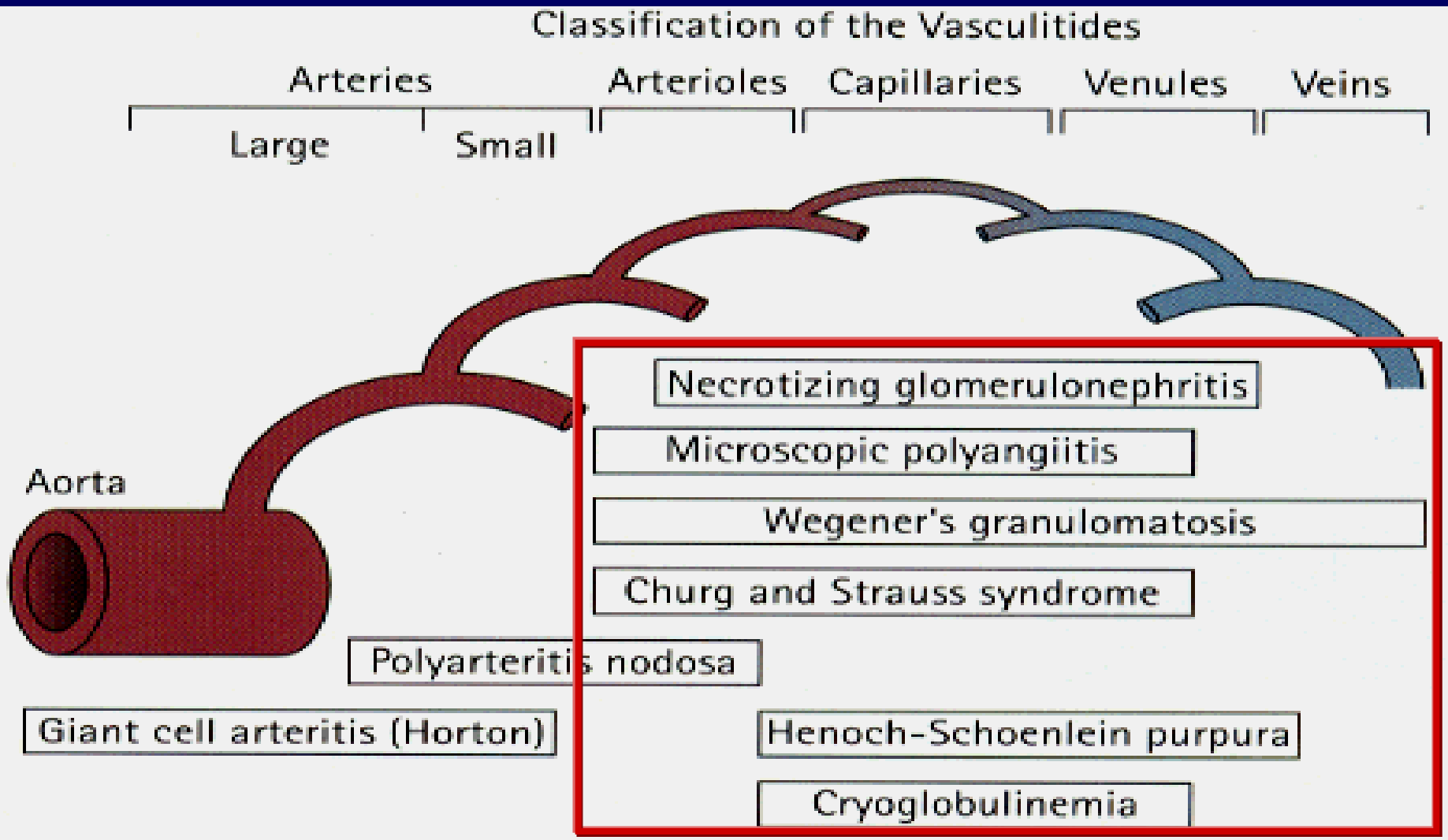
3 of 10 criteria should be present


Nécrose fibrinoïde (media)
Inflammation
Thrombose



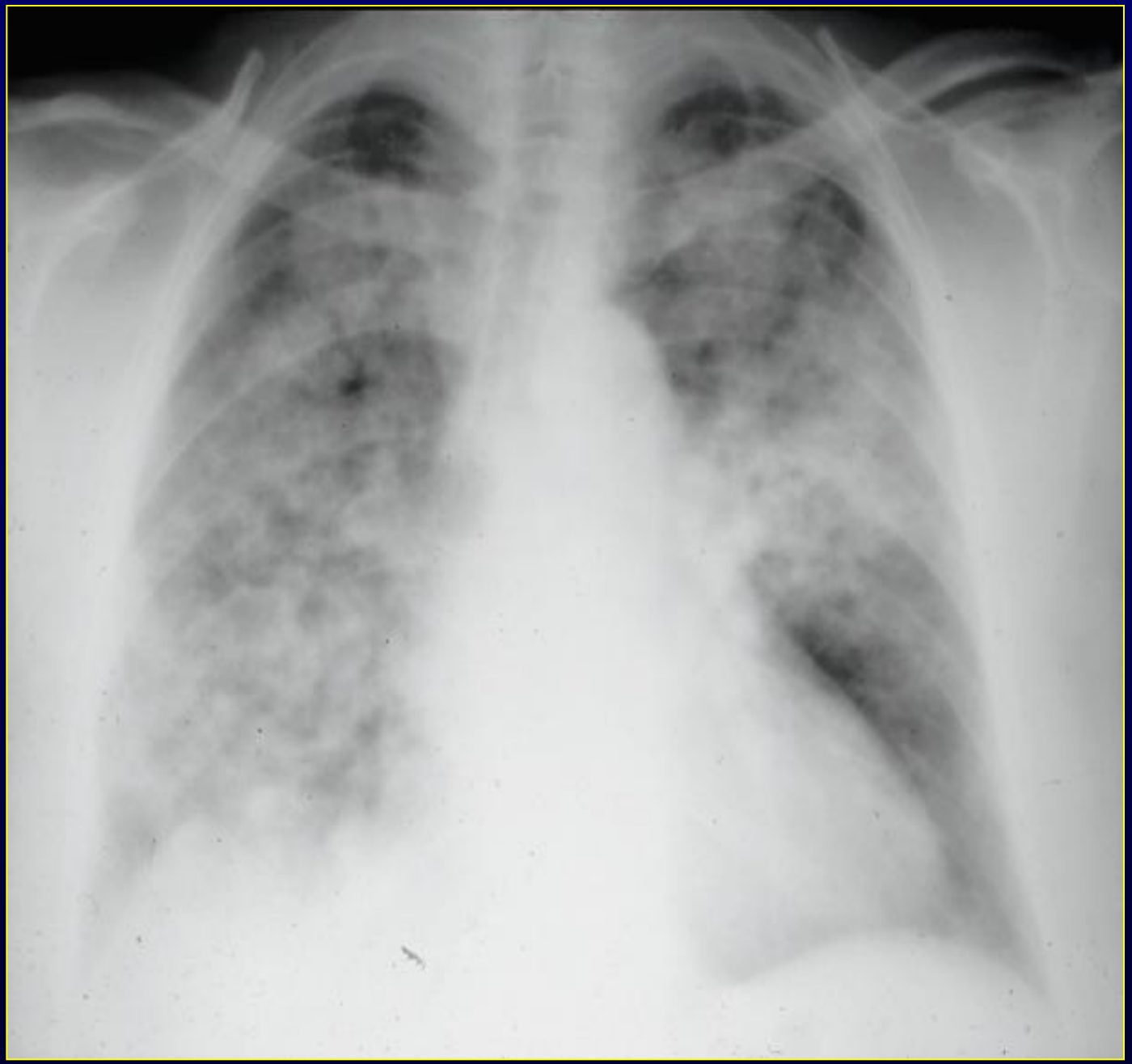


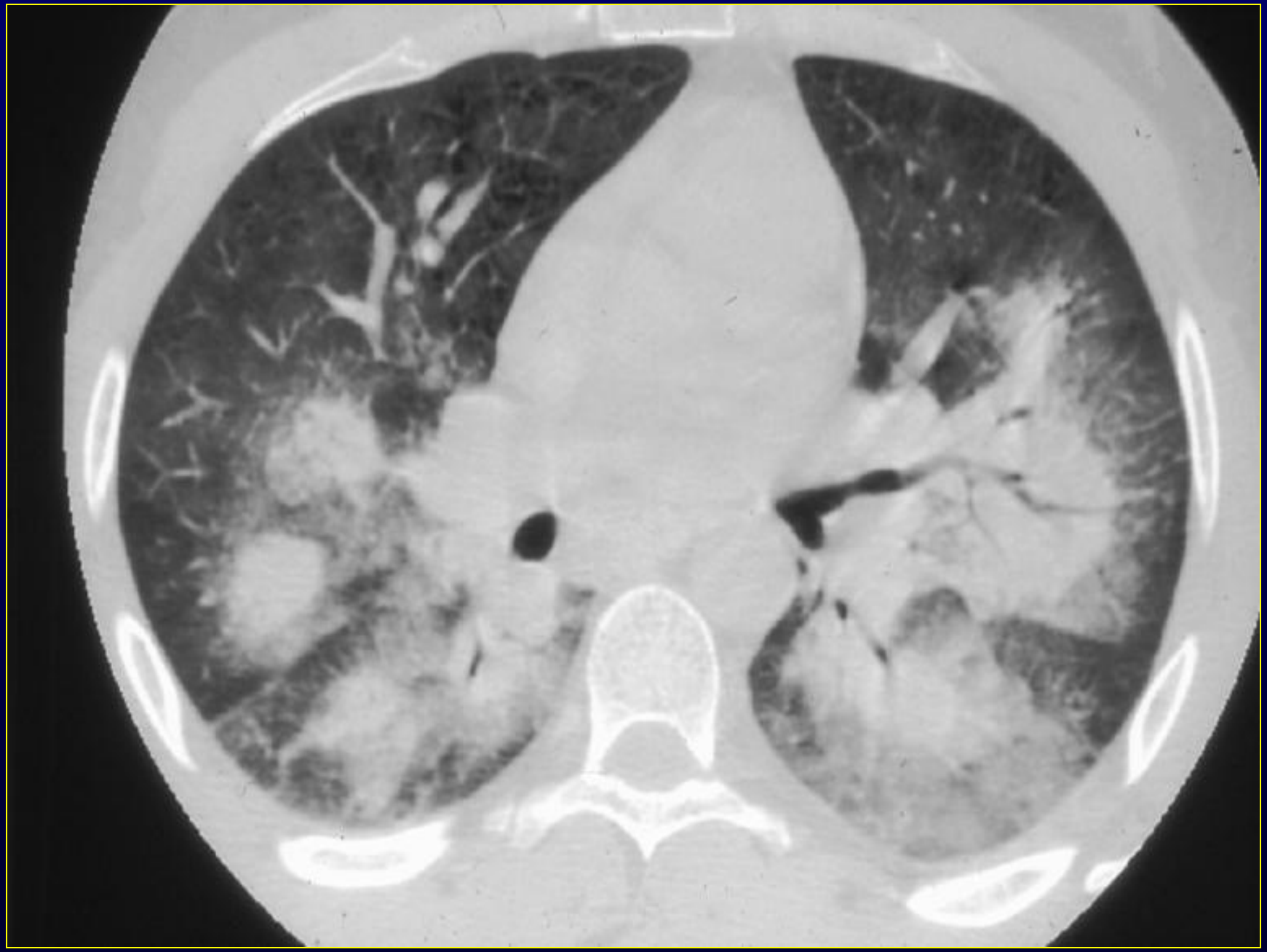
CHAPEL HILL NOMENCLATURE



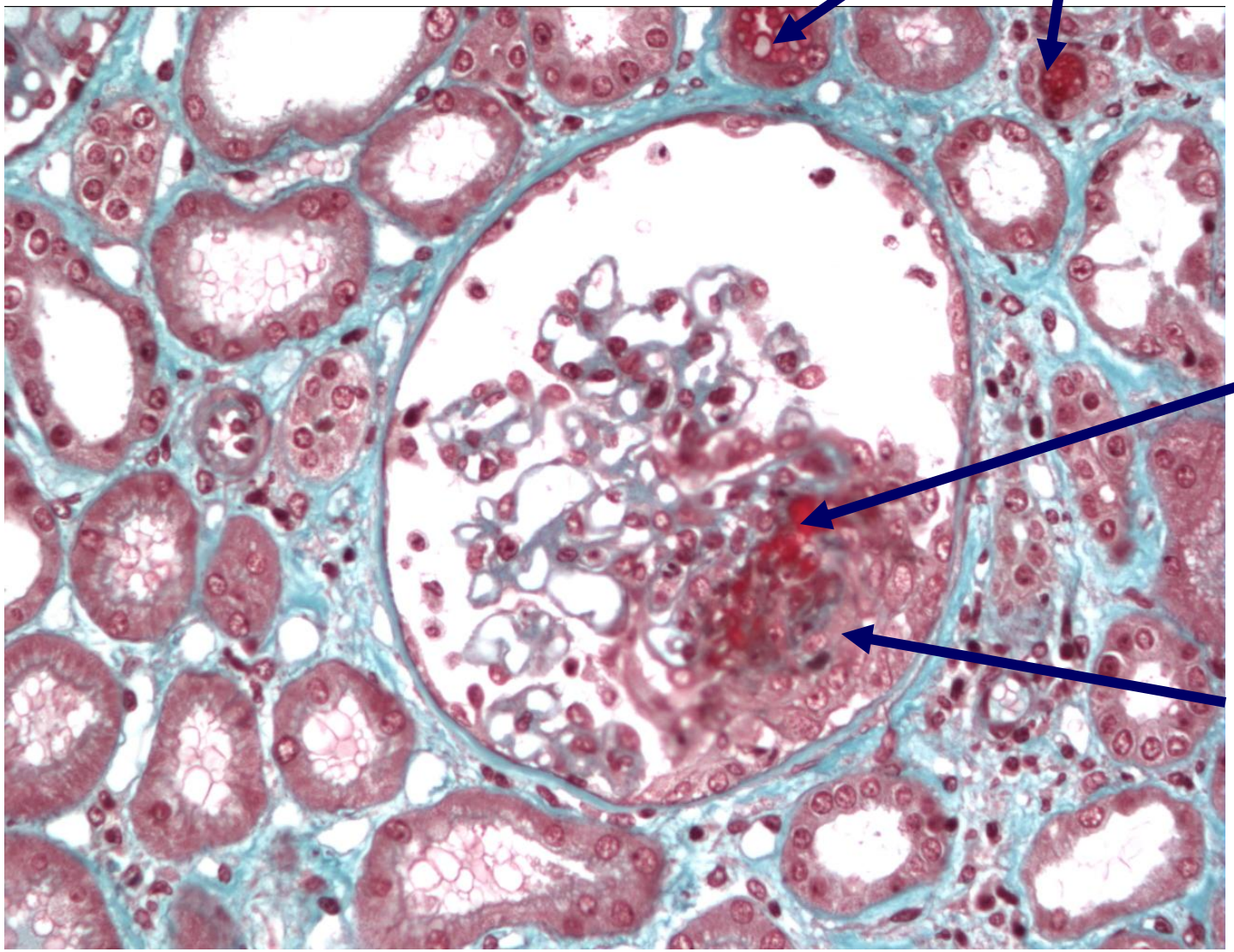
Polyangéite microscopique vs. PAN

	MPA	PAN
Vaisseaux	Petits	Moyens
Granulomes	Non	Rare
VHB	Non	Rare
ANCA	> 50%	Non
Atteinte glomérulaire	Oui	Non
Atteinte vasculaire rénale	Non	Oui
Atteinte pulmonaire	Oui	Non



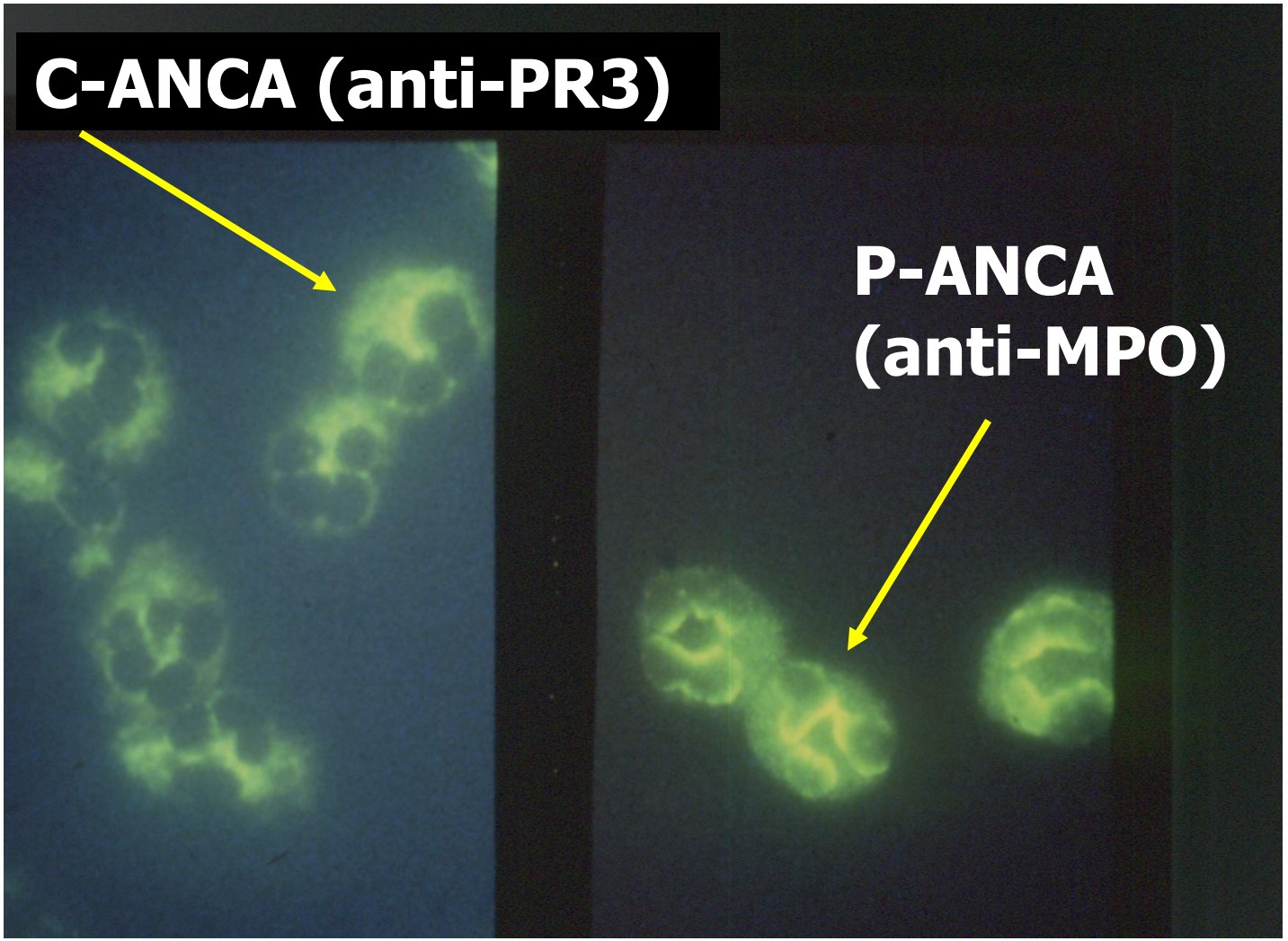


Polyangéite microscopique

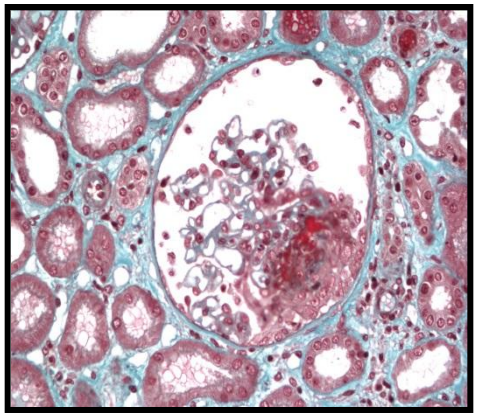


La bandelette urinaire fait partie de l’examen clinique !

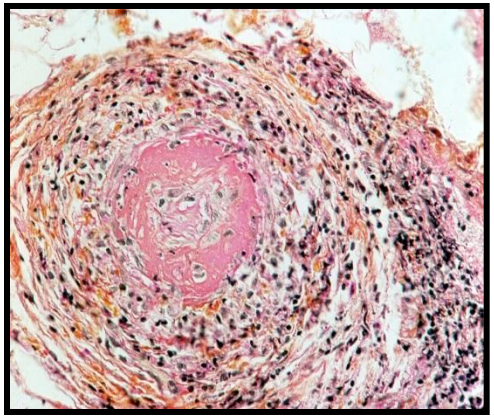
ANCA DANS LES VASCULARITES SYSTEMIQUES

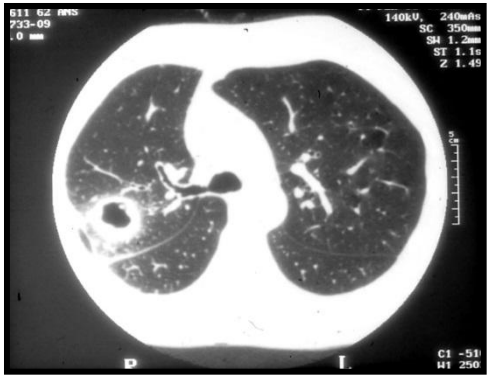


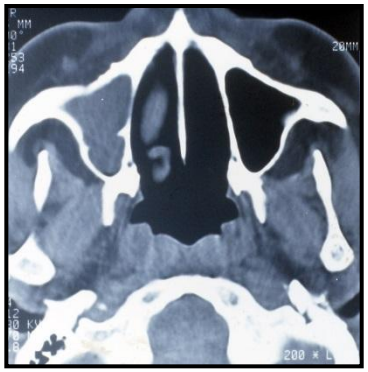
ANCA-associated vasculitides



- Vascular necrosis and perivascular inflammation in small vessels
- Systemic disease because of renal and lung involvements

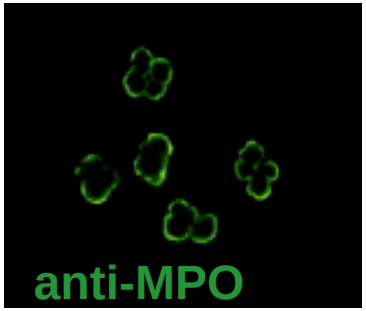


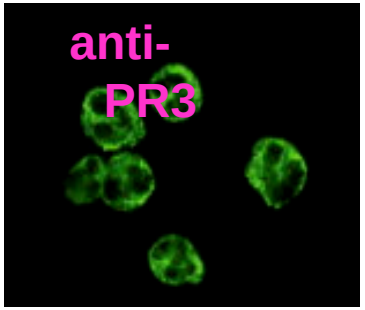




- Diagnostic value of ANCA

	anti-MPO	anti-PR3
Wegener's granulomatosis	10 %	85 %
Microscopic polyangiitis	60 %	30 %
Churg-Strauss	31 %	<10 %





Critères de classification

Critères ACR
Granulomatose de Wegener

Inflammation nasale ou buccale
Anomalies radiologiques pulmonaires
Anomalie du sédiment urinaire
Granulomes inflammatoires à la biopsie

Diagnostic si > 2/4 critères

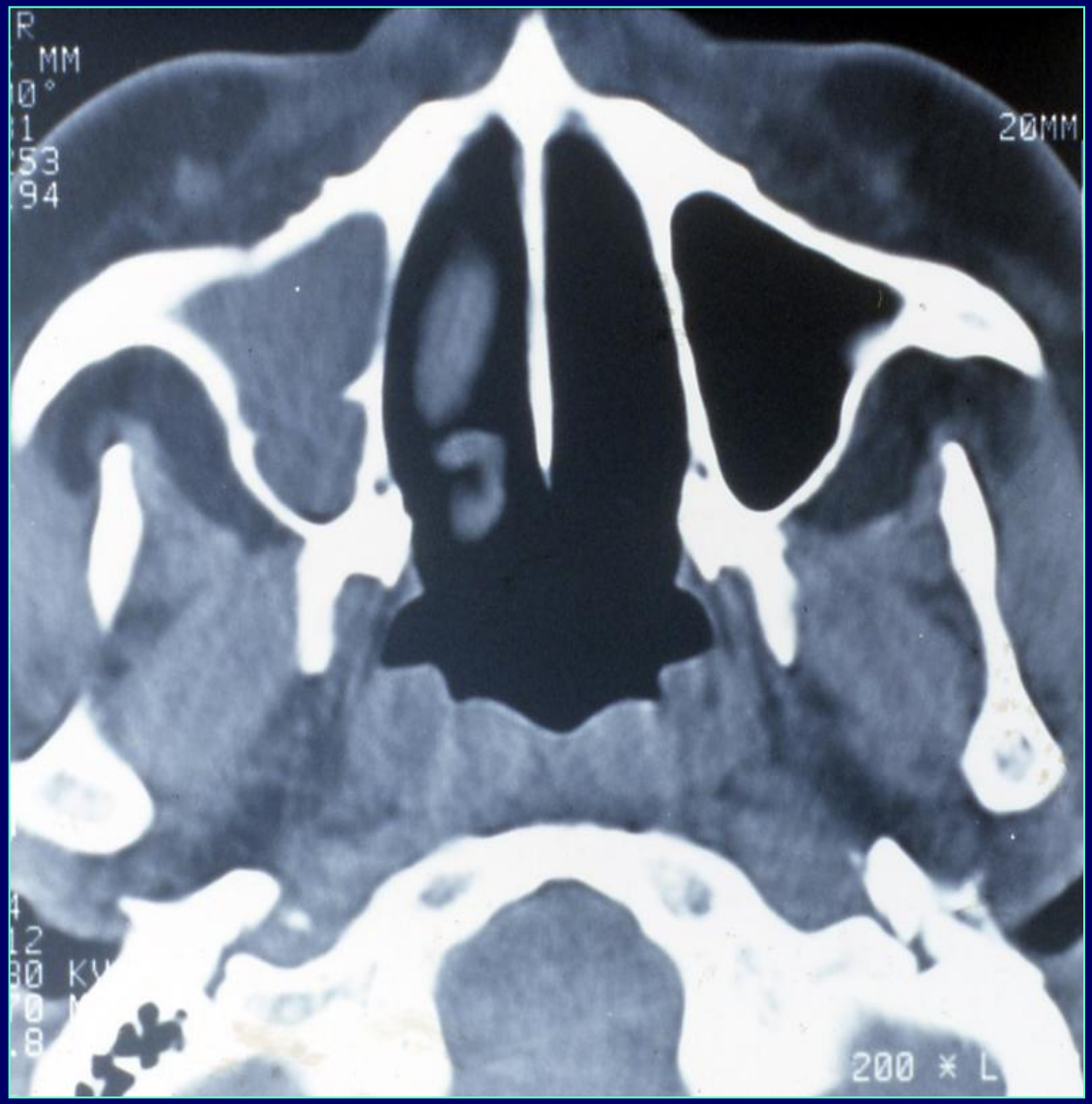
Critères ACR
Churg-Strauss

Asthme
Eosinophilie >10%
Mono ou poly-neuropathie
Infiltrats pulmonaires labiles
Anomalies sinusiennes
Eosinophiles sur biopsie

Diagnostic si > 4/6 critères

Leavitt, Arthritis Rheum, 1990

Granulomatose de Wegener

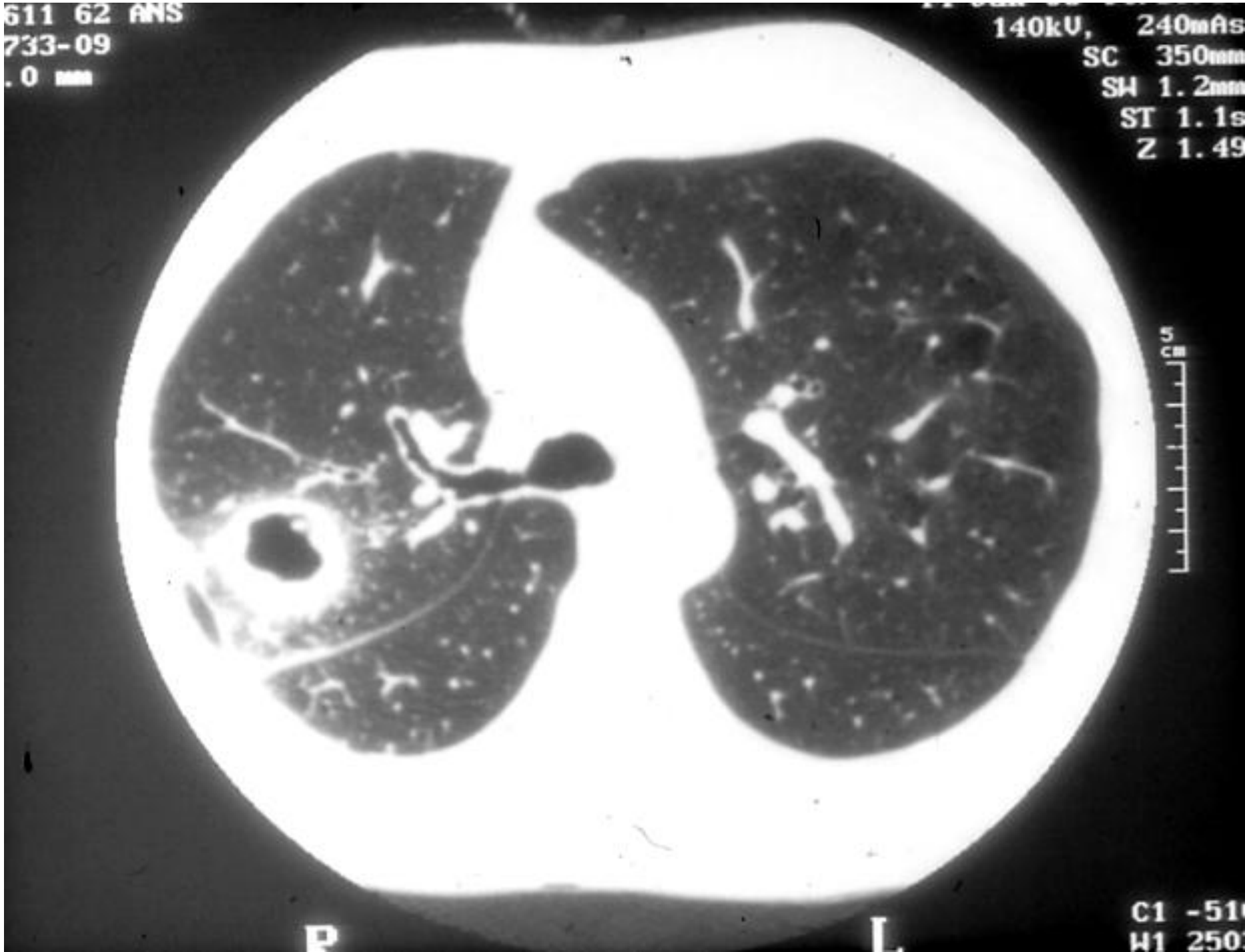


Les signes ORL peuvent révéler la GW

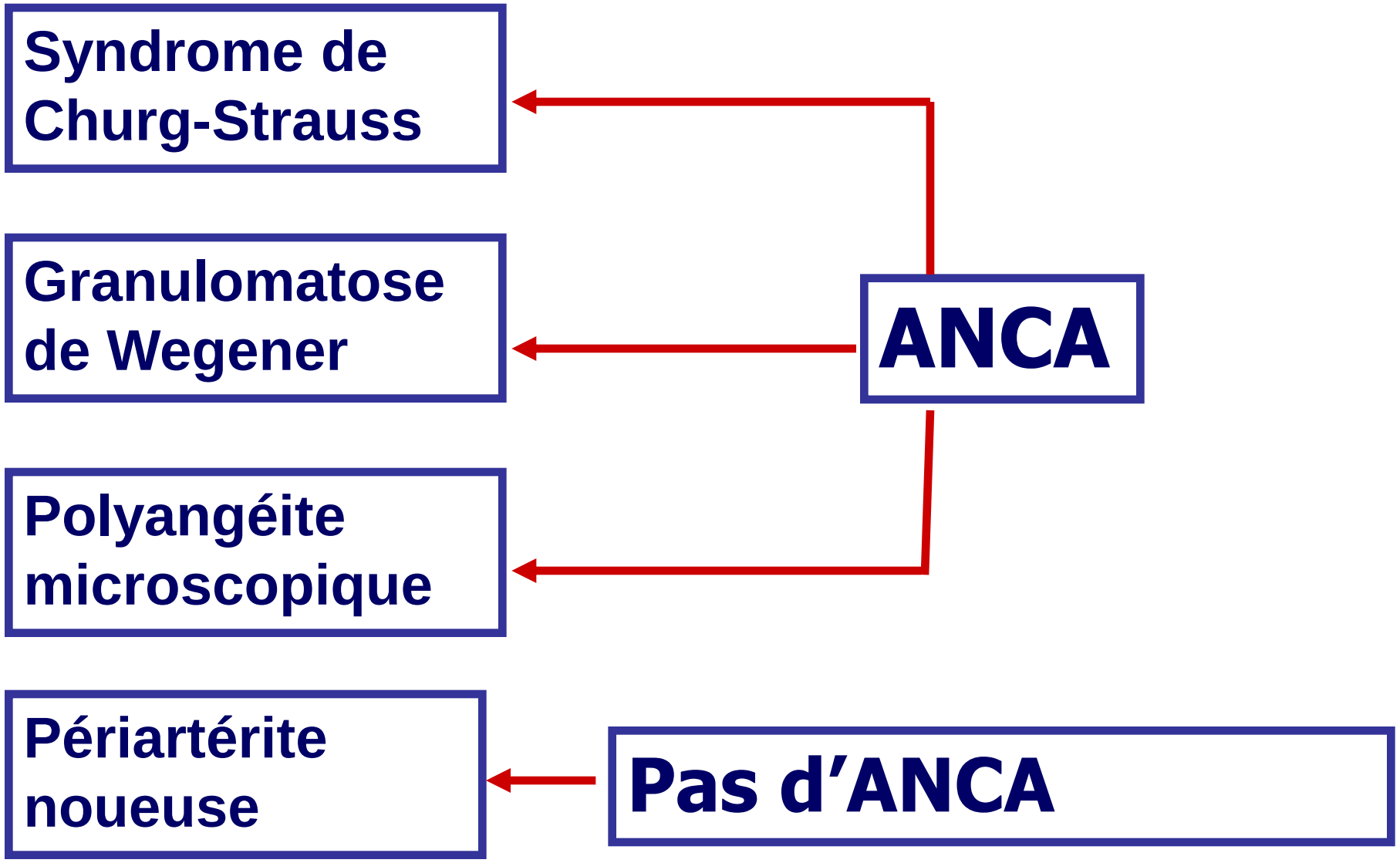
-Destruction

-Sinusite

-Rhinite


611 62 ANS
733-09
.0 mm
140kV, 240mAs
SC 350mm
SH 1.2mm
ST 1.1s
Z 1.49
5 cm
R
L
C1 -51
W1 250

PHYSIOPATHOLOGIE DES VASCULARITES SYSTÉMIQUES TOUCHANT LES VAISSEAUX DE PETIT ET MOYEN CALIBRE



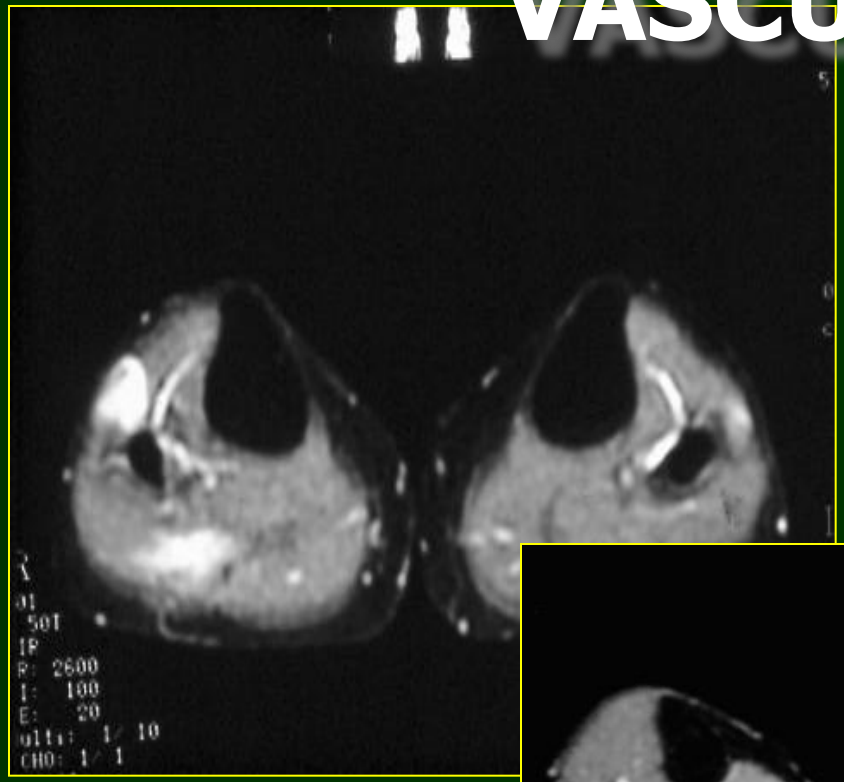
Vascularites des petits vaisseaux: quand y penser ?

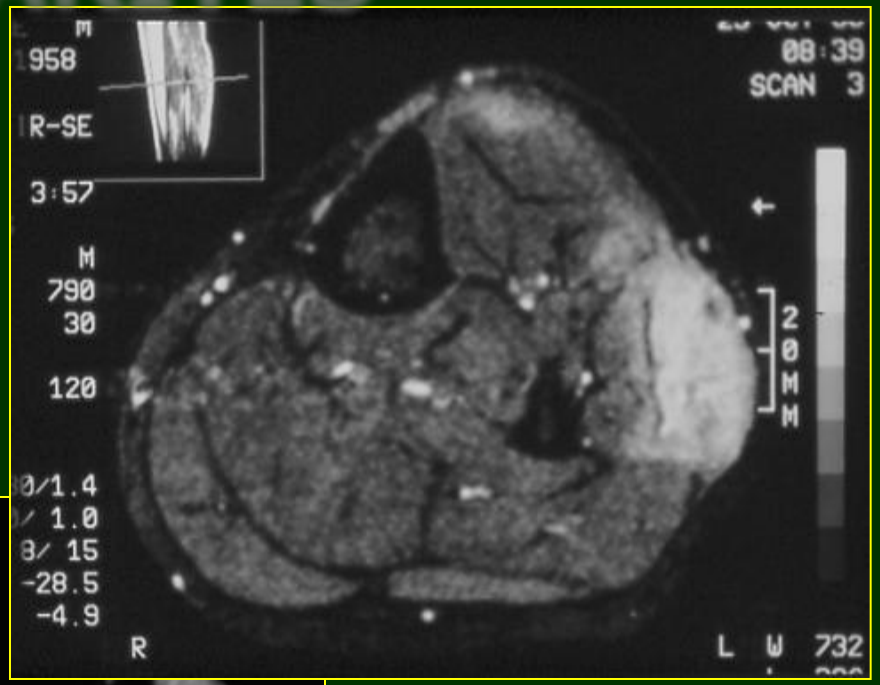
Premiers signes cliniques attribuables à une vascularite (PAN – MPA)

Characteristic	Total
Poor condition	51 (71)
Only general symptoms	6 (8)
Fever	32 (44)
Weight loss	30 (42)
Asthenia	24 (33)
Myalgias	31 (43)
Arthralgias	21 (29)

Agard C et al. Arthritis Care Res 2003

DIAGNOSTIC DES VASCULARITES





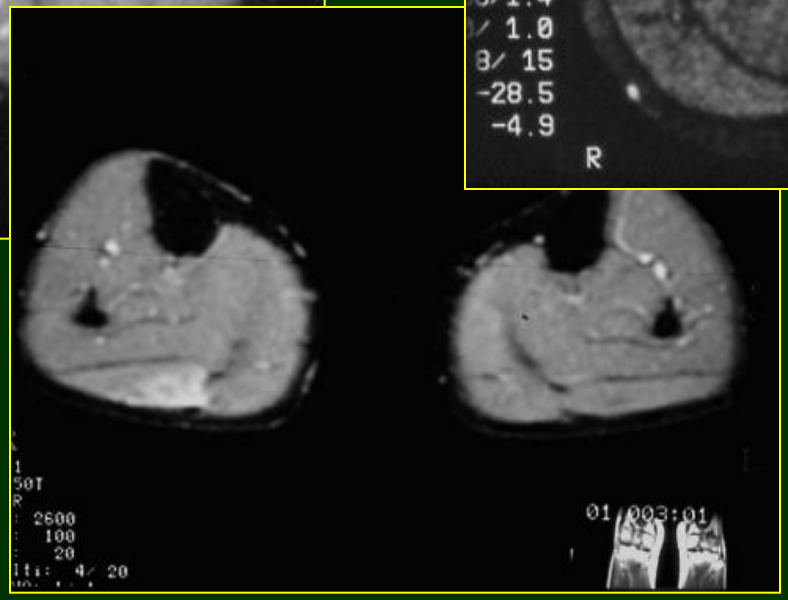


Table 1. First clinical signs attributable to vasculitis and time to diagnosis for 72 patients with PAN (n = 36) or MPA (n = 36)*

Characteristic	Total	PAN	MPA	*P*
Poor condition	51 (71)	29 (81)	22 (61)	0.07
Only general symptoms	6 (8)	0	6 (17)	0.02
Fever	32 (44)	17 (47)	15 (42)	0.64
Weight loss	30 (42)	18 (50)	12 (33)	0.15
Asthenia	24 (33)	15 (42)	9 (25)	0.13
Myalgias	31 (43)	16 (44)	15 (42)	0.81
Arthralgias	21 (29)	10 (28)	11 (31)	0.80
Neurologic involvement	18 (25)	12 (33)	6 (17)	0.10
Peripheral neuropathy	16 (22)	12 (33)	4 (11)	0.02
Cerebral hemorrhage or infarction	3 (4)	1 (3)	2 (6)	0.56
Cutaneous involvement	9 (13)	4 (11)	5 (14)	1.0
Orchitis and/or epididymitis	3 (7)	3 (13)	0	0.24
Nocturnal sweating	5 (7)	1 (3)	4 (11)	0.36
Cardiac involvement	4 (6)	1 (3)	3 (8)	0.61
Headache and/or temporal arteritis	8 (11)	5 (14)	3 (8)	0.45
Gastrointestinal symptoms	10 (14)	9 (25)	1 (3)	0.006
Renal insufficiency (creatininemia $\geq$ 140 μmol/liter)	2 (3)	2 (6)	0	0.49
Raynaud's phenomenon	6 (8)	3 (8)	3 (8)	1.0
Number of involved organs/tissues, mean ± SD	0.66 ± 0.69	0.86 ± 0.59	0.47 ± 0.49	0.02
Time to diagnosis, months, mean ± SD	9.8 ± 19.4	6.6 ± 11.5	12.9 ± 18.9	0.17

* Values are numbers (%) unless otherwise specified. PAN = polyarteritis nodosa; MPA = microscopic polyangiitis.

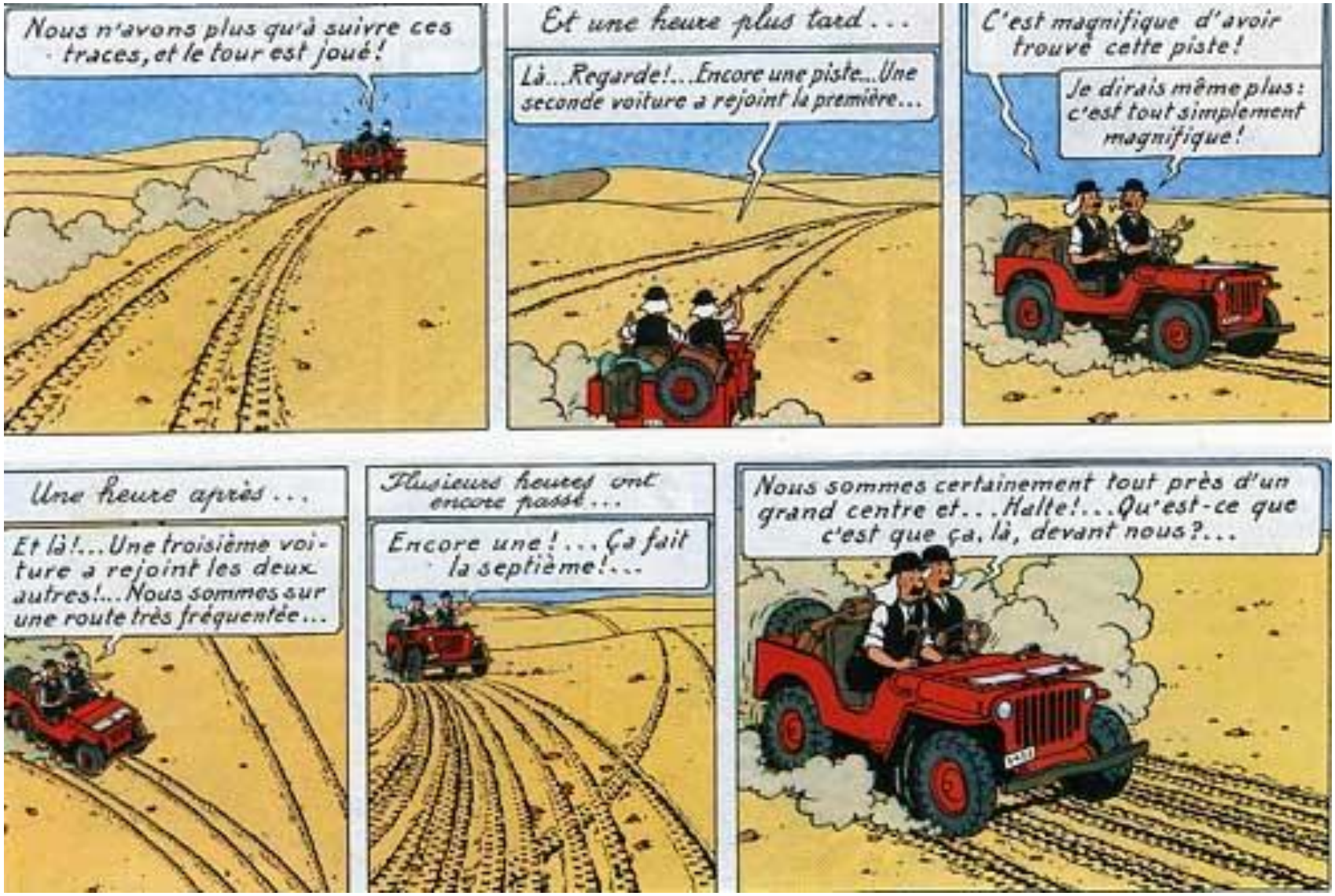
Agard C et al. Arthritis Care Res 2003

Signes cliniques au diagnostic (PAN – MPA)

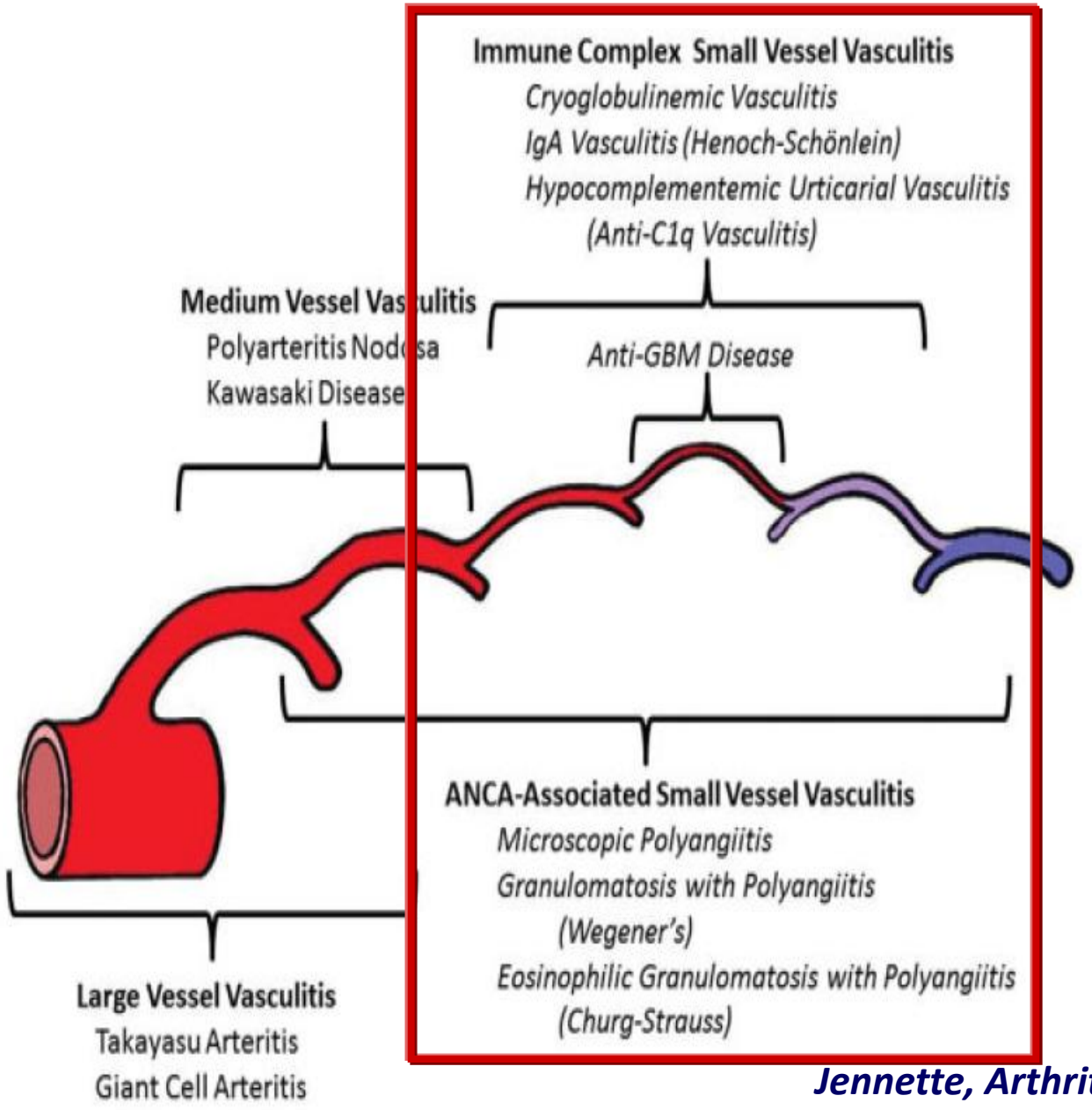
Poor condition	56 (78)
Fever	39 (54)
Weight loss	48 (67)
Myalgias	40 (56)
Arthralgias	36 (51)
Neurologic involvement	59 (82)
Peripheral neuropathy	47 (65)
Cerebral hemorrhage/infarction	6 (8)
Pulmonary involvement	10 (14)
Alveolar hemorrhage	4 (6)
Renal involvement	15 (21)
Gastrointestinal symptoms	27 (38)
Cutaneous involvement	29 (40)
Orchitis/epididymitis	6 (13)
Cardiac involvement	12 (17)
Acute ischemia of the extremities	7 (10)

Agard C et al. Arthritis Care Res 2003

Ce que Chapel Hill 2012 n'est pas….



Chapel Hill nomenclature: revision



Jennette, Arthritis Rheum, 2012

2012 Chapel Hill nomenclature

Abbreviation	Definition	Previous terminology
GPA	Granulomatosis with polyangiitis	Wegener's granulomatosis
EGPA	Eosinophilic granulomatosis with polyangiitis	Churg-Strauss
MPA	Microscopic polyangiitis	
IgA vasculitis	IgA vasculitis	Henoch-Schonlein purpura

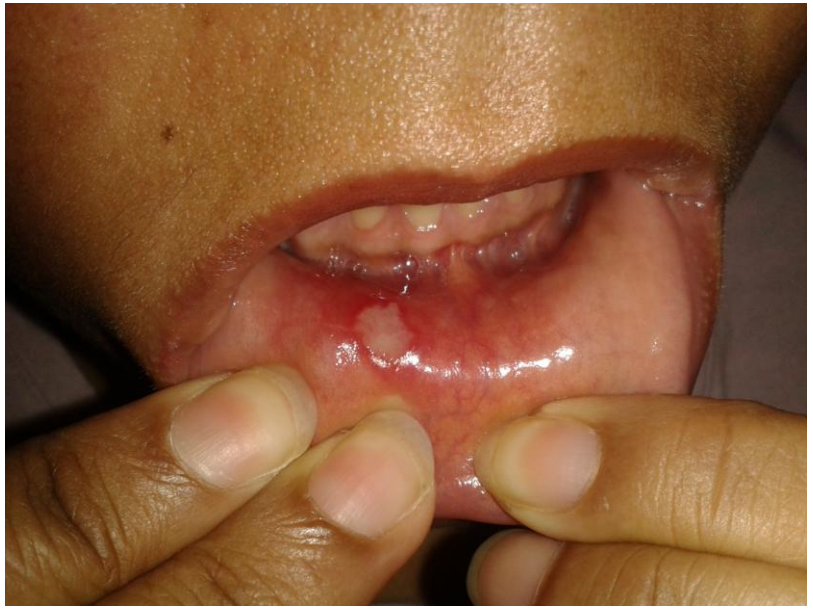
Other new terminology and minor modifications	Previous terminology
Anti-GBM disease	Goodpastures disease
Cutaneous arteritis	Cutaneous PAN
Cutaneous leukocytoclastic angiitis	
Hypocomplementemic urticarial vasculitis	

Jennette, Arthritis Rheum, 2012

2012 Chapel Hill nomenclature

- Formes localisées de vascularites des petits vaisseaux

  GPA et EGPA



- Variable Vessel Vasculitis

  Behcet, Cogan

- Single-organ vasculitis

  peau, testicules, SNC, SNP, rein

- Vascularites secondaires (PR, lupus, virus)

Jennette, Arthritis Rheum, 2012

Vascularites secondaires

- Atteinte cutanée ++
- Étiologies;

  -Infections: bactériennes (streptocoque), virales (hépatites)

  -Paranéoplasiques: cancers, hémopathies

  -Médicaments

  -Connectivites

-Ttt: celui de la cause

Vascularites des petits vaisseaux: Diagnostic différentiel

Diagnostic des vascularites

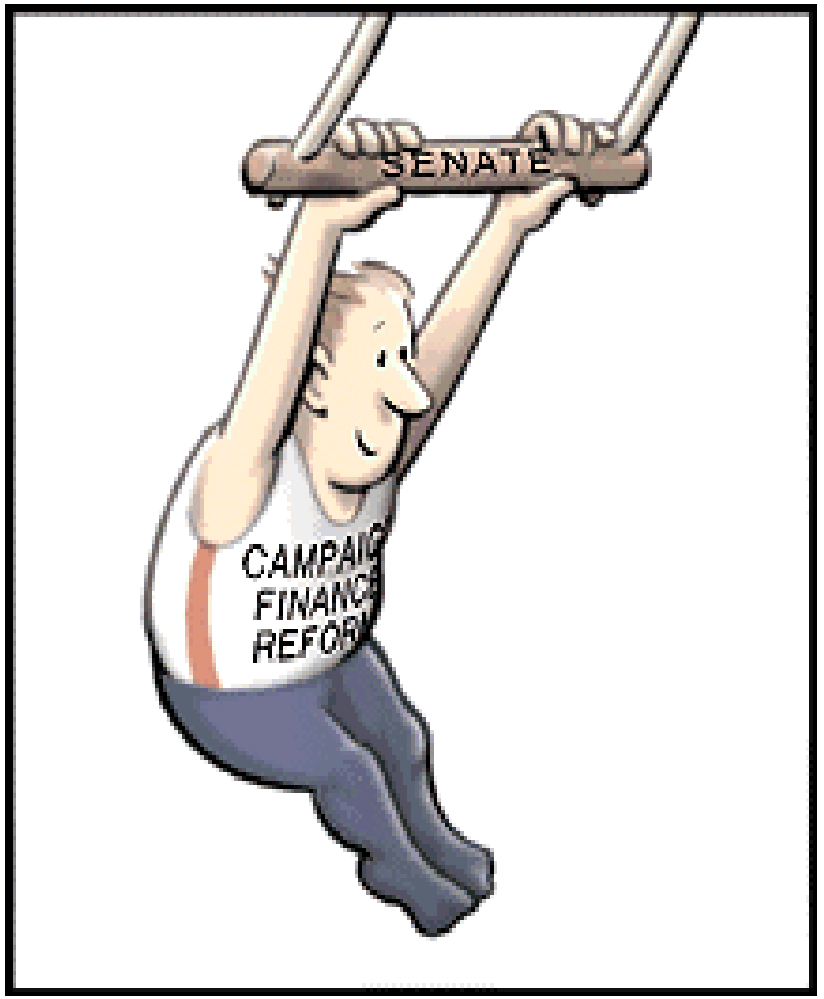
Attention aux diagnostics « au chausse pieds »

Vascularites: diagnostic différentiel

- Embolies septiques des endocardites
- Embols de cholestérol
- Micro-angiopathie thrombotique
- Syndrome des anti-phospholipides
- Localisation angiotrope des lymphomes
- Ergotisme chronique

Conclusions

- Les premiers signes cliniques des vascularites intéressant les vaisseaux de petit et/ou moyen calibre sont non spécifiques: altération de l'état général fébrile avec arthralgies et myalgies
- La biopsie reste un élément déterminant
- Les ANCA constituent une aide importante au diagnostic
- Attention aux diagnostics différentiels: endocardite; embols de cholestérol
- Penser aux vascularites secondaires: médicaments



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