Les neuropathies dysimmunitaires (partie 1)

Cours DES
Guillaume Taieb, Neurologie,
Gui de Chauliac, Montpellier

Cas clinique n°1

Femme 35 ans,

- engourdissement + paresthésies > III doigt G
  > I et V doigts G
  > main G avec maladresse
  > MS G jusqu'au 2/3 du bras
- installation chronique (> 2mois)
- évolution progressive sur 1 an

Contexte de perte de 5 kg

ATCD tabac, reste néant

Clinique: anesthésie en de C5 à C8 G + aréflexie MSG

Video retirée

- Neurographie motrice des 4 membres normale, en dehors d'une onde F absente sur le SPE droit

- Neurographie sensitive absence de potentiel sensitif

Au MS gauche : médian, ulnaire, radial, brachial cutané interne, musculo cutané.

- Myographie normale

- PL = 0.52 g/l sans cellule

Quelle hypothèse diagnostique vous semble la plus probable ?

A atteinte centrale des voies lemniscales

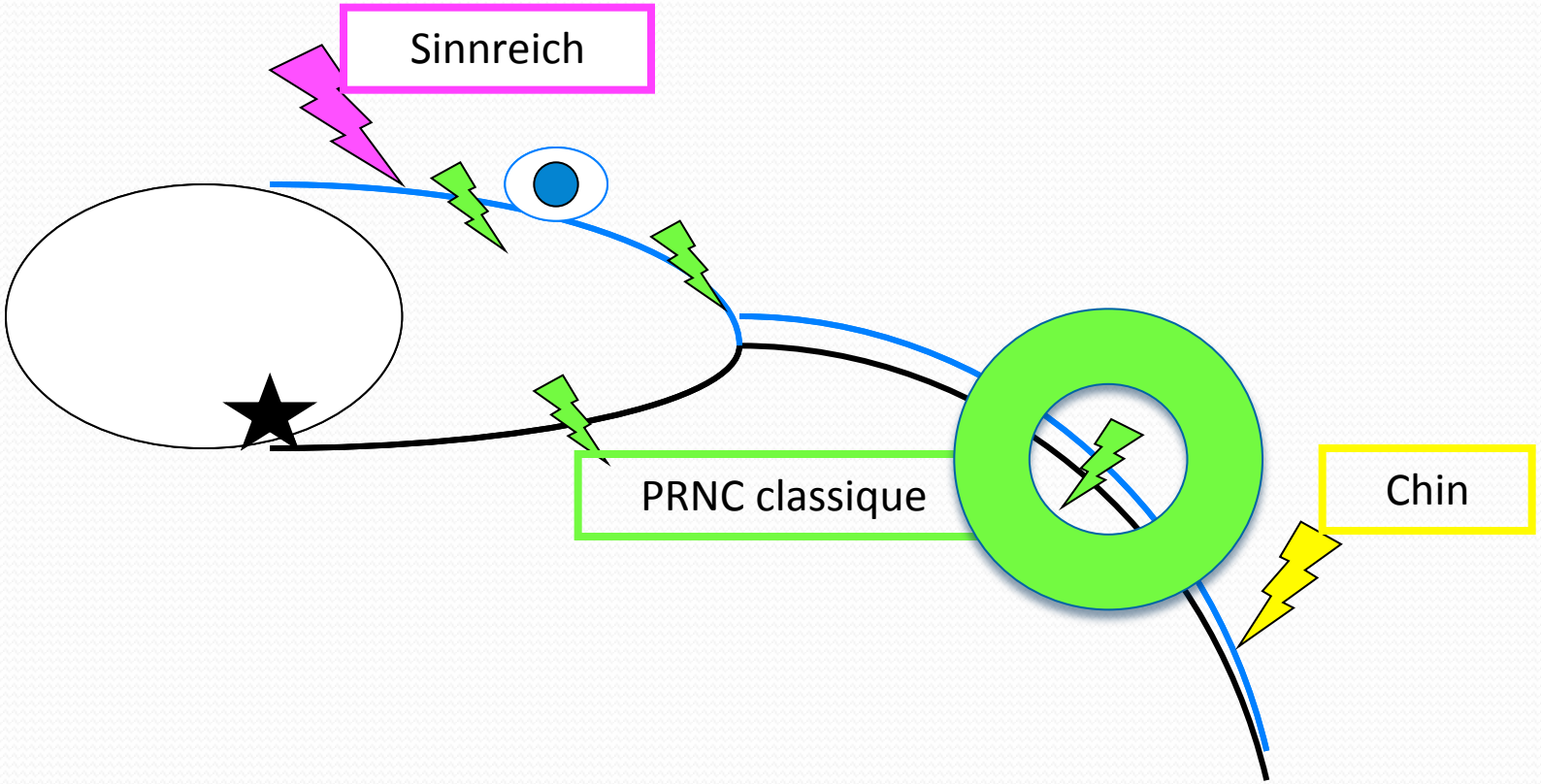
B PRNC monomélique sensitive pure pré ganglionnaire


Ganglionopathie


PRNC monomélique sensitive pure post ganglionnaire

E Multinévrite sensitive

PRNC sensitive pré et post ganglionnaire
Neurographie ss explore les fibres A bêta/gamma



Atteinte proximale sans DW complète

doi:10.1093/brain/awp136 Brain 2009: 132; 1723–1733 | 1723

BRAIN
A JOURNAL OF NEUROLOGY

The pattern and diagnostic criteria of sensory neuronopathy: a case–control study

Jean-Philippe Camdessanché,[1,2,3,4,5] Guillemette Jousserand,[1,2] Karine Ferraud,[1,2,3,4,5] Christophe Vial,[1,6] Philippe Petiot,[1,7] Jérôme Honnorat[5,6,8] and Jean-Christophe Antoine[1,2,3,4,5]

A In a patient with a clinically pure sensory neuropathy a diagnosis of SNN is considered as possible if score >6.5

	Yes	Points
a—Ataxia in the lower or upper limbs at onset or full development	☐	+3.1
b—Asymmetrical distribution of sensory loss at onset or full development	☐	+1.7
c—Sensory loss not restricted to the lower limbs at full development	☐	+2.0
d—At least 1 SAP absent or 3 SAP <30% of the lower limit of normal in the upper limbs, not explained by entrapment neuropathy	☐	+2.8
e—Less than two nerves with abnormal motor nerve conduction studies in the lower limbs	☐	+3.1

If >6.5, a diagnosis of SNN is possible Total

B A diagnosis of SNN is probable if the patient's score is >6.5 and if:

1. The initial workup does not show biological perturbations or ENMG findings excluding SNN and
2. The patient has one of the following disorders: onconeural antibodies or a cancer within 5 years (Graus *et al.*, 2004), cisplatin treatment, Sjögren's syndrome (Vitali *et al.*, 2002).
3. Or MRI shows high signal in the posterior column of the spinal cord

C A diagnosis of SNN is definite if dorsal root ganglia degeneration is pathologically demonstrated although dorsal root ganglia biopsy is not recommended.

European Journal of Neurology 2010, 17: 356–363 doi:10.1111/j.1468-1331.2009.02930.x

EFNS TASK FORCE/CME ARTICLE

European Federation of Neurological Societies/Peripheral Nerve Society Guideline on management of chronic inflammatory demyelinating polyradiculoneuropathy: Report of a joint task force of the European Federation of Neurological Societies and the Peripheral Nerve Society — First Revision

Members of the Task Force: P. Y. K. Van den Bergh[a], R. D. M. Hadden[b], P. Bouche[c], D. R. Cornblath[d], A. Hahn[e], I. Illa[f], C. L. Koski[g], J.-M. Léger[h], E. Nobile-Orazio[i], J. Pollard[j], C. Sommer[k], P. A. van Doorn[l] and I. N. van Schaik[m]

Clinique	a+b	a+b	a+b
EMG	Définie	Probable	Possible
Supportive	0	1	2

Clinique (a ou b)

Forme typique (a)

déficit ss-moteur > 2mois, proximo-distal, diminution ou abolition des ROT

Formes atypiques (b)

distal, asymétrique, focal, sensitif pure, moteur pure

Exclure

lymphome, amylose, POEMS…CMT, HNPP, Lyme.

Critères de démyélinisations sur la neurographie motrice : aucun

Critères complémentaires

- PL +
- IRM +
- Anomalies sensitives ≥ 1 nerf:

médian ou radial anormal avec sural normal, ou VCS < 80%, ou PES anormal

- Réponse aux IgIV
- Biopsie nerveuse (ME ou teasing)

A ce stade, ganglionopathie possible vs CISP (chronic inflammatory sensory polyradiculoneuropathy) ?, Quels autres ECP demandez vous ?


IRM médullaire avec et sans gadolinium


Triple stimulation pour le médian et l'ulnaire


PES


Anti onconeuronaux + Pet scanner, + examen gynécologique


anti SSA, SSB, BGSA

ECPs

- anti YO positif

(immunologie, infectieux, BGSA, PL normaux)

- Pet scan:

fixation > quandrant supero externe sein droit

> 2 ganglions axillaires

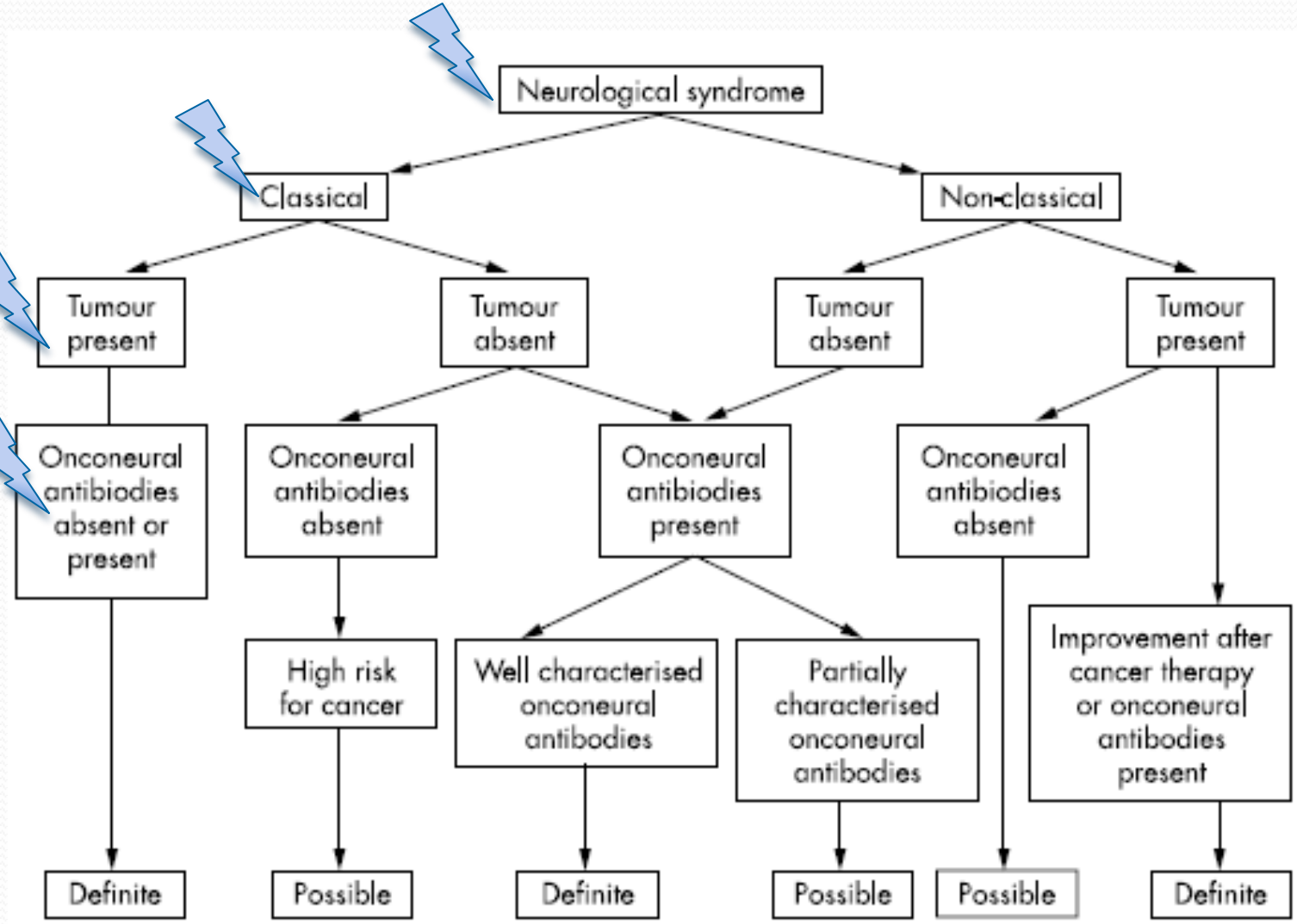
- Carcinome canalaire infiltrant grade SBR3,
  pT1b(3)N2,
  RH-,
  HER2 score 3+

Recommended diagnostic criteria for paraneoplastic neurological syndromes

F Graus, J Y Delattre, J C Antoine, J Dalmau, B Giometto, W Grisold, J Honnorat, P Sillevis Smitt, Ch Vedeler, J J G M Verschuuren, A Vincent, R Voltz, for the Paraneoplastic Neurological Syndrome Euronetwork

See Editorial Commentary, p 1090

J Neurol Neurosurg Psychiatry 2004;75:1135–1140. doi: 10.1136/jnnp.2003.034447



CLINICAL FEATURES AND PATHOPHYSIOLOGICAL BASIS OF SENSORY NEURONOPATHIES (GANGLIONOPATHIES)

THIERRY KUNTZER, MD,[1] JEAN-CHRISTOPHE ANTOINE, MD,[2] and ANDREAS J. STECK, MD[3]

[1] Department of Neurology, Centre Hospitalier Universitaire Vaudois, Lausanne, Switzerland
[2] Department of Neurology, Hôpital Bellevue, Saint-Etienne, France
[3] Department of Neurology, University Hospital Basel, Petersgraben 4, CH-4031 Basel, Switzerland

Accepted 13 April 2004

Table 1. Comparison of sensory neuronopathies.

Entity	Onset	Distinctive clinical signs	Distinctive ancillary tests	Response to treatment
Paraneoplastic subacute sensory neuronopathy	Days to weeks; progressive; multifocal or asymmetrical	Pseudoathetoid movements or unsteady gait; autonomic signs; association with central involvement	Anti-Hu antibodies	Disappointing; early diagnosis of cancer gives the best chance of stabilizing
Dysimmune sensory ganglionopathies	Abrupt to indolent progression; asymmetrical	Sicca syndrome; pseudoathetoid movements or unsteady gait; autonomic signs	Rheumatoid factor; antinuclear or anti-SSA/SSB antibodies; lip biopsy	Improvement from courses of IVIg may occur
Acute autoimmune ataxic syndromes	Days to 4 weeks	Areflexia, ophthalmoplegia, ataxia; may be drowsiness or limb weakness	IgG anti-GQ1b antibody	IVIg or plasmapheresis
Chemotherapy-induced peripheral neurotoxicity	First signs appear ~1 month after therapy	Large-fiber sensory involvement; gait disturbance may occur	None	Supportive
Pyridoxine-induced sensory neuronopathy	Subacute	Large-fiber sensory involvement; gait disturbance may occur	None	Stop intake of pyridoxine
Friedreich's ataxia	Chronic, over years	Large-fiber sensory involvement and gait disturbance	Genetic testing	Supportive and idedenone

Cas clinique n°2

Patiente de 31 ans, tabac 10 PA

HDM: engourdissement des 4 extrémités
remontant le long des 4 membres
intéressant ensuite le tronc, et la région péribuccale
instabilité à la marche
installation en 4 jours

Clinique: force normale, AROT généralisé,
apallesthésie des 4 membres,
ataxie des 4 membres, Romberg +, tandem impossible

Absence d'autre signe neurologique/ extra neurologique

Video retirée

Examens complémentaires

- ENMG à J5

neurographie motrice : normale

neurographie sensitive : normale

myographie: normale

- IRM cérébrale et médullaire normales

- PL normale

Quelles hypothèses diagnostiques vous semblent possibles ?

PRNA type SGB ataxique (anti-GD1b)

B Syndrome de Miller Fisher (anti-GQ1b)

Ganglionopathie avant la dégénérescence Wallérienne

D Multinévrite sensitive

PRNC sensitive à début aigu pré ganglionnaire

Quelles explorations proposeriez vous ?


PES

B Biopsie neuro-musculaire


Anti-gangliosides


Refaire une deuxième PL


Refaire un deuxième ENMG

Examens complémentaires

- ENMG à J15

<u>neurographie motrice :</u> normale

<u>neurographie sensitive des 4 membres:</u>

médian, ulnaire, radial, sural, musculo-cutané = 0 µV

<u>myographie:</u> normale

- PL n°2 normale

- Anti-gangliosides négatifs
- Anti-neuronaux négatifs
- Facteur nucléaires 1/1280 de type anti-nucéosome
- BGSA: focus 1

- Pet Scanner normal
- Ig IV 3 cures => aucune efficacité

Quelle hypothèse diagnostique retenez vous?

A PRNA type SGB ataxique (anti-GD1b)

B Syndrome de Miller Fisher (anti-GQ1b)

Ganglionopathie **avec** dégénérescence Wallérienne

D Multinévrite sensitive

E PRNC sensitive à début aigu pré ganglionnaire

The pattern and diagnostic criteria of sensory neuronopathy: a case–control study

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If >6.5, a diagnosis of SNN is possible	Total	

B A diagnosis of SNN is probable if the patient's score is >6.5 and if:

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3. Or MRI shows high signal in the posterior column of the spinal cord

Cas clinique n°3

Patiente de 65 ans,

Depuis 6 ans

Sensation brulure des 4 extrémités, pieds > main, permanant

ATCD anneau gastrique il y a 16 ans, xérostomie

Cliniquement: allodynie des pieds, diminution de la sensation chaud / froid sous les malléoles, reste RAS

IRM cérébrale et médullaire normales

ENMG normal

Hémogramme, bilan rénal + hépatique normaux,

HGPO négative, TSH, B12, folate normaux

EPP: hypergamma polyclonale;

FR +, SSA +,

BGSA Focus 1,

Dans ce contexte de Sjögren, quelle pathologie du SNP suspectez vous?

A PRNC sensitive pré ganglionnaire

B Ganglionopathie

Neuropathie des petites fibres

D Multinévrite sensitive

E Syndrome des jambes sans repos

Les petites fibres….

Fibres A delta:

- froid
- douleur
- SNA pré GG

Fibres C:

- chaud (froid)
- douleur (seuil haut): mécanique, thermique, chimique,
- SNA post GG

Quelles explorations complémentaires demandez vous ?

Etude quantifiée de la sensibilité thermique
(chaud: fibre C, froid: fibre A delta)

B Biopsie du nerf sural droit

La recherche d'une dysautonomie (Ewing)

Biopsie de peau (cheville + hanche)

PES laser (fibre A delta)

Diagnosis of small fiber neuropathy: A comparative study of five neurophysiological tests

Diagnostic des neuropathies des petites fibres : une étude comparative de 5 tests neurophysiologiques

J.-P. Lefaucheur[a,b,h,*], A. Wahab[h], V. Planté-Bordeneuve[b,c], D. Sène[d], I. Ménard-Lefaucheur[h], D. Rouie[h], D. Tebbal[h], H. Salhi[a,c], A. Créange[a,c], H. Zouari[a,e], S. Ng Wing Tin[a,f,g]

Neurophysiologie Clinique/Clinical Neurophysiology (2015) 45, 445–455

Sensibilité :

- **PES laser 79% (A delta)**
- **Sudoscan 61% (C sympathique)**
- **Warm Detection Treshold 55% (C)**
- RCS 41% (B sympathique)
- Cold Detection treshold 32% (A delta)

La combinaison des 3 premiers examens augmente la sensibilité

Les explorations demandées…

- PES laser altération des fibres A delta

-Biopsie de peau

Densité des fibres nerveuses intra-épidermique:

Effondrée en distalité

Abaissée en proximal

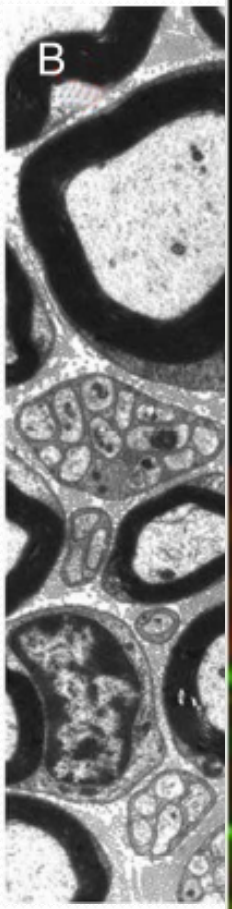
Critères diagnostiques d'une neuropathie des petites fibres

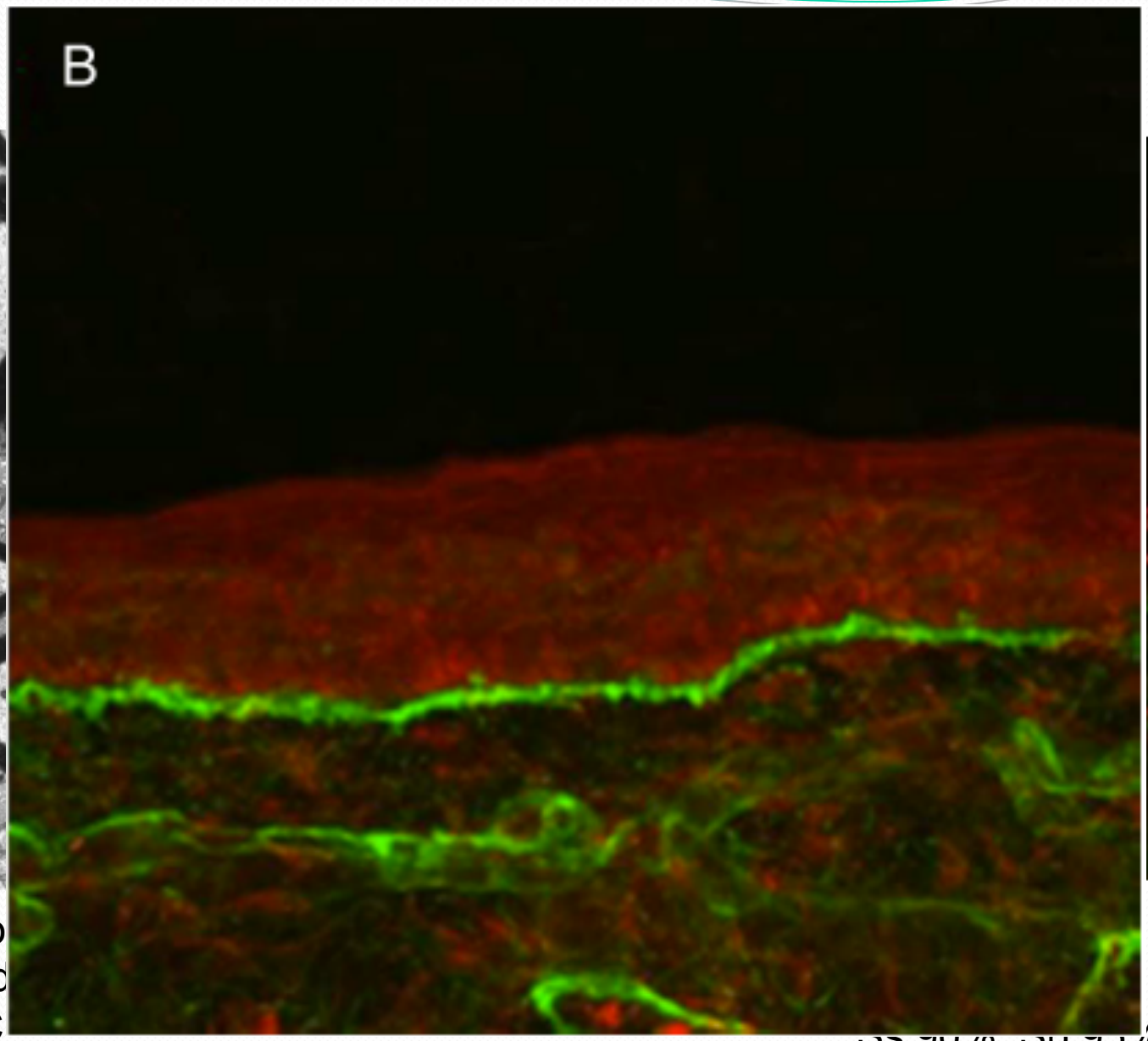
- Possible—length-dependent symptoms and/or clinical signs (pinprick and thermal sensory loss and/or allodynia/hyperalgesia).
- Probable—length-dependent symptoms, clinical signs of small fibre damage and normal nerve conduction studies.
- Definite—length-dependent symptoms, clinical signs of small fibre damage, normal nerve conduction studies, and altered intra-epidermal nerve fibre density at the ankle and/or abnormal quantitative sensory testing of thermal thresholds at the foot.

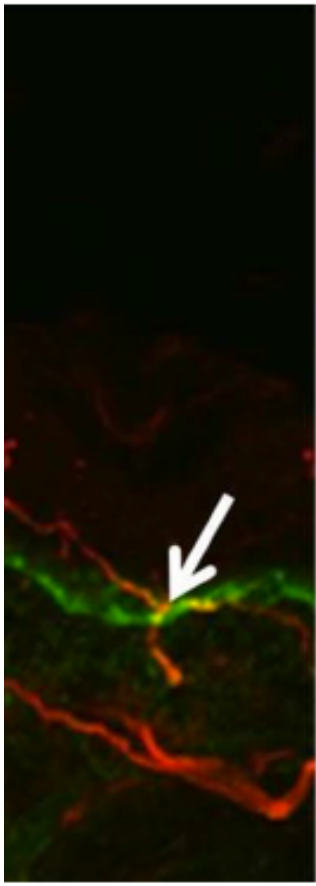
En fait: signes et/ou symptômes longueur dépendant ou non
ENMG normal
Biopsie de peau pathologique

Patient VIH avec burning feet syndrome

Sujet







Bleu : gro

Violet: A d

Rouge: C

vert

Ss 90 %; Sp 95 %

Causes des neuropathies des petites fibres pour la plupart => grosses fibres

Table 1 Causes of small fibre neuropathy

Primary	Secondary
Idiopathic	Metabolic
▶ Idiopathic small fibre neuropathy	▶ Impaired glucose tolerance
▶ Burning mouth syndrome	▶ Diabetes mellitus
	▶ Rapid glycaemic control
	▶ Vitamin B12 deficiency
	▶ Dyslipidaemia
	▶ Hypothyroidism
	▶ Chronic kidney disease
Hereditary/genetic	Infections
▶ $Na_v1.7$ mutations	▶ HIV
▶ $Na_v1.8$ mutations	▶ Hepatitis C
▶ Familial amyloid polyneuropathy	▶ Influenza
	Toxins and drugs
▶ Fabry's disease	▶ Anti-retrovirals
▶ Tangier's disease	▶ Antibiotics—metronidazole, nitrofurantoin, linezolid
	▶ Chemotherapy—bortezomib
	▶ Flecainide
	▶ Statin
	▶ Alcohol
	▶ Vitamin B6 toxicity

Souvent non longueur dpt

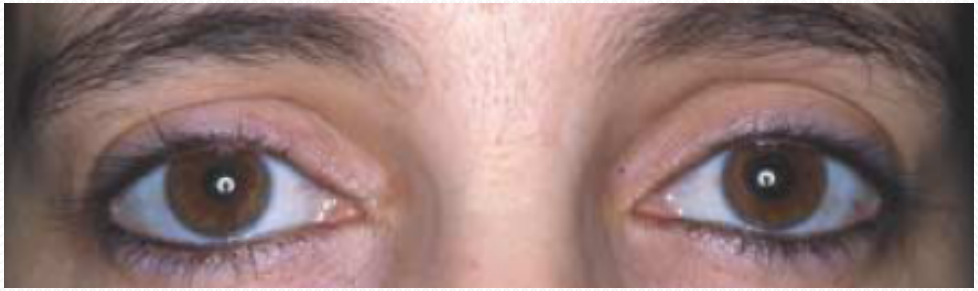
Immune mediated

- Coeliac disease
- Sarcoidosis
- Sjögren's syndrome
- Rheumatoid arthritis
- Systemic lupus erythematosus
- Vasculitis
- Inflammatory bowel disease
- Paraneoplastic
- Monoclonal gammopathy/amyloid

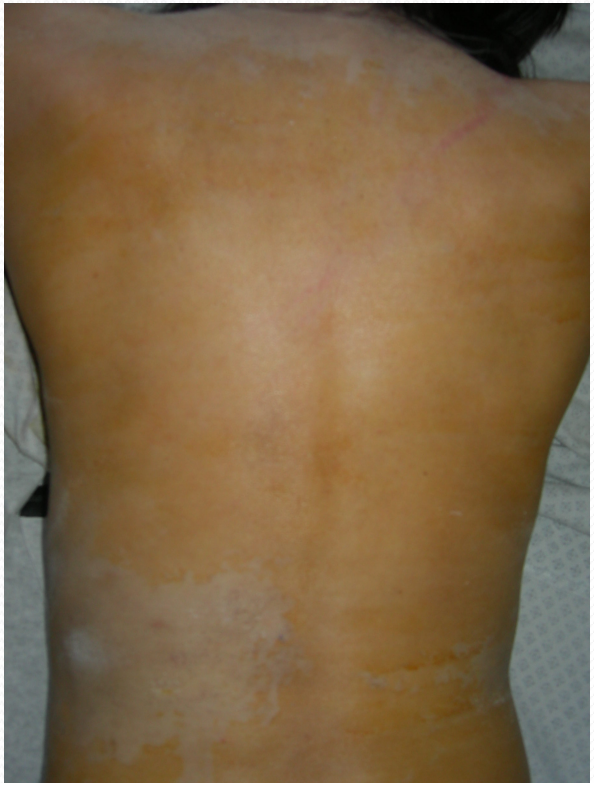
HSAN
Lipome de la queue de cheval
Lèpre

Cas clinique n°4 (cas de la littérature)

- Patiente de 32 ans, Gougerot depuis 2 ans,



Hyporéflexie



Ross syndrome, an entity included within the spectrum of partial disautonomic syndromes

M Ballestero-Díez,*† I García-Río,† E Daudén,† MJ Corrales-Arroyo,‡ A García-Díez†

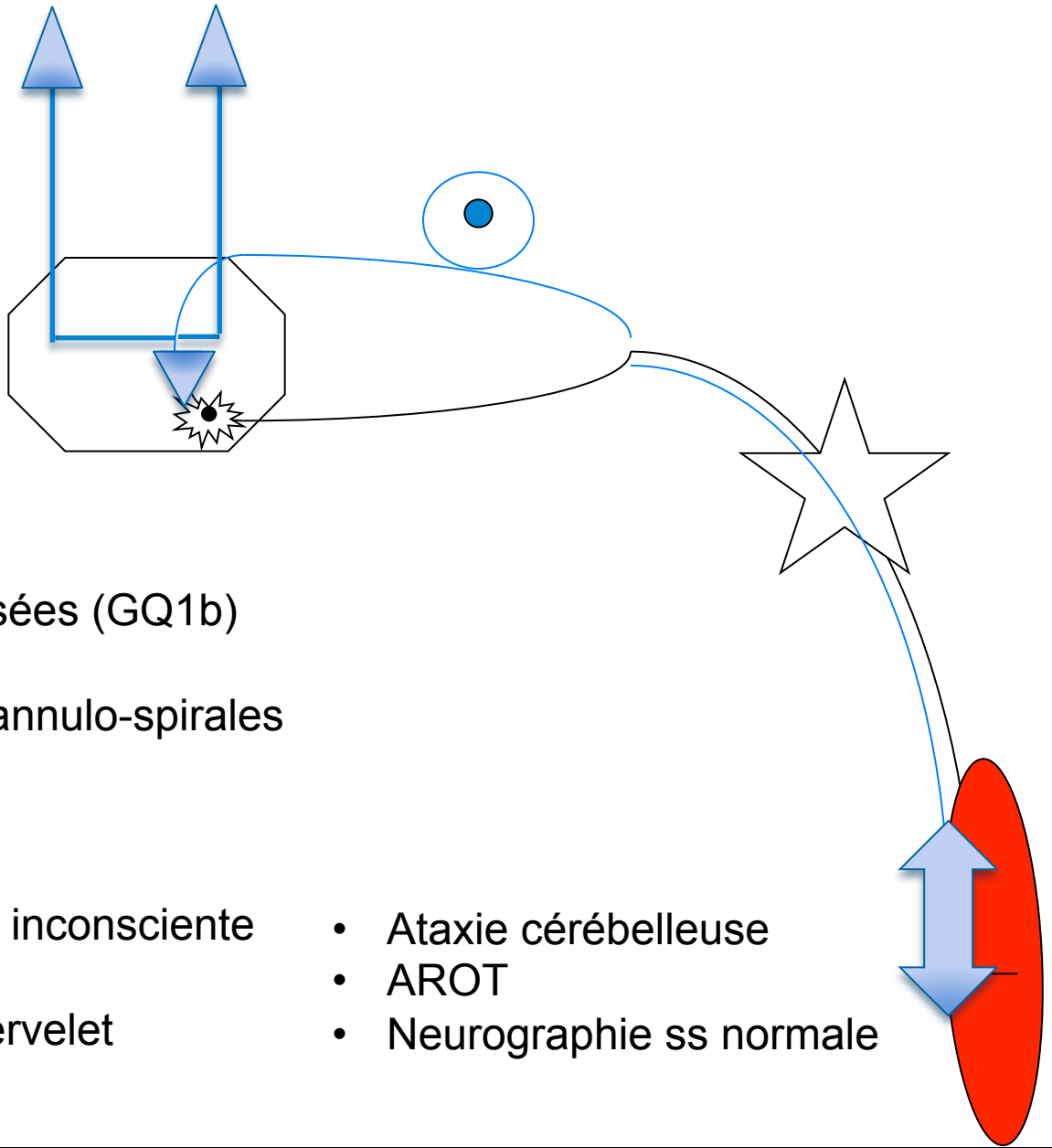
Les maladies dysimmunes peuvent donner des neuropathies sensitives à neurographie sensitive normale ….

- 3 neurones sensitifs (A alpha, A beta-gamma, A delta C)

Sur la neurographie conventionnelle :

- 1 sur trois est détectable
- seulement en distalité

Trois neurones sensitifs: 1. A alpha



Fibres myélinisées (GQ1b)

Terminaisons annulo-spirales

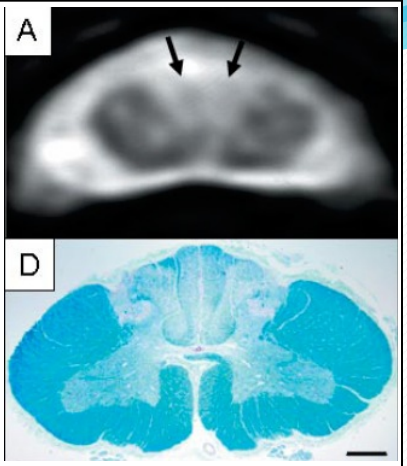
ROT

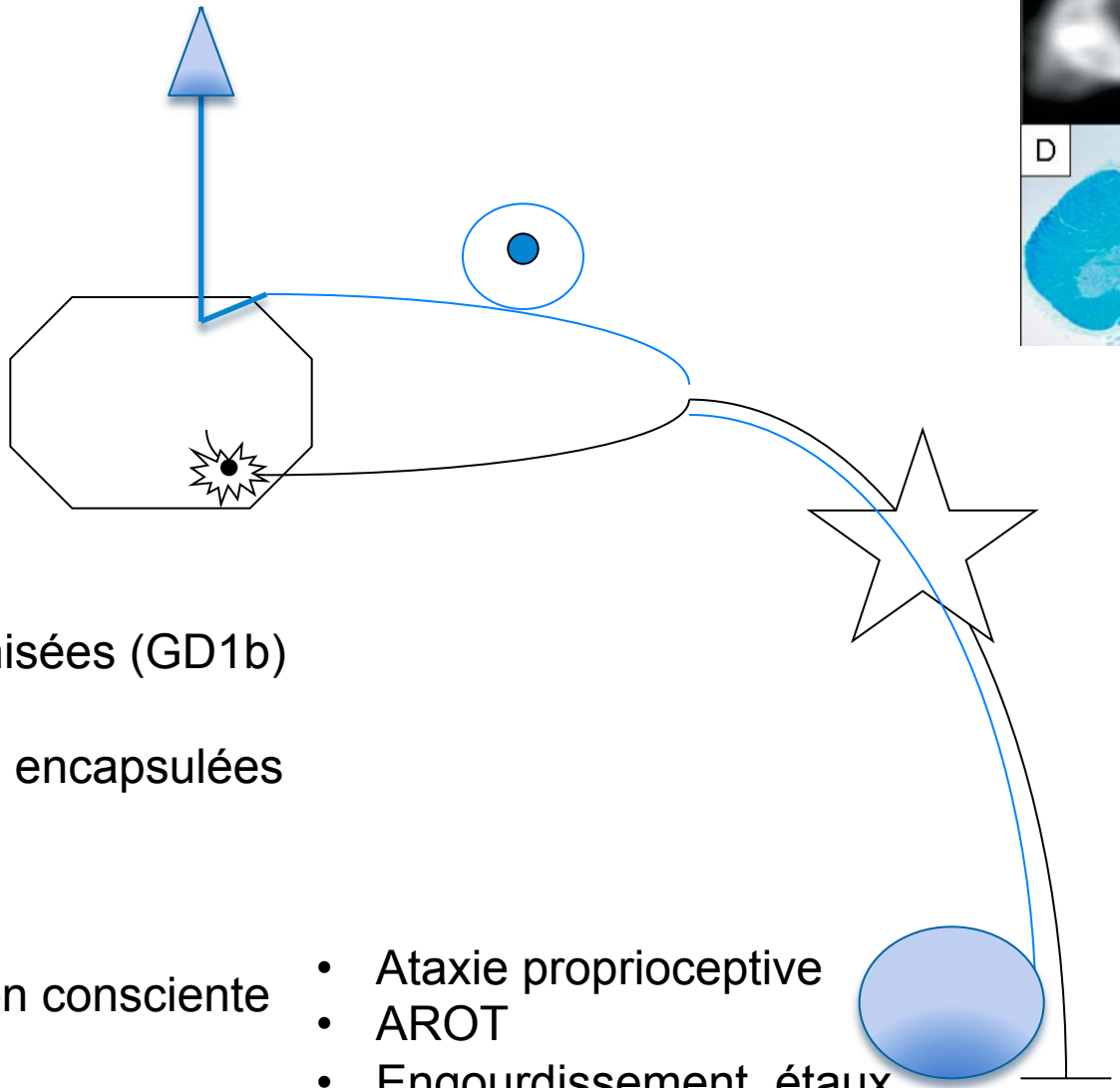
Proprioception inconsciente

Informent le cervelet

- Ataxie cérébelleuse
- AROT
- Neurographie ss normale

Trois neurones sensitifs: 2. A bêta/gamma





Fibres myélinisées (GD1b)

Terminaisons encapsulées

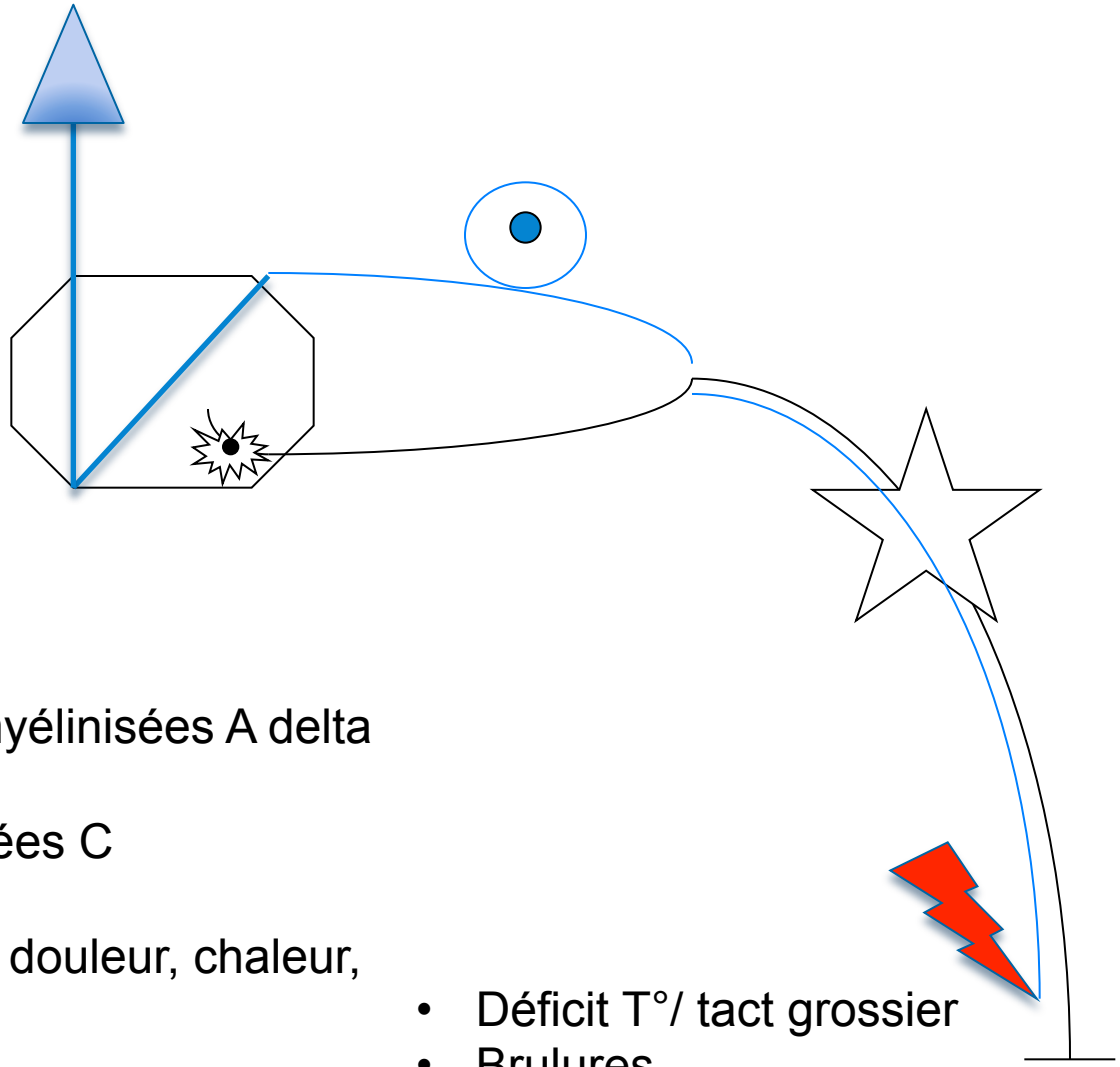
ROT ?

Proprioception consciente

=> aire somesthésique I

- Ataxie proprioceptive
- AROT
- Engourdissement, étaux…
- Diminution de la sensibilité épicritique…
- Neurographie sensitive altérée sauf si

Trois neurones sensitifs: 3. A delta/C



Petites fibres:

Faiblement myélinisées A delta

Non myélinisées C

Tact grossier, douleur, chaleur,

SNA

- Déficit T°/ tact grossier
- Brulures
- dysSNA
- ROT présents, Neurographie sensitive normale

Les neuropathies dysimmunitaires (partie 2)

Cours DES
Guillaume Taieb, Neurologie,
Gui de Chauliac, Montpellier

Cas clinique n°5

57 ans, ATCD néant

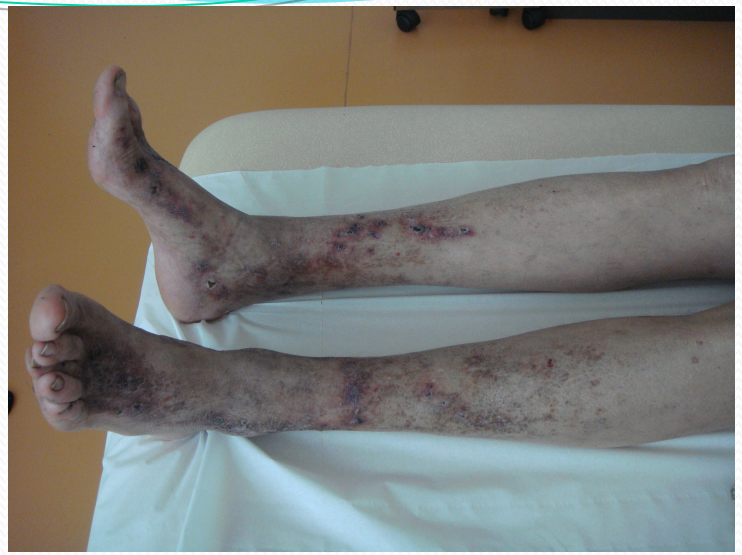
- 2009

- douleurs sévères plante des pieds => 2/3 mollet (droite>G)
- brulures, constantes
- installation < 2 mois

+ poussées

- lésions des jambes
- erythémato-papulleuses violacées
- ulcérées, crouteuses d'âges différents
- fréquences 3/ an
- durée 1 mois
- FF non

Cliniquement



marche pointes/ talons RAS

tandem instable

Romberg: oscillations YF

amyotrophie des muscles pédieux,

extenseurs communs des orteils 4/5,

hyporéflexie achilléenne

apallesthésie des chevilles, hypopallesthésie des chevilles

ENMG

- **neurographie motrice** :

amplitude SPE X2 recueil pédieux effondrée (Droit << Gauche)
amplitude SPI X2 effondrée
SPE recueil TAX2 normaux
Médian, ulnaire X2 normaux
Latence des ondes F normale SPE-TA et MSup

- **neurographie sensitive** :

Sural, MSC X2 = 0
Médian, ulnaire, radial X2 normaux

- **Sur la myographie** :

AS: pédieux, court fléchisseur du GO, TS
Tracé SA: pédieux, court fléchisseur du GO, TS
Autres muscles: TA, vaste interne, premier interosseux normaux

Diagnostic

- Topographie PNP asymétrique ou MNM
- Installation subaiguë
- Clinique sensitive (SF> GF) > motrice
- ENMG axonal
- Evolution évolutif
- Clés peau

Hypothèse ?

Bilan

standard normal
auto immun négatif (connectivite, BBS, vascularite)
Paranéoplasique, GAD, Anti transglutaminase négatif
infectieux négatif
métabolique/ vitaminique normal
Hématologie/ thrombophilie acquise/ congénital

- BGSA
- PL
- Pet
- Echo doppler veineux artériel

Quelle exploration proposez vous ?

Biopsie cutanée

au contact d'une ulcération superficielle, une importante hyperplasie vasculaire dermique associée à une fibrose et une sidérose, l'ensemble réalisant un aspect morphologique proche de lésions d'acro-angio-dermatite. On ne voit pas d'authentique image de vascularite des vaisseaux dermiques dans les limites de ce prélèvement.

Que faites vous ?

Biospie neuro musculaire

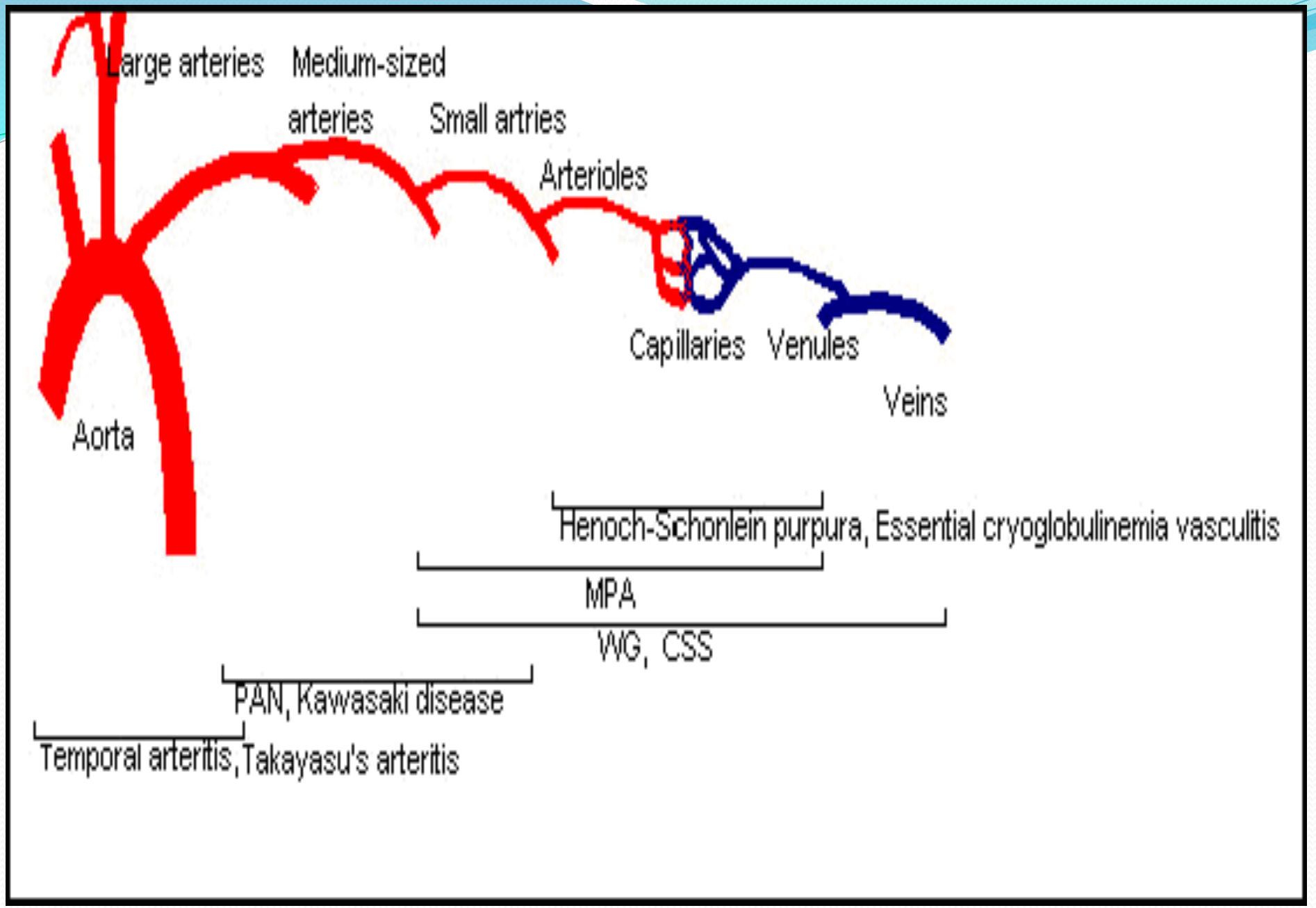
Pourquoi ?

- mécanisme
- évolutivité
- étiologie

BIOPSIE DU NERF SURAL GAUCHE

Sur les diverses recoupes réalisées, on observe des lésions de neuropathie axonale sévère associées à des lésions de vascularite mises en évidence par l'infiltrat mononucléé lymphocytaire autour des vaisseaux. On notera également que certains vaisseaux comportent une nécrose fibrinoïde de leur paroi associée à la formation de micro-thrombi.

CONCLUSION :
Biopsie nerveuse mettant en évidence une neuropathie axonale sévère associée à des lésions de vascularite.



Classification de Chapel Hill 1994

Journal of the Peripheral Nervous System 15:176–184 (2010)

PNS NSVN GUIDELINE

Peripheral Nerve Society Guideline* on the classification, diagnosis, investigation, and immunosuppressive therapy of non-systemic vasculitic neuropathy: executive summary

Michael P. Collins (Chair), USA; P. James B. Dyck, USA; Gary S. Gronseth, USA; Loïc Guillevin, France; Robert D. M. Hadden, UK; Dieter Heuss, Germany; Jean-Marc Léger, France; *N.C.* Notermans, The Netherlands; John D. Pollard, Australia; Gérard Said, France; Gen Sobue, Japan; *A.F.J.E.* Vrancken, The Netherlands; John T. Kissel (Co-Chair), USA.

Table 1. Classification of vasculitides associated with neuropathy.

I. Primary systemic vasculitides
  1. Predominantly small vessel vasculitis
    a. Microscopic polyangiitis*
    b. Churg–Strauss syndrome*
    c. Wegener's granulomatosis*
    d. Essential mixed cryoglobulinemia (non-HCV)
    e. Henoch–Schönlein purpura
  2. Predominantly medium vessel vasculitis
    a. Polyarteritis nodosa (PAN)
  3. Predominantly large vessel vasculitis
    a. Giant cell arteritis

II. Secondary systemic vasculitides associated with one of the following
  1. Connective tissue diseases
    a. Rheumatoid arthritis
    b. Systemic lupus erythematosus
    c. Sjögren's syndrome
    d. Systemic sclerosis
    e. Dermatomyositis
    f. Mixed connective tissue disease
  2. Sarcoidosis
  3. Behcet's disease
  4. Infection (such as HBV, HCV, HIV, CMV, leprosy, Lyme disease, HTLV-I)
  5. Drugs
  6. Malignancy
  7. Inflammatory bowel disease
  8. Hypocomplementemic urticarial vasculitis syndrome

III. Non-systemic/localized vasculitis
  1. Non-systemic vasculitic neuropathy (includes non-diabetic radiculoplexus neuropathy and some cases of Wartenberg's migrant sensory neuritis)
  2. Diabetic radiculoplexus neuropathy
  3. Localized cutaneous/neuropathic vasculitis
    a. Cutaneous PAN
    b. Others

*Anti-neutrophil cytoplasmic antibody (ANCA)-associated vasculitides.
CMV, cytomegalovirus; HBV, hepatitis B virus; HCV, hepatitis C virus; HIV, human immunodeficiency virus; HTLV, human T-lymphotropic virus.

Table 2. Diagnostic criteria for pathologically definite vasculitic neuropathy*.

I. *Active lesion:* Nerve biopsy showing collection of inflammatory cells in vessel wall AND one or more signs of acute vascular damage:
   1. Fibrinoid necrosis;
   2. Loss/disruption of endothelium;
   3. Loss/fragmentation of internal elastic lamina;
   4. Loss/fragmentation/separation of smooth muscle cells in media (can be highlighted with anti-smooth muscle actin staining);
   5. Acute thrombosis;
   6. Vascular/perivascular hemorrhage; OR
   7. Leukocytoclasia.

II. *Chronic lesion with signs of healing/repair:* Nerve biopsy showing collection of mononuclear inflammatory cells in vessel wall AND one or more signs of chronic vascular damage with repair:
   1. Intimal hyperplasia;
   2. Fibrosis of media;
   3. Adventitial/periadventitial fibrosis; OR
   4. Chronic thrombosis with recanalization.

III. No evidence of another primary disease process that can mimic vasculitis pathologically, such as lymphoma, lyphomatoid granulomatosis, or amyloidosis.

*Presence of a chronic lesion does not exclude active vasculitis (vasculitides are usually segmental and multifocal, producing lesions of different ages in the same tissue or end-organ).

Is it acceptable to not treat NSVN?

Si stable depuis 3 mois

What is first-line immunosuppressive therapy in NSVN?

Bolus MP => forme sévère
CT po 1 mg/kg/j
Sevrage CT: 25 mg à 3 mois
20 mg à 4 mois
10 mg à 6 mois
7.5 à 5 mg pour 6-18 mois

When should combination therapy be used in NSVN?

of the risk for developing transitional-cell carcinoma of the bladder many years after discontinuation of therapy, all patients treated with CYC should undergo screening urinalyses (for non-glomerular hematuria) and urine cytologies every 6 months indefinitely. In patients with a history of CYC-induced cystitis, routine cystoscopy should be considered every 1 or 2 years.

How should the therapeutic response be monitored?

Clinique tous les mois au moins, EMG, bio..

When can patients with NSVN be concluded to be in clinical remission?

no evidence of clinical worsening by any objective measure and some evidence of improvement by at least one objective measure after 6 months of observation (improvement often delayed for several months due to slow pace of axonal regeneration).

What treatment should be administered for patients in probable clinical remission?

the use of maintenance therapy for 18–24 months to reduce relapses. First-line options for remission maintenance are AZA 1.0–2.0 mg/kg/day or MTX 20–25 mg/week.

How should patients refractory to the initial therapy be treated?

In patients not already treated with CYC, CYC should be used. In patients refractory to CYC, the diagnosis should be reconsidered. Patients with

Si déjà sous CYC, faites votre choix inclusion protocole si possible

Quelle prise en charge proposez vous ?

- corticothérapie 18 mois,
- méthotrexate 18 mois,
- sous traitement

- amélioration des douleurs
- Clinique et ENMG stable
- Mais persistance de 3 poussées cutanées modérées/ an

biopsie de peau: vasculopathie livédoïde (vasculopathie thrombosante)

CASE OF THE MONTH

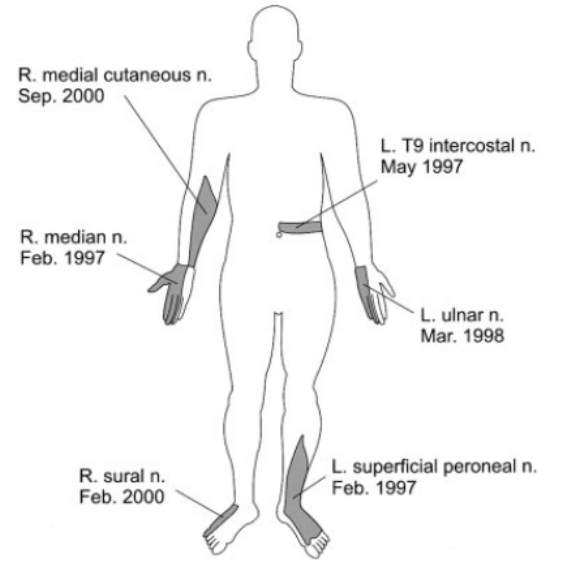
ABSTRACT: Livedoid vasculitis is a chronic dermatological disorder associated with petechiae and recurrent, unusually shaped ulcers that heal to form hyperpigmentated areas and atrophie blanche. This condition is more correctly termed a vasculopathy, rather than a vascultis, and is often associated with an underlying hypercoagulable disorder. We report a patient with livedoid vasculitis and mononeuropathy multiplex. We propose that peripheral nervous system involvement arises from multifocal areas of ischemia due to fibrin and thrombin deposition within both the wall and lumen of vasa nervorum.

Muscle Nerve **27:** 634-639, 2003

MONONEUROPATHY MULTIPLEX IN ASSOCIATION WITH LIVEDOID VASCULITIS

CORY TOTH, MD,[1] MARTIN TROTTER, PhD, MD,[2] ARTHUR CLARK, MD,[2] and DOUGLAS ZOCHODNE, MD[1]

MNM ss
Bilan négatif
BNM: perte axonale chronique
Traitement: corticoides, AA, endoxan, AVK



Causes VL:
- thrombophilies acquises ou congénitales
- poussées cutanées X,
- rare atteinte du SNP, occlusion des vasa nervorum ?

Ischemic Neuropathy Associated with Livedoid Vasculitis

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Background Livedoid vasculitis is a chronic dermatological problem with an unclear etiology. Clinical findings are petechiae with painful ulcers in both lower extremities, which heal to become hyperpigmented and porcelain-white satellite lesions. There are only a few reported cases of livedoid vasculitis presenting in combination with peripheral neuropathy.

Case Report We report the first case of a Korean patient presenting with mononeuritis multiplex combined with livedoid vasculitis, which was confirmed by electrophysiological and pathological studies.

Conclusions Our report supports the possible vaso-occlusive etiology of livedoid vasculitis in multifocal ischemic neuropathy.

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Key Words livedoid vasculitis, livedoid vasculopathy, mononeuritis multiplex, multifocal ischemic neuropathy.

MNM ss > m
Bilan neg
BNM neuropathie ischémique + ILPV
ss Plavix + pentoxifylline

Case of livedoid vasculopathy with peripheral neuropathy successfully treated with low-dose warfarin

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ABSTRACT

We report herein a case of a 28-year-old woman with persistent livedo racemosa and recurrent ulcerations on the lower extremities. The clinical presentation, together with histopathological findings of vascular occlusion without overt vasculitis in the dermis, led to the diagnosis of livedoid vasculopathy. The patient experienced recurrence of ulcerations and developed peripheral neuropathy affecting the distal extremities during the course of treatment with sarpogrelate hydrochloride. The skin lesions and neurological symptoms improved dramatically after adding low-dose warfarin potassium to the treatment regimen. This case suggests that administration of low-dose warfarin is an effective therapy of choice for patients with livedoid vasculopathy.

Key words: livedo racemosa, livedoid vasculopathy, peripheral neuropathy, warfarin.

MNM ss pure (fibulaire sup, sural, ulnaire)
ACL +
BNM non effectuée
AVK ss

Livedoid vasculopathy associated with peripheral neuropathy: a report of two cases*

Vasculopatia livedoide associada a neuropatia periférica: relato de dois casos

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Gabriela Fortes Escobar[1]
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Gabriela Maldonado[3]

DOI: http://dx.doi.org/10.1590/abd1806-4841.20132363

Cas n°1

PNP ss > m

ACL +

BNM neuropathie chronique + ILPV

Traitement ?

Cas n°2

MNM ss et m

Bilan neg

BNM non

Traitement ?

Evolution

- Echec de la réinstauration des corticoïdes
- Essaie des Ig IV echec
- HBPM dose curative : disparition des poussées cutanées, pas de nouvelle poussée neurologique

Neuropathies dysimmunitaires

Groupe A

•Polyradiculonévrites aigües

•Polyradiculonévrites chroniques
(Motrice/ sensitive/ Lewis et Sumner/ distale/ focale)

•Polyneuropathie démyélinisante anti-MAG

•Neuropathie motrice multifocale avec bloc de condution

Groupe B

•Vascularites
•Sarcoïdose
•Connectivite
•Maladies autoimmunes spécifiques
•Paranéoplasiques
•Gammapathies

Groupe C

•Anti-TNF alpha
•INF alpha…

Vascularites

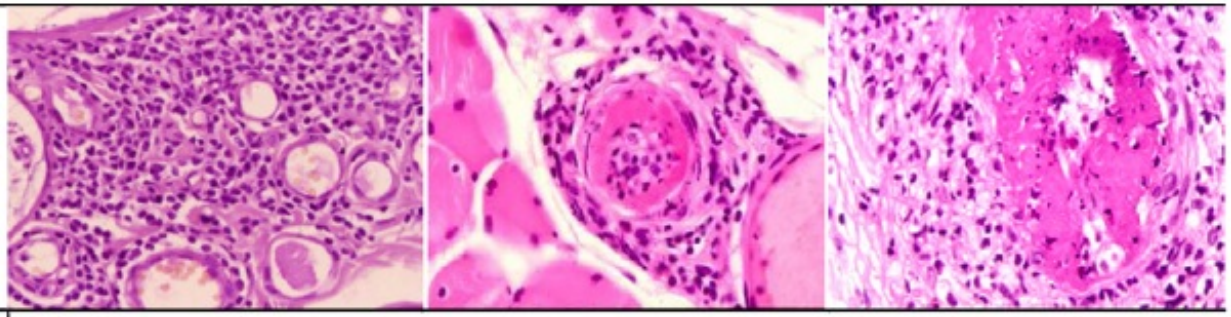
- Multinévrite (SPE, SPI, ulnaire…), PNP asymétrique
- Subaiguë
- Moteur = sensitif, douloureux
- Evolutif
- +/- cutanée, articulaire

- Séronégative: PAN, vascularite restreinte du nerf, BSS
- ANCA, CG dépendante
- Autres SAPL…

Corrélations anatomocliniques des neuropathies périphériques cryoglobulinémiques secondaires à l'hépatite C. Série consécutive de 22 cas

Cryoglobulinemic peripheral neuropathy in hepatitis C virus infection: Clinical and anatomical correlations of 22 cases

G. Taieb [a,*], T. Maisonobe [b], P. Cacoub [c], P. Bouche [b]



Biopsie (x540)			
Résultat	Infiltrat lymphocytaire périvasculaire	Vascularite de petit calibre (<80µm)	Vascularite de moyen calibre (>80µm)
Neuropathie	POLYNEUROPATHIE	MONONEUROPATHIE MULTIPLE	
Handicap	Faible	Modéré	Modérément sévère

Fig. 1 – Topographie et sévérité de la neuropathie en fonction de la biopsie neuromusculaire (résumé schématique), photos de biopsies du nerf sensitif musculocutané, inclus en paraffine, coloration hématéine éosine, coupe transversale. *Neuromuscular biopsy and features of the neuropathy (schematic summary), pictures of superficial peroneal nerve biopsy, paraffin embedded tissue, transversal section, hematein eosin stain.*

Connectivites, sarcoïdose, MICI

Pathologies	V	GG	PRNC	MNM ss-m	PNP ss-m
LEAD	+		+	+ (viteII)	+/ SNA
PR	+			+ (canal/kystes/ viteII)	+/SNA
Goujerot	+	++++	+	+ (viteII,CG)	+/ SMF/ SNA
Connect. Mixtes	+++		+	+	+/SNA
Sclérodermie	+			+ (viteII, X intima)	+
Sarcoïdose	+		+	+	+

Crohn/RCH pdt poussée digestive	PC	B12	+/plexopathie	+	+ (flagyl)