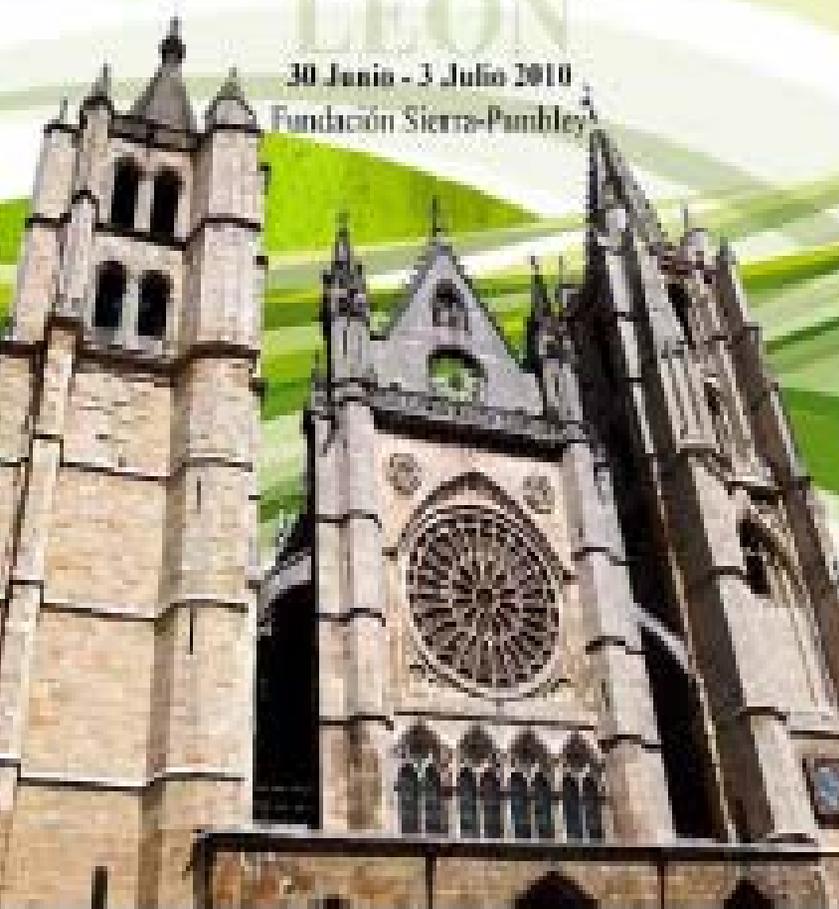


Escuela de Verano de Medicina Interna

LEON

30 Junio - 3 Julio 2010

Fundación Sierra-Pamplona



Conferencia clínico- patológica

Sergio Serrano Villar
Hospital Clínico San Carlos
Madrid

Moderador: Miquel Aranda

CASO 1

- Varón de 57 años
- **MC:** fiebre
- **Antecedentes personales:**
 - Tabaquismo (30 paquetes/año), sin otros hábitos tóxicos.
 - Transportista de materias inflamables
 - Contacto con pájaros
 - Neumopatía por exposición a tóxicos
 - Ulcus duodenal
 - Hipertrofia benigna de próstata
 - No recibe tratamiento

ENFERMEDAD ACTUAL

- Fiebre, náuseas, vómitos, tos seca (tto con jarabe por MAP)
- Cede la fiebre y la clínica respiratoria, aunque persiste la astenia y aumento de sudoración sin predominio horario
- Sigue trabajando

En ocasiones fiebre de 39°+escalofríos y cefalea

Tiempo

-2 meses

ENFERMEDAD ACTUAL

Urgencias Traumatología

- Parestesias a nivel de muslo izquierdo de carácter oscilante.
- EF normal, alta con miorrelajante y complejo vit B

Tiempo

-2 meses

-9 días

ENFERMEDAD ACTUAL

Desconexión transitoria del medio + parestesias mientras conducía el camión

- Disnea progresiva
- Tos
- Fiebre de 4 días de evolución
- Pérdida ponderal de 5 kg en 2 meses.

Tiempo

-2 meses

Ingreso



EXPLORACIÓN FÍSICA

- Taquipnea
- Fiebre 38°
- Subictericia conjuntival
- Hepatomegalia homogénea
- Neurológico normal

Tiempo

-2 meses

Ingreso

PRUEBAS COMPLEMENTARIAS

•**Rx TÓRAX:** patrón alveolointersticial bilateral de predominio apical.

•**SO:** normal

•**EKG:** normal

ANALÍTICA

•Plaquetas 98000

•Urea 87 mg/dL

•Creatinina: 1.72 mg/dL

•Bilirubina total: 1.26 mg/dL

•Triglicéridos: 217 mg/dl

•AST: 57 UI/L

•LDH: 2217 UI/l

•Proteína C reactiva: 144.55 mg/l

•VSG: 61 mm/h

Tiempo

-2 meses

Ingreso

PRUEBAS COMPLEMENTARIAS

Serologias

- IgG *Mycoplasma pneumoniae*
 - IgG VHS tipo II
- }  **débil** **IgM negativas**

- Chlamydia pneumoniae*
- Chlamydia psitacci*
- Legionella*
- Coxiella burnetti* (realizada PCR)
- Leptospira*
- CMV
- VEB
- Adenovirus
- HIV
- Virus hepatitis A, B y C

- Antígeno de *Legionella* en orina: negativo.

PRUEBAS COMPLEMENTARIAS

- PPD (2 determinaciones en 7 días)
- Ziehl-Nielsen esputo
- Cultivo de Löwenstein en orina, sangre y heces
- Hemocultivos y urocultivos

- ECA x2, ADA x2
- Beta2 microglobulina 3.2 mgs/l (N<2.1)

- ANAs,
- ANCAs,
- CEA, CA 19.9, CA 125, PSA

•**GAB**: pH 7.43, pCO₂: 34.4, pO₂: 67, bicarbonato 22.9, EB: -1.6.
Sat: 93.6%.

ENFERMEDAD ACTUAL

TAC torácica

- Patrón alveolointersticial bilateral difuso de predominio superior
- Mínimas adenopatías mediastínicas (paratraqueal derecha baja de 1.5 cm, pre y subcarinales de 1 cm e hiliares bilaterales < 1 cm)

Broncoscopia

- Macroscópicamente normal
- Lavado alveolar: 70% macrófagos, 20% Linfocitos, 10% polimorfonucleares
- Citología: no células malignas.

Tiempo

-2 meses

Ingreso



EVOLUCIÓN

Dx al ingreso: Probable neumonía atípica

- Tratamiento con cefotaxima y claritromicina.
- Evolución clínica tórpida
- Aumento de transaminasas x20
- Se sustituye claritromicina x doxiciclina
- **Eco abdominal:** hepatoesplenomegalia homogénea, sin otras alteraciones.

- Alucinaciones visuales
- Bradipsiquia
- Ataxia
- Paresia braquial izquierda aislada
- Episodios de hemiparesia facio-braquio-crural izquierdas
- Hiperreflexia con clonus y parestesias en hemicuerpo izquierdo
- Dolor neuropático residual
- Disfagia
- 13º día: síndrome confusional agudo y focalidad neurológica transitoria y oscilante de diferentes territorios y disartria.

Tiempo

-2 meses

Ingreso

Día 13º

EVOLUCIÓN

Pruebas Complementarias

•Fondo de ojo y TC craneal

•Punción lumbar:

- Líquido en “cristal de roca”
- Presión normal
- Ligera hiperproteíorraquia (0.87 g/l)
- Glucosa, ADA, LDH, recuento y fórmula
- Micro: cultivos, Ziehl-Nielsen y Löwenstein: negativos.

•Nueva PL:

- 16 leucocitos
 - PMN 46%
 - MN 54%
- Hiperproteíorraquia
- Citología (ambas muestras): no malignidad.

PCR

VHS I, II
VVZ
CMV
VEB

Tiempo

-2 meses

Ingreso

Día 13º

EVOLUCIÓN

Pruebas Complementarias

RMN CRANEAL

- Infartos isquémicos agudos y crónicos en diferentes territorios (tanto profundos como superficiales)
- Arterias cerebrales anteriores medias y posteriores.

EEG

Actividad paroxística sobre el ritmo de base.

• Ecocardiograma: normal (no cardiop estruc embolig)

• Doppler TSA

• Estudio de trombofília

• Aspirado medular MO no infiltrada, inmunofenotipo CD 20+

• Biopsia de cresta iliaca acumulación de linfocitos de aspecto reactivo sin atipia.

Tiempo

-2 meses

Ingreso

Día 13º

RESUMEN

Camionero
Pájaros
Materias inflamables

Fiebre
Parestesias
Disnea

Hepatitis aguda
Hepto-esplenomegalia

MICRO, ANA, ANCA
Marc. Tumorales
NEGATIVOS

**Focalidad neurológica
fluctuante
con infartos cerebrales
múltiples**

ECA
ADA
LDH
B2-MICROBLOBULINA

Meningitis aséptica

Infiltrado
Intersticial
vértices

**Cuadro
subagudo-crónico
y fluctuante**



DIAGNÓSTICO DIFERENCIAL

**ENFERMEDADES
NEUROLÓGICAS
DISEMINADAS
EN TIEMPO
Y ESPACIO**

**AFECTACIÓN PULMONAR
(INFILTRADO INTERSTICIAL
SIN RESPUESTA A ANTIBIÓTICOS)**

ENFERMEDADES NEUROLÓGICAS DISEMINADAS EN TIEMPO Y ESPACIO

- 1. Esclerosis múltiple**
- 2. Infecciosas:**
 - **Neuroborreliosis**
 - **Neurolúes**
 - **VIH**
- 3. Reumatológicas**
 - **Cerebritis lúpica**
 - **Sjögren**
 - **Síndrome antifosfolípido**
- 4. Enfermedades genéticas**
 - **MELAS (Mitochondrial myopathy, encephalopathy, lactic acidosis, and strokelike symptoms)**
 - **Moya-Moya**
 - **Drepanocitosis**
 - **CADASIL (cerebral autosomal dominant arteriopathy with subcortical infarcts and leukoencephalopathy)**
- 5. Cocaína**
- 6. Vasculitis SNC (primaria, secundaria)**
- 7. Linfoma no hodgkiniano**

Vasculitis secundaria del SNC-Causas

INFECCIOSAS

BACTERIANAS

- *M. pneumoniae*
- *C. psittaci*
- *C. burnetii*
- *M. tuberculosis*
- *T. pallidum*

VIRALES

- Varicella zoster
- VIH
- VHC
- VHB
- JC

FÚNGICAS

- *Candida sp*
- *Aspergillus*
- *Histoplasma*
- *Balstomyces*
- *Criptosporidium*
- *Criptococcus*

PARÁSITOS

- Plasmodium
- Taenia
- Schistosoma

AUTOINMUNES

COLAGENOVASCULARES

- LES
- Wegener
- Beçhet
- PAN
- Artritis reumatoide
- Sjögren

MISCELÁNEA

- Sarcoidosis
- Toxinas
 - Efedra
 - Cocaína
 - Tacrolimus
 - Hidralacina
 - Material inflamable?

Neumonía de evolución tórpida

Causas

INFECCIOSAS

BACTERIANAS

- *S. pneumoniae*
- *M. pneumoniae*
- *C. Psittaci*
- *C. burneti*
- *M. tuberculosis*
- ~~• *Nocardia*~~
- *Actynomices*

NO BACTERIANAS

- Virales
- Hongos
- Parásitos
 - *Paragonimus westermani*
 - *Necator americanus*
 - *Ascaris lumbricoides*
 - *Strongyloides stercoralis*

FACTORES HOSPEDADOR

- ~~• Inmunodeficiencias~~

~~COMPLICACIONES~~

- Empiema
- Absceso

NO INFECCIOSAS

TUMORALES

- ~~• CA broncogénico~~
- ~~• CA bronquioalveolar~~
- Linfoma no-hodgkin

INFLAMATORIAS

- Vasculitis sistémicas
 - LES
 - Wegener
 - Hemorragia alveolar

~~• BONO~~

~~• Neumonía eosinofílica~~

~~• SDRA~~

~~• Proteinosis alveolar~~

• Sarcoidosis

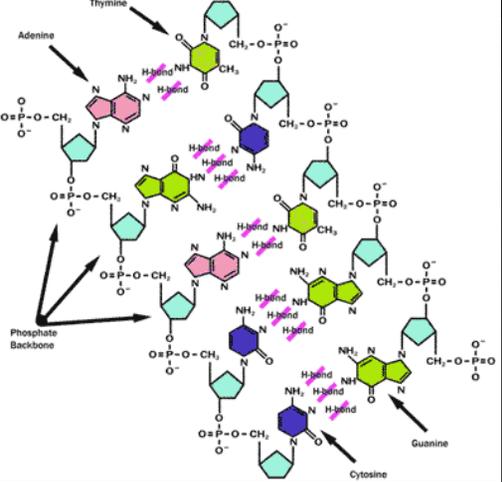
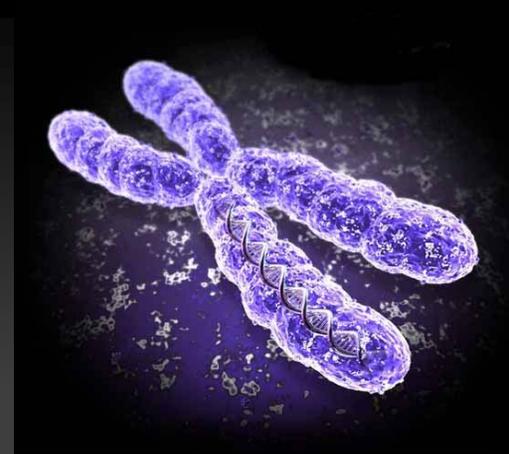
~~INDUCIDO POR DROGAS~~

- Amiodarona
- Metotrexate
- Bleomicina
- Nitrofurantoína

~~EMBOLISMO PULMONAR~~

~~EDEMA HIDROSTÁTICO~~

ENFERMEDADES GENÉTICAS



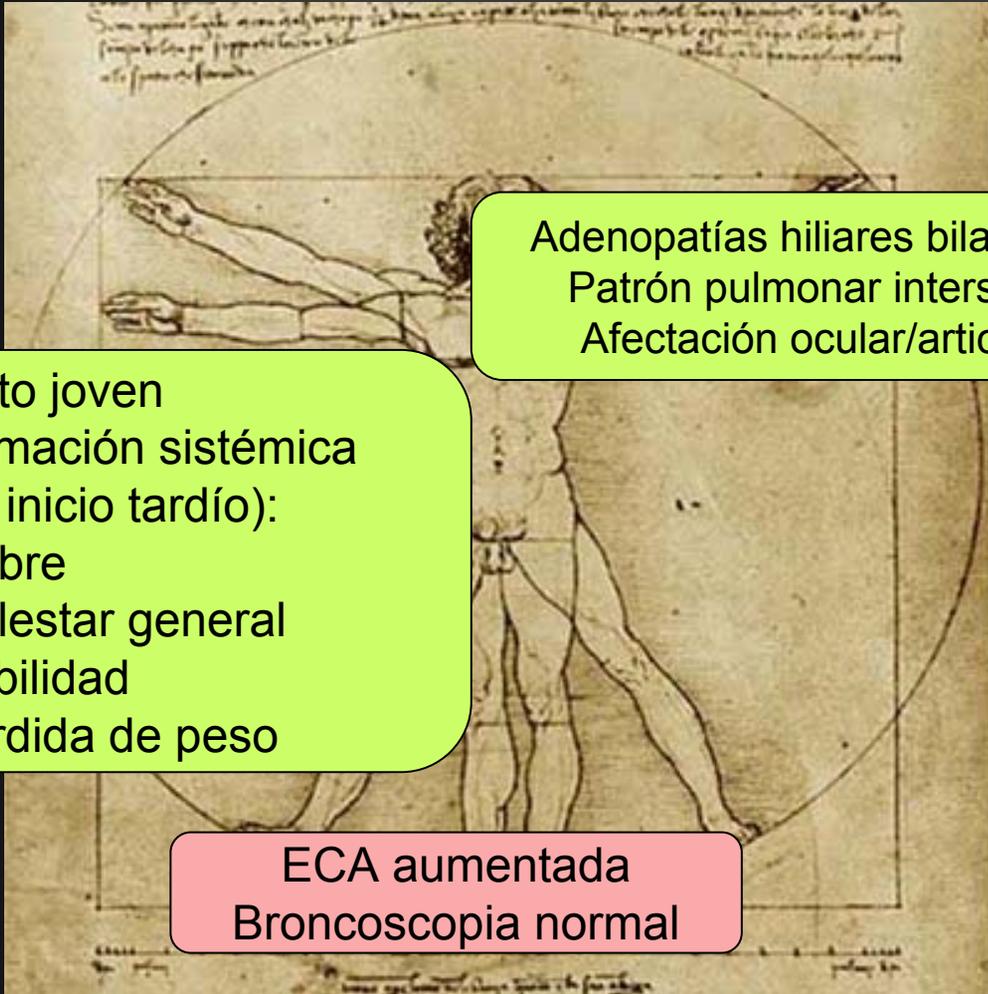
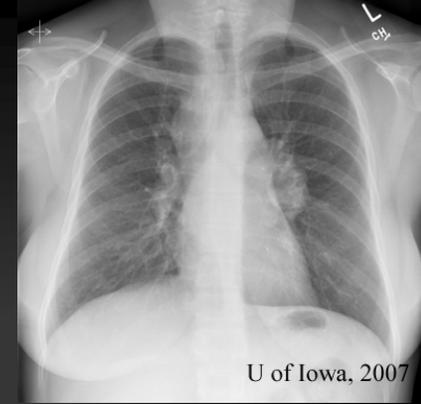
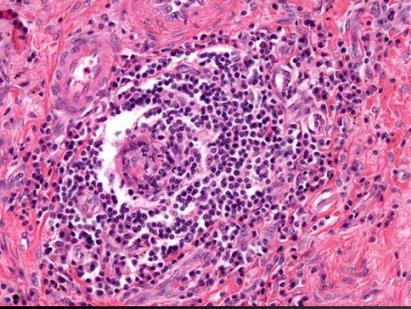
Moya-Moya

- Estenosis-oclusión progresiva de las arterias del polígono de Willis
- Descrita en Japón
- Incidencia 0.3/1000000
- Se asocia a tr. autoinmunes/vasculitis
- Pico edad: 10-14, 40-49
- ACVs recurrentes

CADASIL

- HAD, gen NOTCH3
- 85% Ictus de repetición
- Otros Sx: migrañas con aura, deterioro cognitivo, trastornos psiquiátricos

SARCOIDOSIS

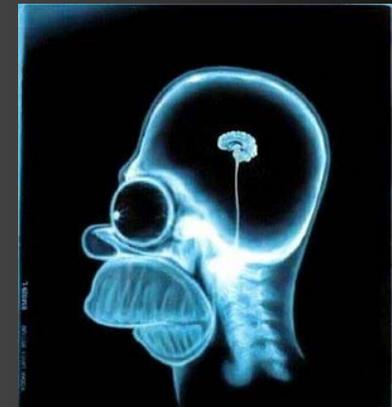


Adenopatías hiliares bilaterales
Patrón pulmonar intersticial
Afectación ocular/articular

Adulto joven
Inflamación sistémica
(+ si inicio tardío):

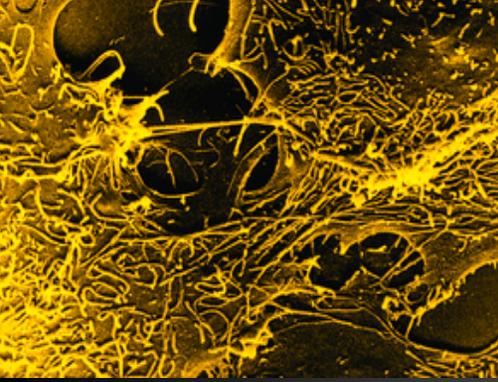
- Fiebre
- Malestar general
- Debilidad
- Pérdida de peso

ECA aumentada
Broncoscopia normal



Neurosarcoidosis (5%)

- Neuropatía craneal
- Disfunción hipotalámica
- Granulomas perivasculares (encefalopatía y vasculopatía)
- Neuropatías periféricas
- Meningitis auda/crónica

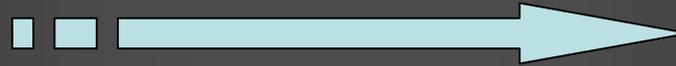


CAUSAS INFECCIOSAS

**Mycoplasma
pneumonie**



Cefotaxima
Claritromicina

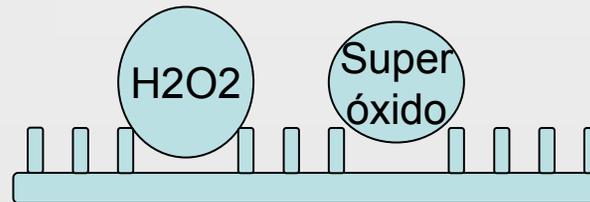


Cefotaxima
Doxiciclina

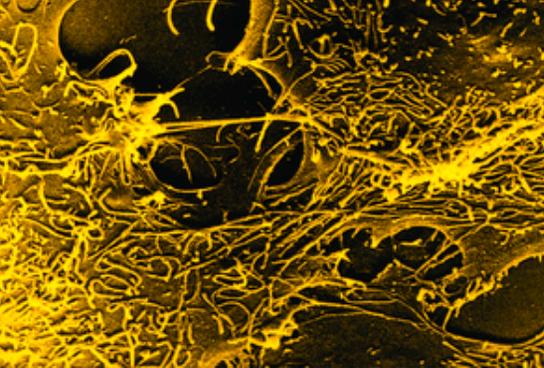
Micro: forma filamentosa la más frecuente, afinidad por epitelio respiratorio.

Patogenia:

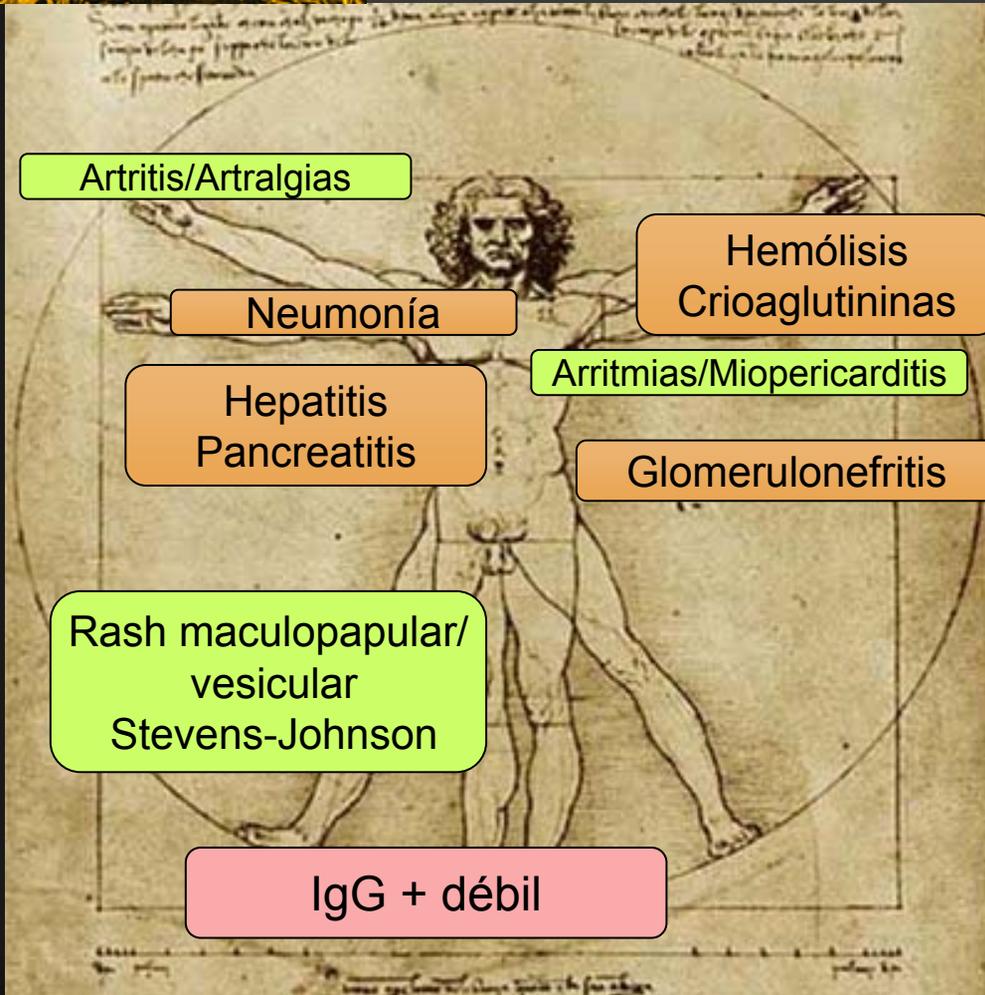
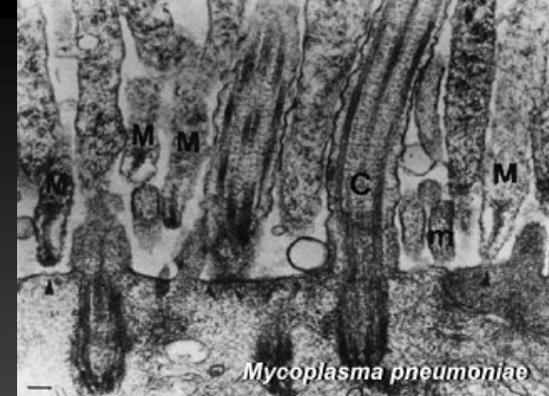
A

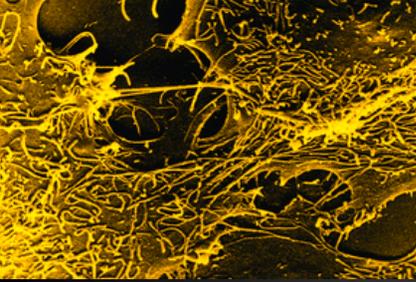


B Autoinmune: Ac frente a Ag glicolípidos de *M. pneumoniae* ejercen como autoAc (reacciones cruzadas con eritrocitos y células SNC).



Mycoplasma pneumoniae





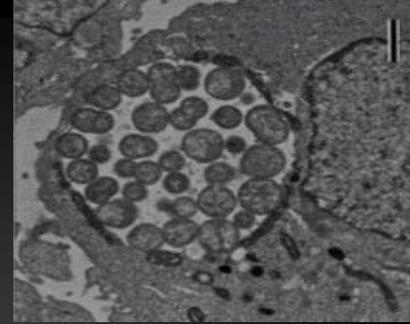
Mycoplasma pneumoniae

Diagnóstico



- 1. Cultivo esputo: 2-3 semanas**
- 2. Serología: necesaria seroconversión**
 - **Inicio 9-10 días tras infección**
 - **Pico a las 3-4 semanas, título x4**
- 3. EIA: S 99%, E 98%**
 - **Similar sensibilidad que PCR, si ha pasado el suficiente tiempo**
- 4. PCR: muy S y E, mejor que serología en fases iniciales, S 48% en primeros**
- 5. Detección de antígeno en esputo (Ag-EIA) días**
 - **+ en los primeros 7 días**

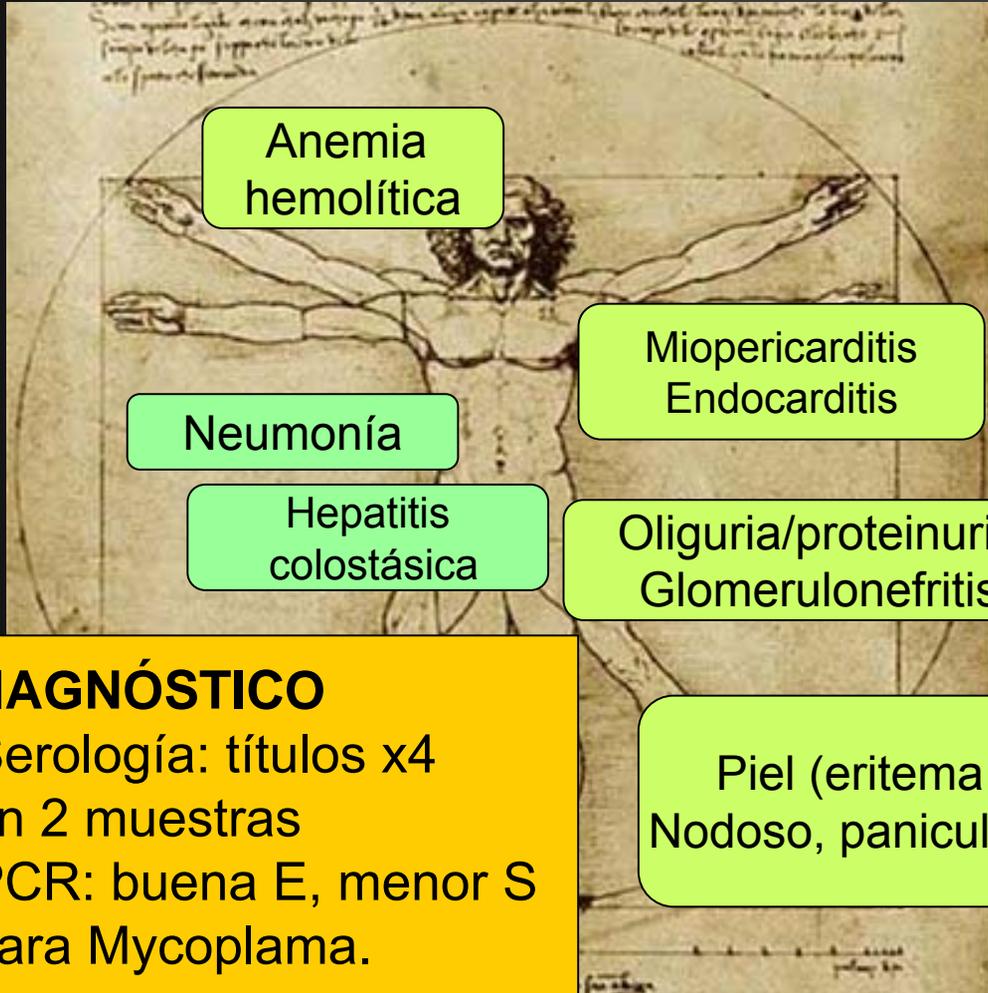
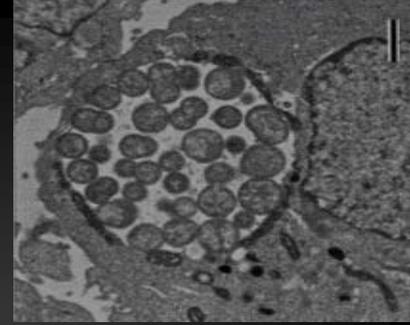
Chlamydia psittaci



Cu
cl



Chlamydia psittaci



Anemia hemolítica

Neumonía

Hepatitis colostásica

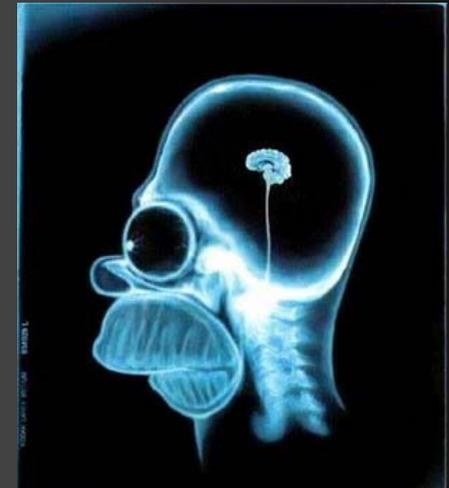
Miopericarditis
Endocarditis

Oliguria/proteinuria
Glomerulonefritis

Piel (eritema
Nodoso, paniculitis)

DIAGNÓSTICO

- Serología: títulos x4 en 2 muestras
- PCR: buena E, menor S para Mycoplasma.



Meningoencefalitis
Mielitis transversa
Parálisis PC
Excepcional vasculitis



Coxiella burnetii

Fiebre Q – Forma aguda



Exantema

Cuadro pseudogripal
Fiebre 2-3 semanas

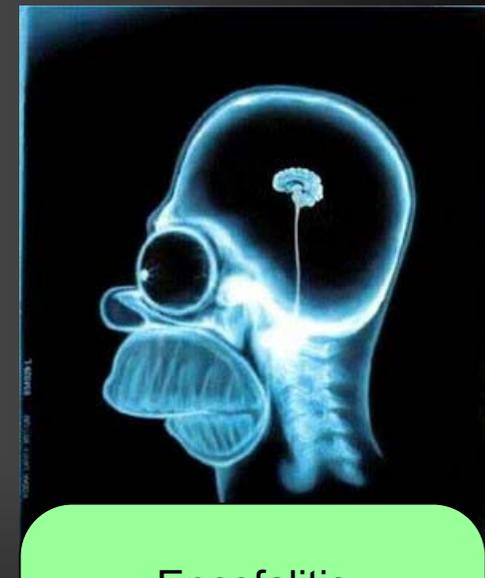
Neumonía

Hepatitis

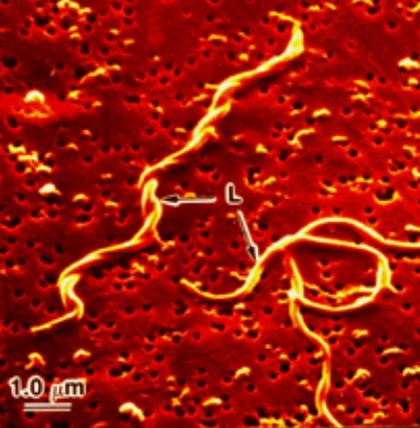
Miopericarditis
Endocarditis
(forma crónica)

DIAGNÓSTICO

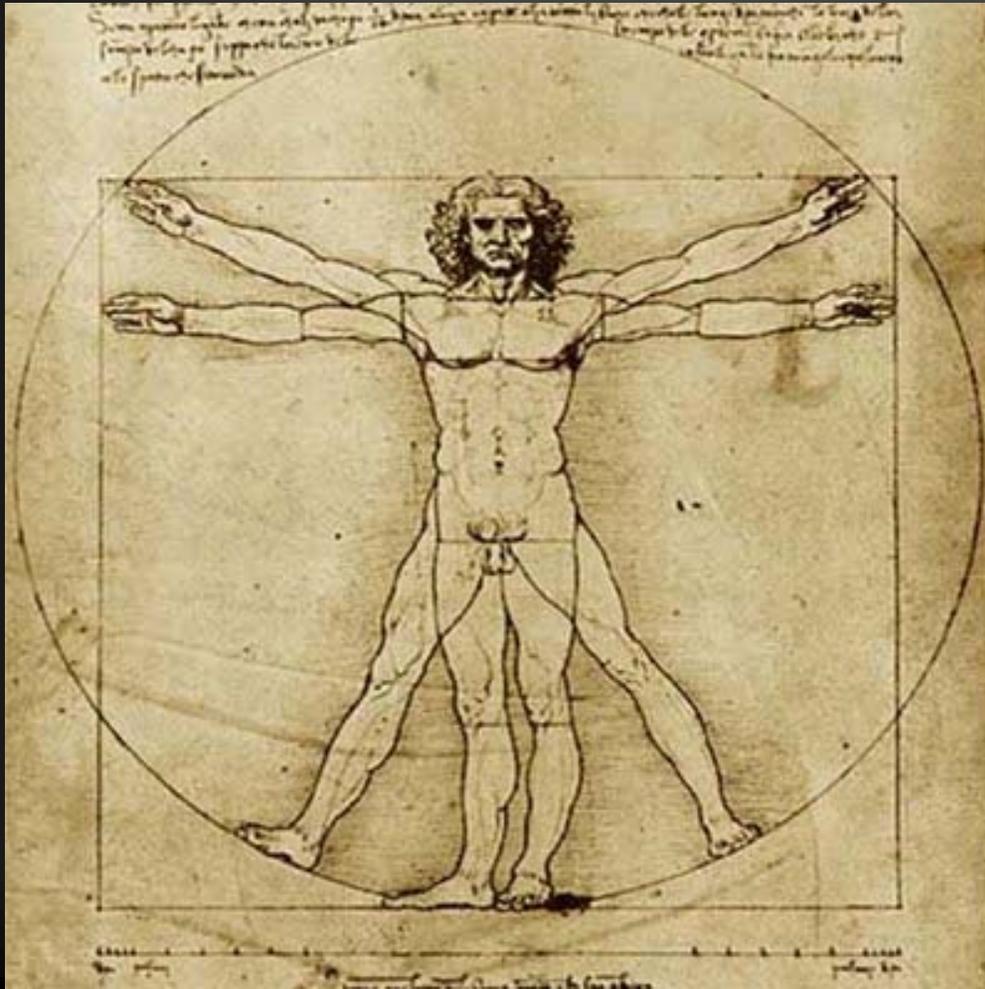
- Serología: títulos x4 en 2 muestras
- PCR
- Cultivo



Encefalitis
Meningitis aséptica
ACVs múltiples por endocarditis

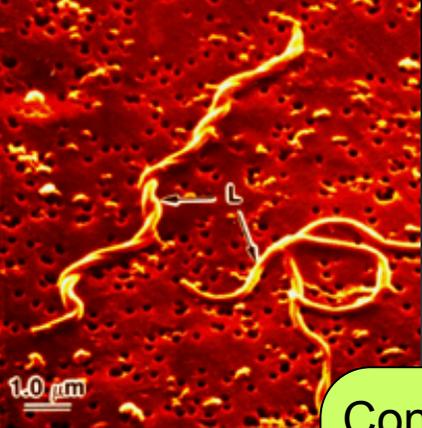


Borrelia burgdorferi
Enfermedad de Lyme
Fase precoz



Artromialgias, astenia
Adenopatía locorregional





Borrelia burgdorferi

Enfermedad de Lyme

Fase diseminada



Conjuntivitis
Iritis
Coroiditis
Vitritis
retinitis

(5%) Miopericarditis
Bloqueo AV

Neumonitis

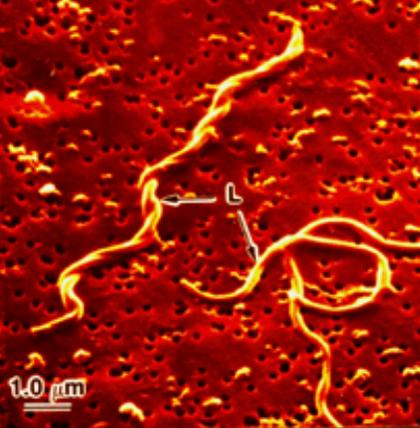
Hepatitis

60%
Artralgias migratorias

Adenopatías
regionales/genralizadas



Meningitis linfocitaria
Encefalitis
Neuropatías
(PC, facial, bilateral)
Polirradiculitis
Mielitis
Ataxia cerebelosa
Vasculitis



Borrelia burgdorferi

Enfermedad de Lyme



Fatal course of cerebral vasculitis induced by neuroborreliosis

Fredrik Buchwald¹, Kasim Abul-Kasim², Johan Tham³, Bjarne U Hansen⁴

¹ Department of Neurology, Faculty of Medicine, University of Lund, Malmö University Hospital, Malmö, Sweden

Volume 81, Issue 4, Pages 519-521 (April 2000)

Lyme neuroborreliosis mimics stroke: A case report☆☆☆

Yutong Zhang, MD, PhD, Gilbert Lafontant, MD, Francis J. Bonner Jr., MD

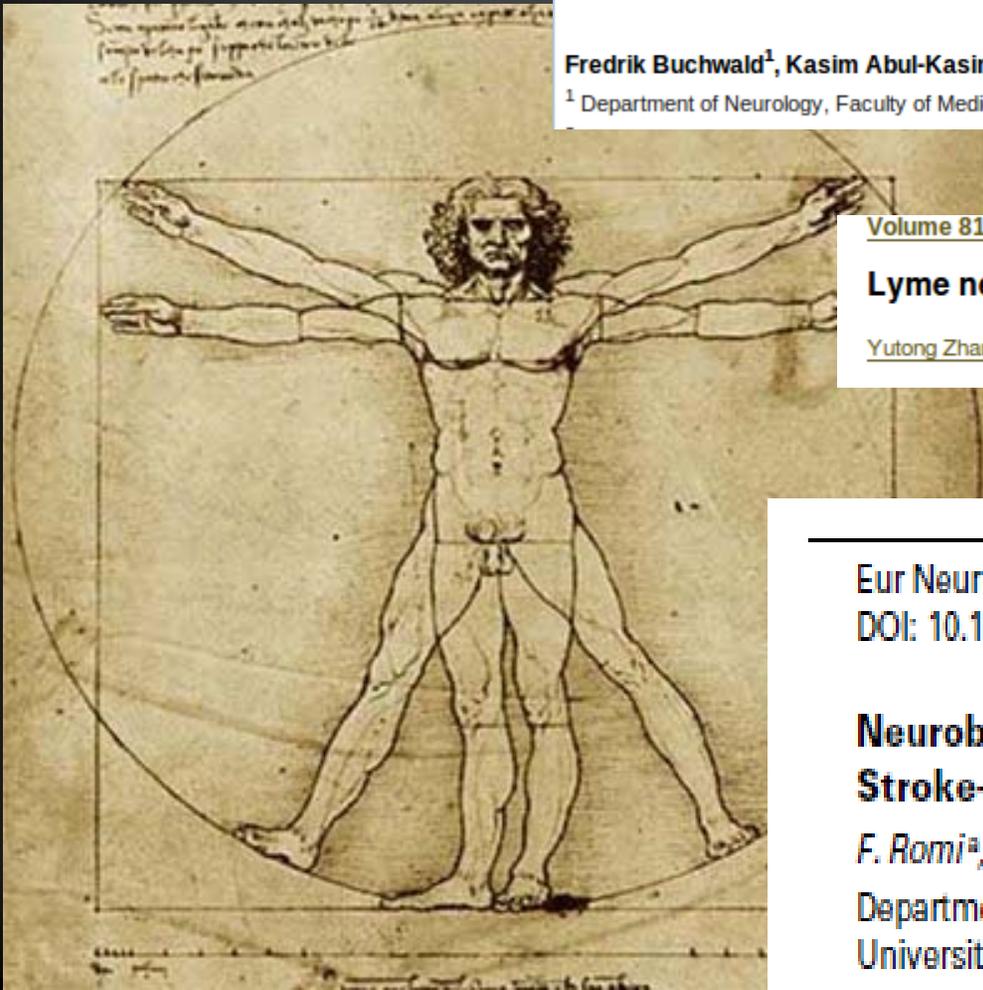
Eur Neurol 2004;51:49-50

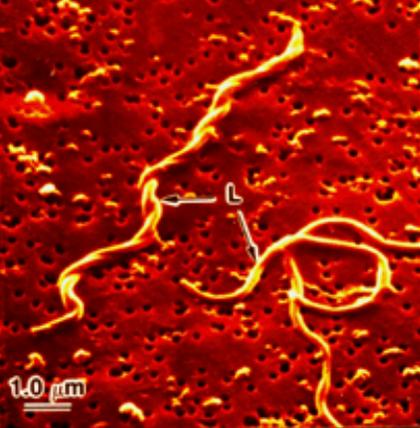
DOI: 10.1159/000075090

Neuroborreliosis with Vasculitis Causing Stroke-Like Manifestations

F. Romi^a, J. Kråkenes^b, J.A. Aarli^a, O.-B. Tysnes^a

Departments of ^aNeurology and ^bRadiology, Haukeland University Hospital, Bergen, Norway



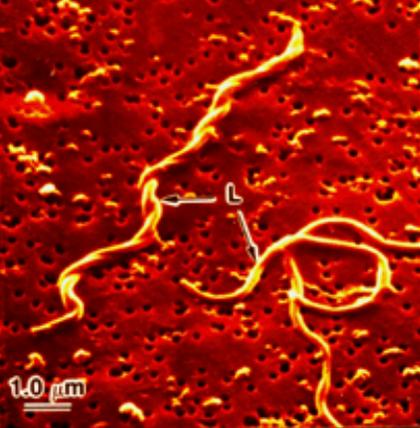


Borrelia burgdorferi
Enfermedad de Lyme
Diagnóstico



•CLÍNICO

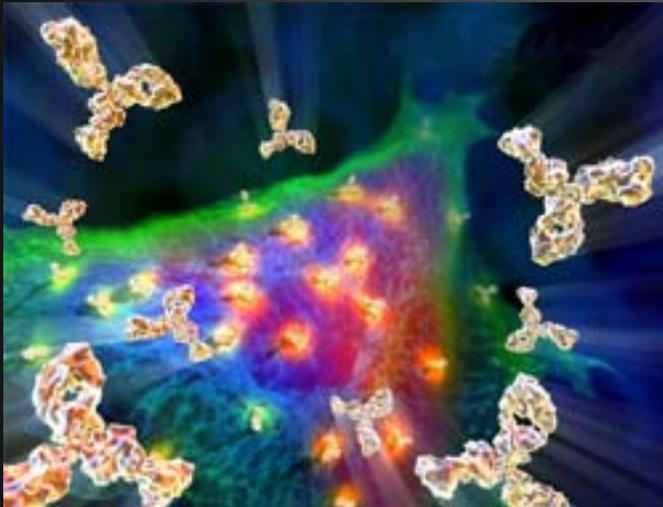
- Fiable
- Eritema migrans tras picadura en zona de alta prevalencia
- Ausencia de marcadores serológicos en fase precoz infección



Borrelia burgdorferi

Enfermedad de Lyme

Diagnóstico

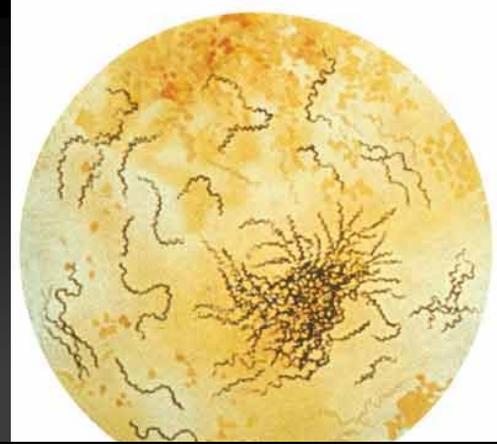


SEROLÓGICO

- Muchos FP (hacer sólo si sospecha clínica, nunca Dx sólo por serología)
- Periodo ventana IgM 1-2 sem, IgG 4-8 sem.
- Dos pasos
 - ELISA
 - Western-Blot
- Neuroborreliosis
 - Serología en LCR
 - PCR en LCR

Treponema pallidum

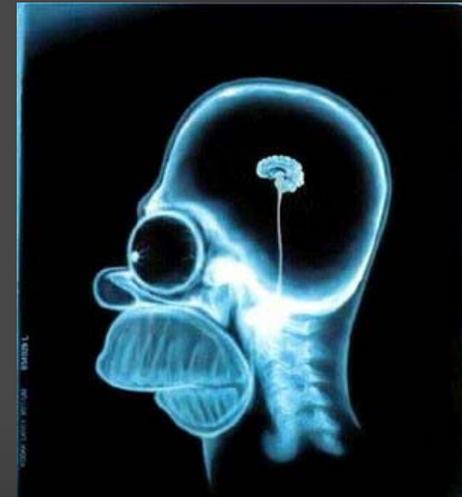
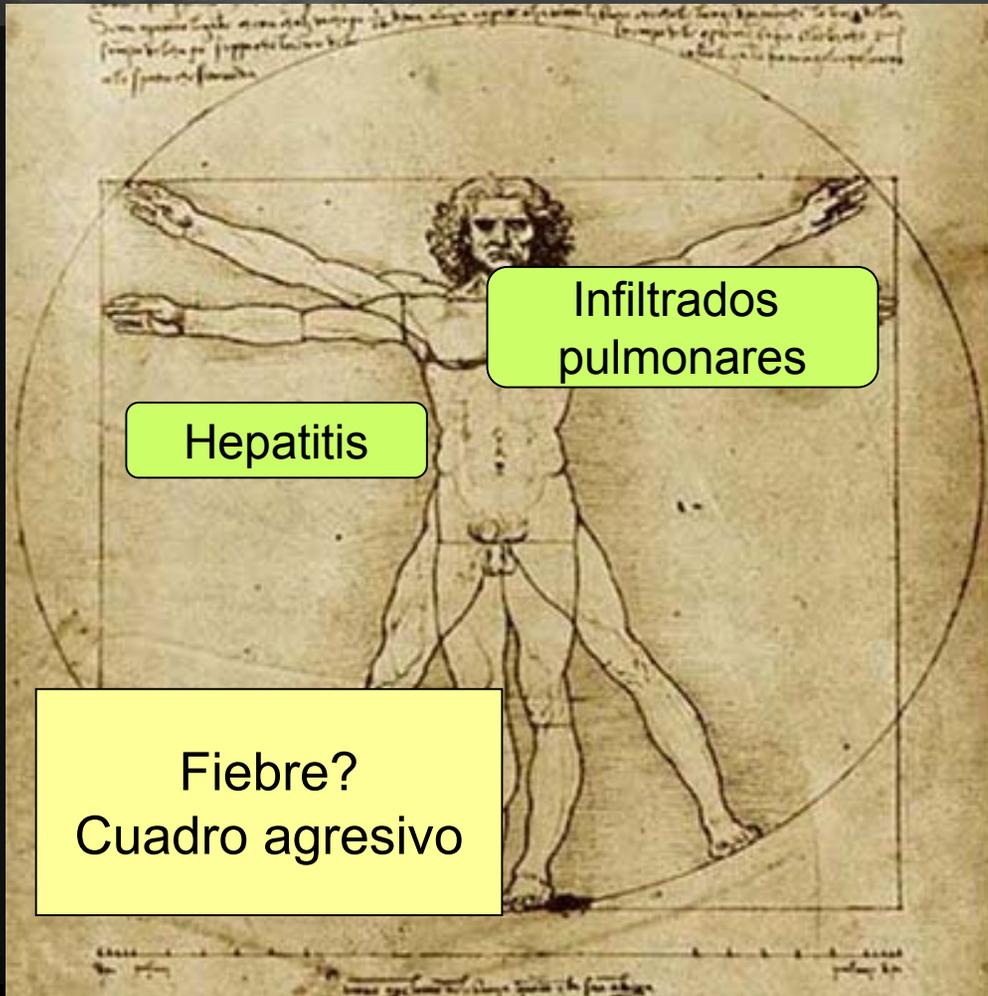
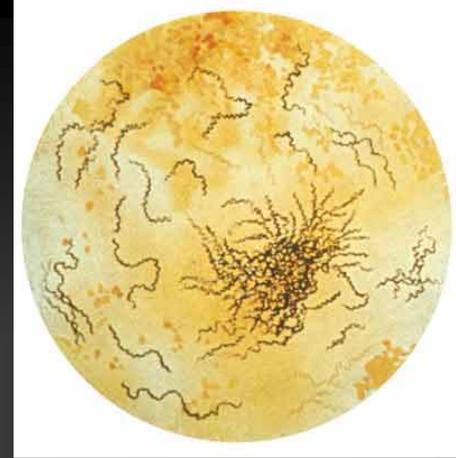
Sífilis



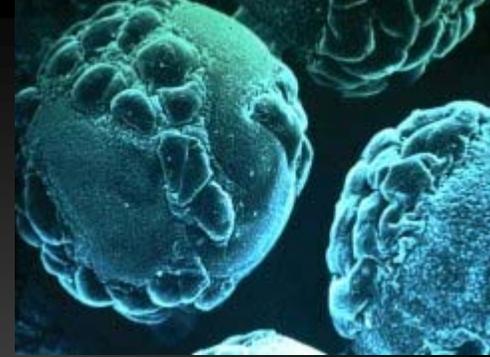
“Es más probable encontrar manifestaciones infrecuentes de enfermedades comunes que manifestaciones frecuentes de enfermedades raras”

Treponema pallidum

Sífilis

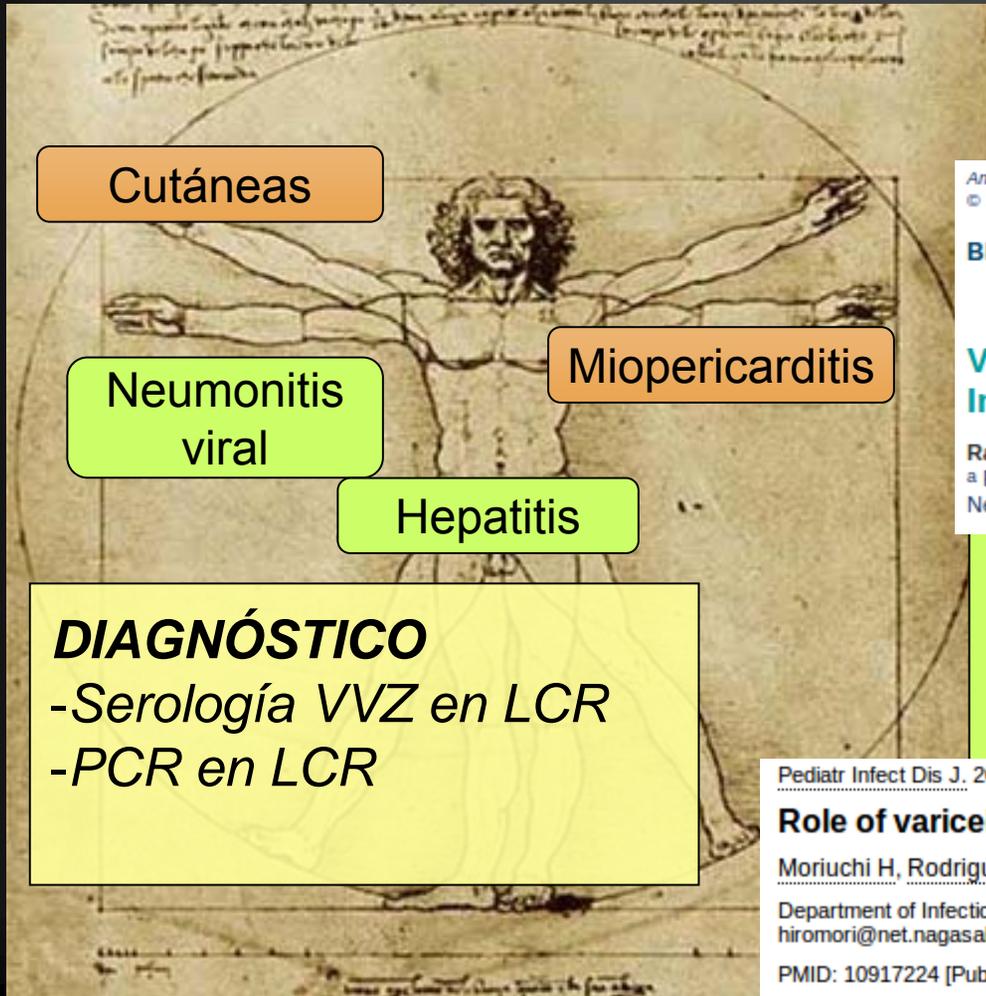


Sífilis secundaria
Meningitis
Sífilis meningovascular



VIRUS

Varicela zoster



Cutáneas

Neumonitis viral

Hepatitis

Miopericarditis

DIAGNÓSTICO
-Serología VVZ en LCR
-PCR en LCR

American Journal of Neuroradiology 24:971-974, May 2003
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BRAIN

Varicella-Zoster Vasculitis Presenting with Intracranial Hemorrhage

Rajan Jain^a, John Deveikis^a, Susan Hickenbottom^a and Suresh K. Mukherji^a
^a From the Division of Neuroradiology and Department of Radiology, Department of Neurology, University of Michigan Health System, Ann Arbor, MI

Meningoencefalitis
Encefalopatía Aguda Desmielinizante (ADEM)
Guillain-Barré

Pediatr Infect Dis J. 2000 Jul;19(7):648-53.

Role of varicella-zoster virus in stroke syndromes.

Moriuchi H, Rodriguez W.

Department of Infectious Diseases, Children's National Medical Center, Washington, DC, USA.
hiromori@net.nagasaki-u.ac.jp

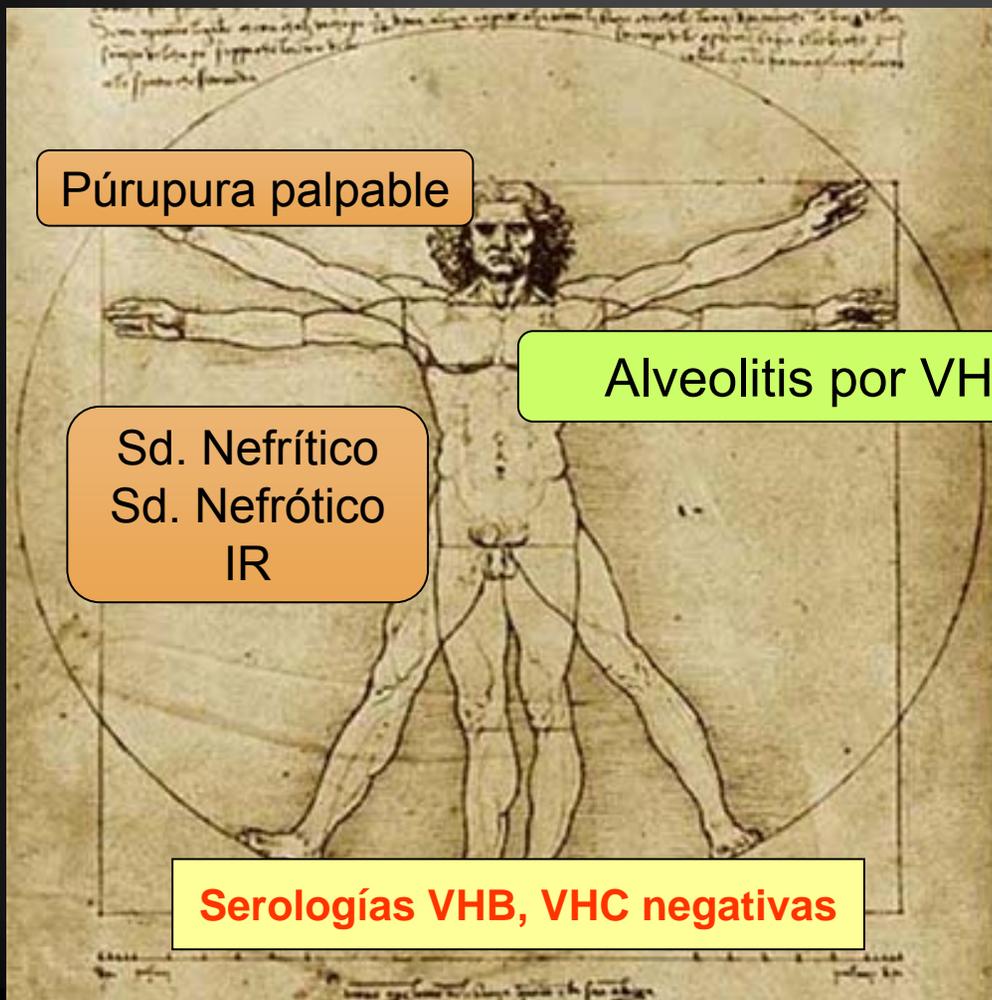
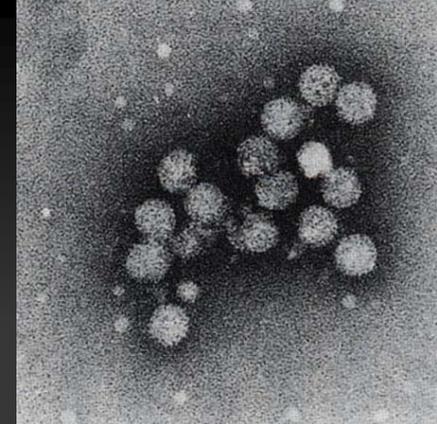
PMID: 10917224 [PubMed - indexed for MEDLINE]



Virus Hepatitis C

Virus hepatitis B

Crioglobulinemia mixta esencial

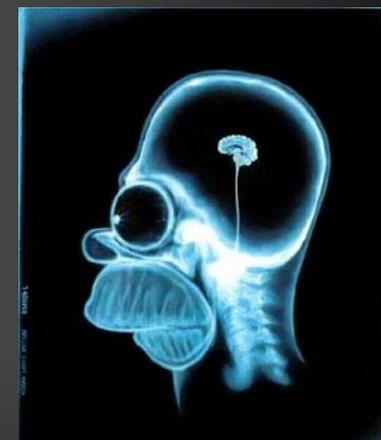


Púrpura palpable

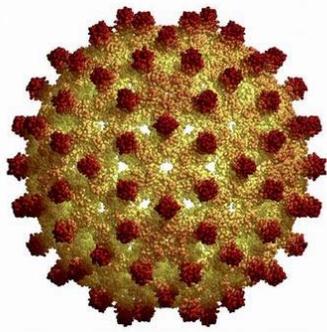
Sd. Nefrítico
Sd. Nefrótico
IR

Alveolitis por VHC

Serologías VHB, VHC negativas

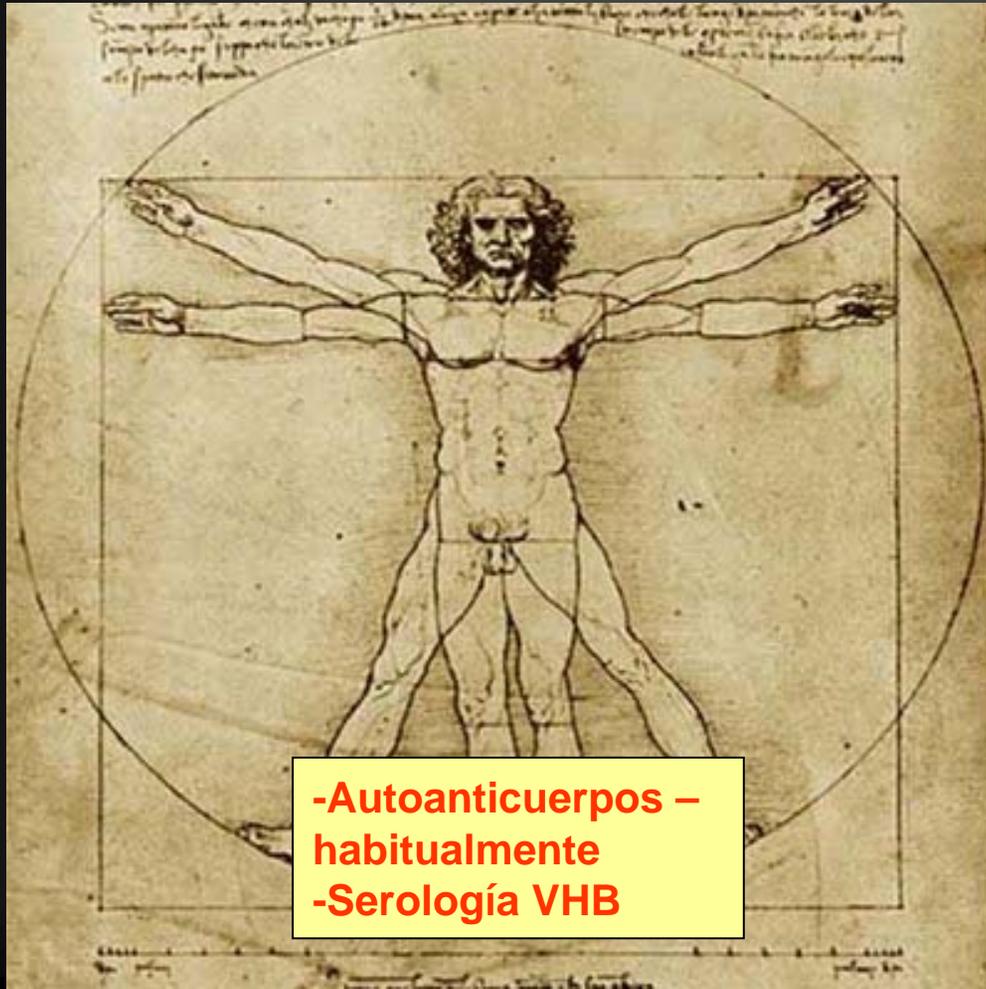


Vasculitis crioglobulinémica

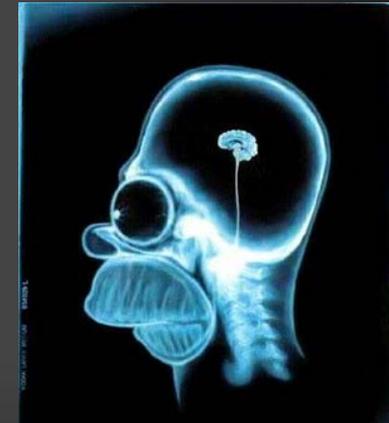


Virus Hepatitis B

Panarteritis nodosa



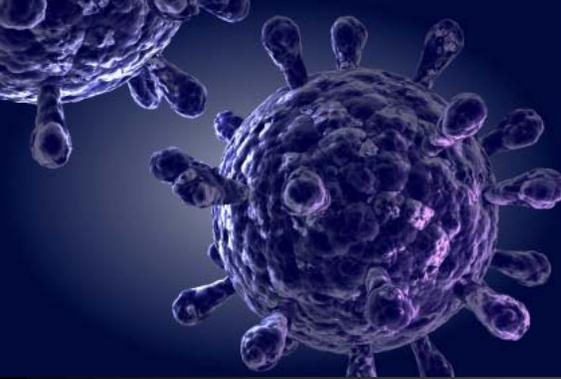
**-Autoanticuerpos –
habitualmente
-Serología VHB**



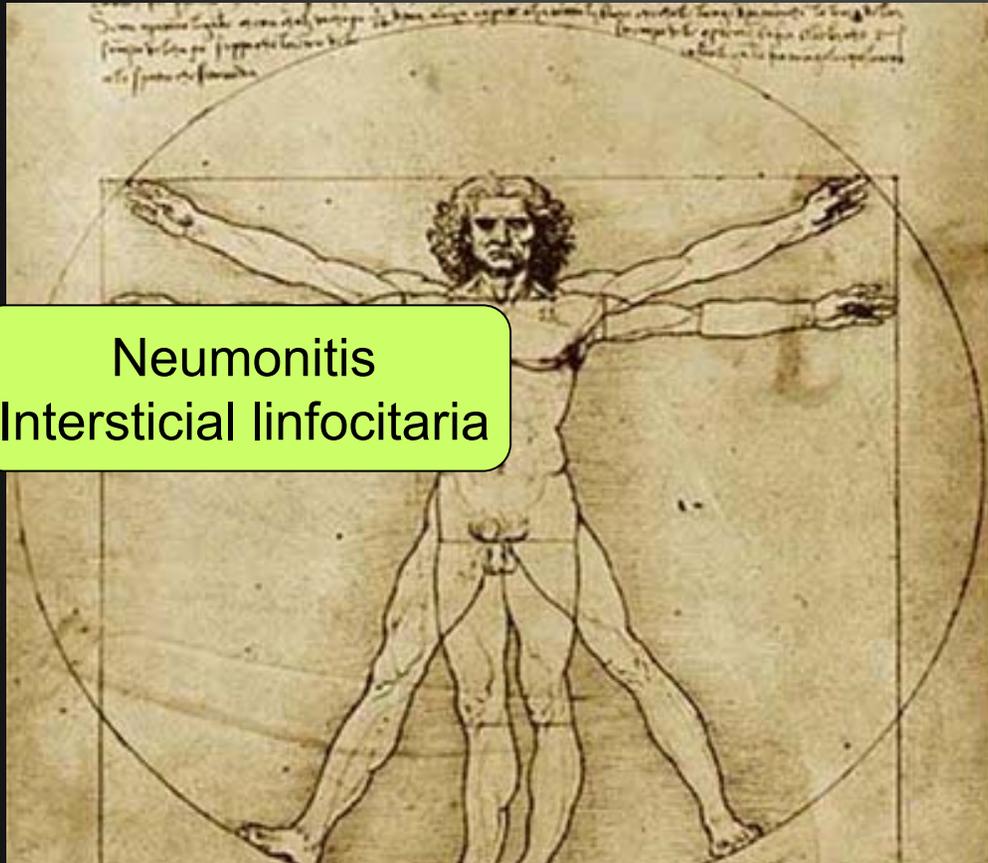
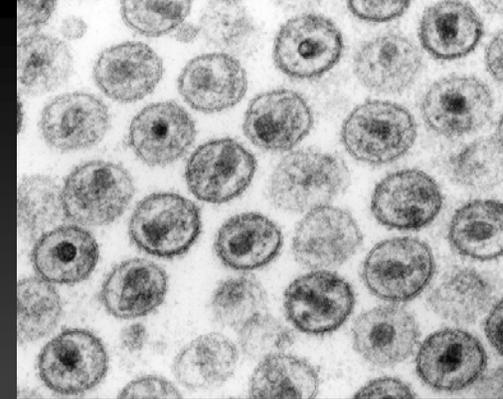
**Mononeuritis múltiple
ACV isquémicos/hemorrágicos
(10%)**

Provenzale JM, Am J Neuroradio 1996

Cohen RD, Mayo Clin Proc 1980

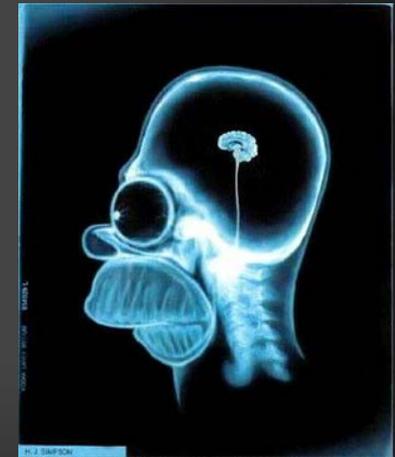


Virus Inmunodeficiencia Humana



Neumonitis
Intersticial linfocitaria

Serología VIH negativa



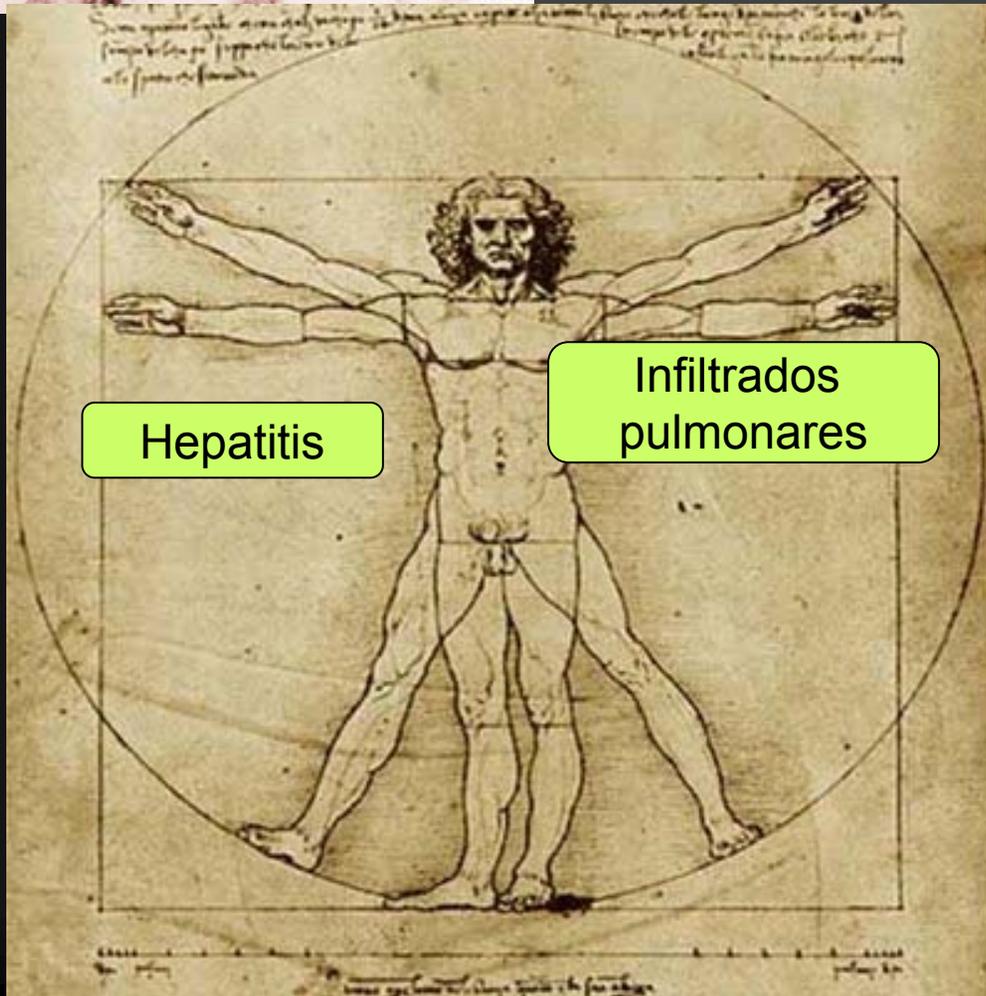
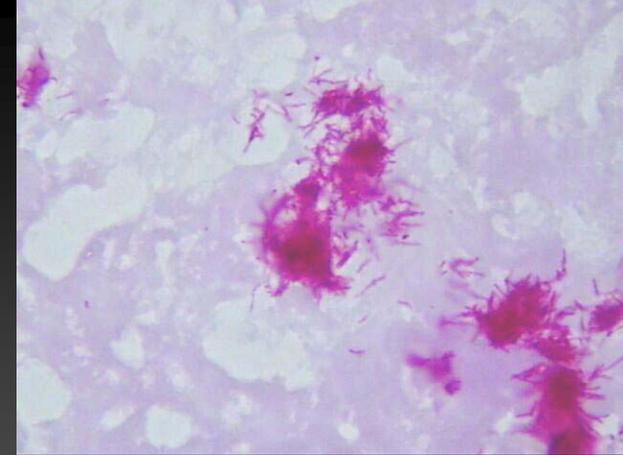
Vasculitis SNC

Neuroradiology. 2000 Jul;42(7):526-8.
MRI in human immunodeficiency virus-associated cerebral vasculitis.
[Berkefeld J](#), [Enzensberger W](#), [Lanfermann H](#).

PUES SERA UN VIRUS,
PERO TIENE UNA CARA
DE BACTERIA QUE NO SE LAME



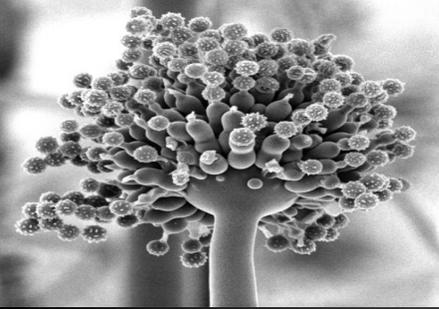
Micobacterium tuberculosis



Hepatitis

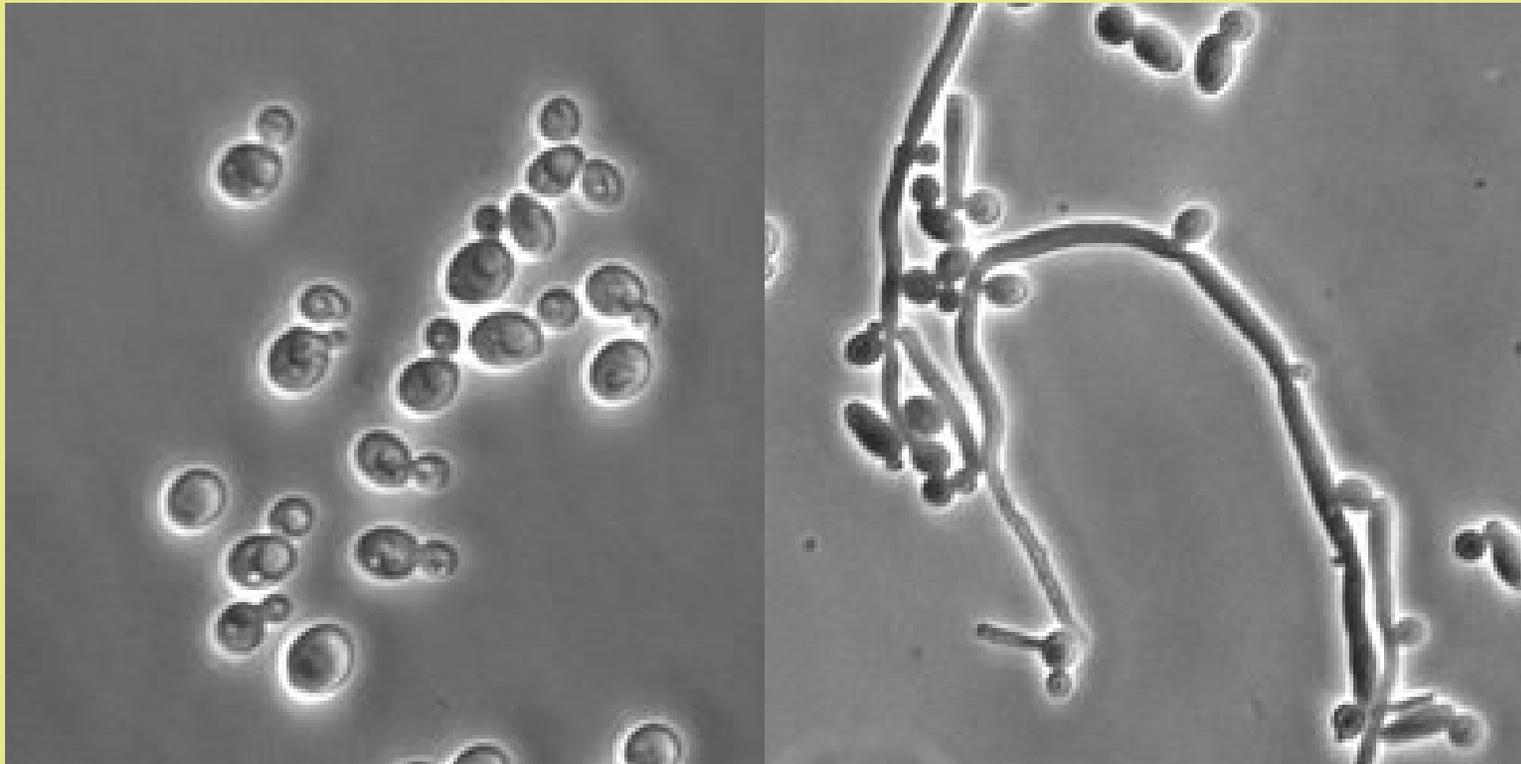
Infiltrados
pulmonares

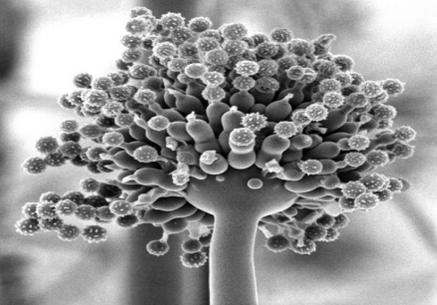
1. Meningitis / Hydrocephalus
2. Tuberculoma
3. Vasculitis / Spinal artery syndromes
4. Potts / paraplegia
5. Myeloradiculitis



Hongos

Candida sp.





Cerebral Vasculitis Caused by *Aspergillus* Species in an Immunocompetent Adult

M. Roberts, A. Carmichael, P. Martin



Abstract

We describe a case of cerebral vasculitis due to *Aspergillus* infection in an immunocompetent adult, in whom the probable source was paranasal sinus infection. At postmortem there was widespread endovascular *Aspergillus* infection of the cerebral arteries with occlusion of the basilar artery. We review the literature regarding intracranial *Aspergillus* infection and new approaches to diagnosis.

Infection 2004; 32: 360–363
DOI 10.1007/s15010-004-3077-1

Case Report

A 71-year-old Caucasian female was admitted with a 5-week history of malaise and intermittent headaches. She had a past history of sinusitis treated with courses of antibiotics and hypertension treated with bendrofluzide and diltiazem. She had no history of immune deficiency or immunosuppressive treatment.

On admission, she was afebrile with a pulse of 80 per min, blood pressure of 135/70 and a soft pansystolic murmur at the cardiac apex. Abdominal and neurological examinations were normal. Initial investigations showed hemoglobin 7.6 g/dl, mean cell volume 76 fl, neutrophil count $6.8 \times 10^9/l$, erythrocyte sedimentation rate 32 mm/h, and C-reactive protein 3 mg/l. A blood film was normal except for some hypochromic, microcytic cells consistent with the development of an iron deficiency anemia with a low serum ferritin. Fecal occult bloods were negative. The blood glucose was normal and the chest X-ray was normal. During a blood transfusion she developed a fever of 38.3 °C that persisted after the transfusion ceased. Three sets of blood cultures and a urine culture showed no bacterial growth. A transthoracic echocardiogram showed minimal mitral regurgitation (subsequent autopsy confirmed normal cardiac valves). She developed an increasingly severe headache, accompanied by intermittent diplopia, confusion, and increasing drowsiness. A head CT scan showed a small area of low attenuation in the right internal capsule, consistent with a lacunar infarct. Lumbar cerebrospinal fluid (CSF) examination showed neutrophils 108/field, lymphocytes 178/field, protein 0.85 g/l, glucose 1.3 mmol/l (compared to blood glucose 5.5 mmol/l). A Gram stain was negative and subsequent CSF bacterial and prolonged cultures for *Mycobacterium tuberculosis* were negative.

Because of the clinical features of a lymphocytic meningoencephalitis of unknown etiology, she was treated with rifampicin, isoniazid, pyrazinamide, ethambutol, acyclovir, and meropenem.

Repeated CSF examinations showed decreasing numbers of neutrophils and lymphocytes, but a persistent hypoglycocoachia and increasing protein concentration. In multiple samples of CSF, no mycobacteria or fungi were seen on microscopy and mycobacteria DNA PCR, herpesvirus DNA PCR and CSF cryptococcal antigen were negative.

Seven days after admission she remained drowsy and developed a left hemiparesis. A cranial MRI scan including diffusion weighting showed the small area of ischemic damage in the right internal capsule. There was mild dilatation of the lateral ventricles, and fluid in the left maxillary antrum and sphenoid sinus.

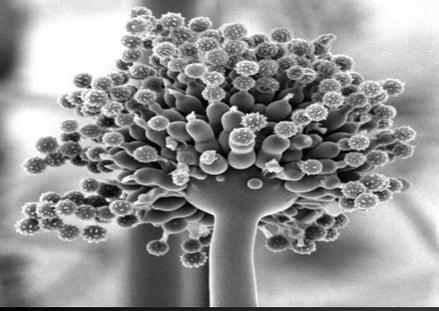
Clinically, it was suspected that she had developed cerebral vasculitis, possibly secondary to tuberculous meningitis. She was treated with intravenous methylprednisolone 1g and an external ventricular drain was inserted, without improvement in her level of consciousness. Cerebral angiography showed diffuse irregularity of right and left middle cerebral, right and left posterior cerebral and basilar arteries, consistent with an arteritis. A right frontal brain biopsy showed histological features suggestive of vasculitis. Despite reservations that an infective process might have triggered her vasculitis, intravenous cyclophosphamide and then intravenous immunoglobulin were administered without improvement. Evidence of sinusitis was noted (Figure 1a). A repeat cranial MRI scan showed an occlusion of the basilar artery and extensive infarction of the brainstem and territory of both posterior cerebral arteries (Figure 1b). The patient died 3 days later.

At postmortem, there was thrombosis of the basilar artery, with infarction of the pons. The wall of the basilar artery was extensively infiltrated with dichotomously branching septate hyphae of *Aspergillus* species (Figure 2). Fungal hyphae were also identified within necrotic granulomas in the cerebral hemispheres and meninges (Figure 3); tissue for culture was not obtained. *Aspergillus* was not identified in any other organs, particularly the lungs. There was no evidence of immunosuppression with a normal total and differential white cell count, normal immunoglobulin profile and human immunodeficiency virus antibody negativity, and no

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P. Martin
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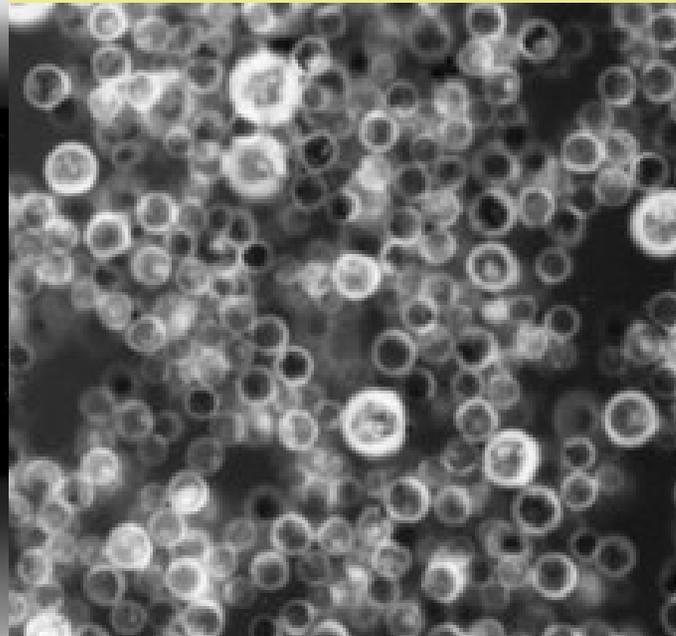
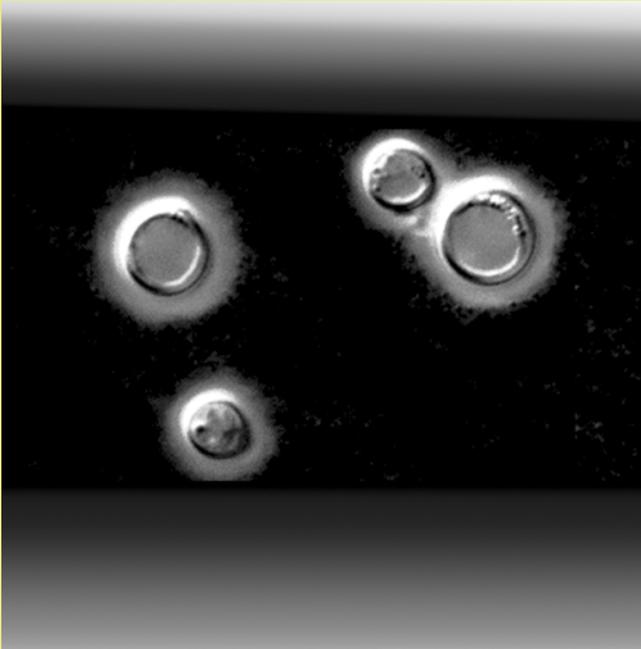
Received: May 5, 2003 • Revision accepted: December 11, 2003



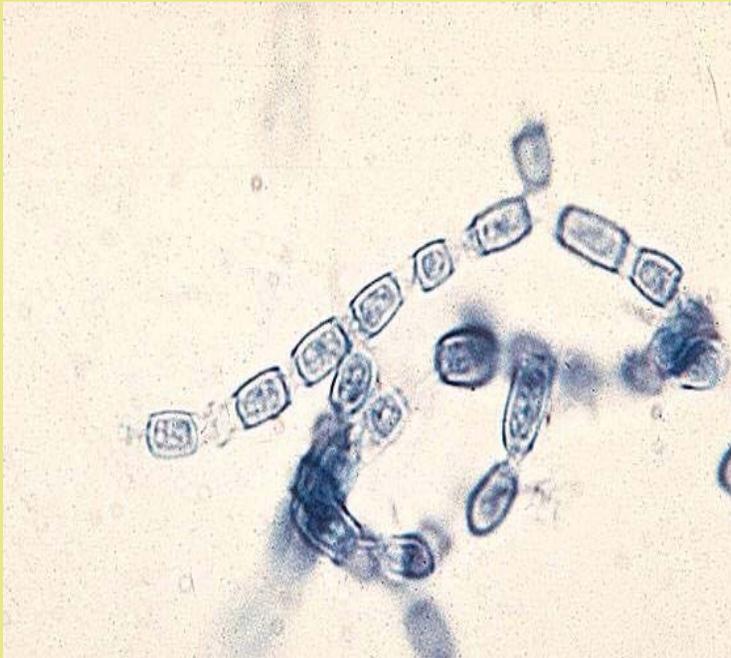
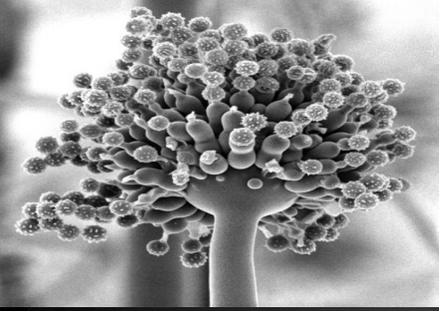


Hongos

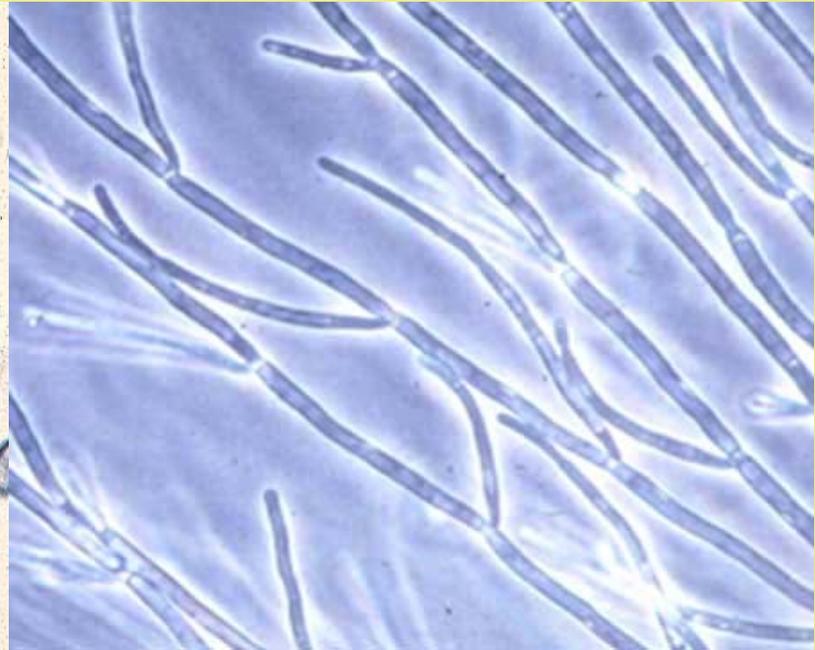
Cryptococcus neoformans



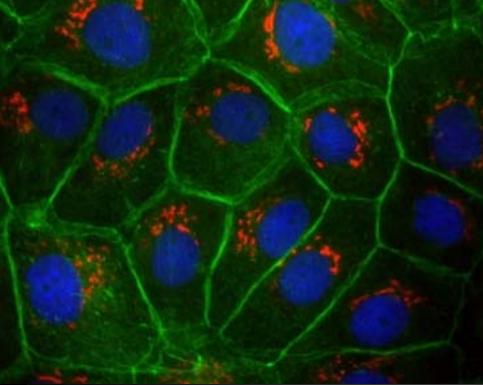
Hongos



Coccidioides immitis



Blastoschizomyces capitatus



Autoinmunes

Colagenovasculares



- LES
- Wegener
- Panarteritis nodosa
- Behçet
- Sjögren
- Artritis reumatoide



Frequency of symptoms of systemic lupus erythematosus

Symptoms	Percent at onset	Percent at anytime
Fatigue	50	74-100
Fever	36	40-80+
Weight loss	21	44-60+
Arthritis or arthralgia	62-67	83-95
Skin	73	80-91
Butterfly rash	28-38	48-54
Photosensitivity	29	41-60
Mucuous membrane lesion	10-21	27-52
Alopecia	32	18-71
Raynaud's phenomenon	17-33	22-71
Purpura	10	15-34
Uticaria	1	4-8
Renal	16-38	34-73
Nephrosis	5	11-18
Gastrointestinal	18	38-44
Pulmonary	2-12	24-98
Pleurisy	17	30-45
Effusion		24
Pneumonia		29
Cardiac	15	20-46
Pericarditis	8	8-48
Murmurs		23
ECG changes		34-70
Lymphadenopathy	7-16	
Splenomegaly	5	
Hepatomegaly	2	
Central nervous system	12-21	
Functional		
Psychosis	1	
Convulsions	0.5	



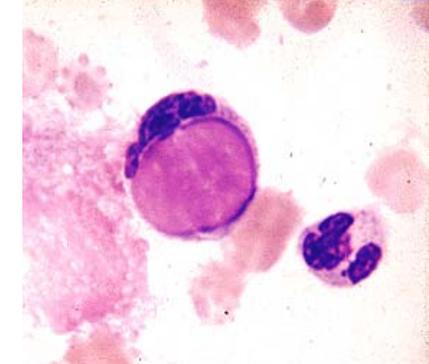
Trombosis- Sd. Antifosfolípido
 Aterosclerosis acelerada
 Hemorragia subaracnoidea
 Endocarditis Libman-Sacks
 Vasculitis

Adapted from: Von Feldt, JM, Postgrad Med 1995; 97:79.

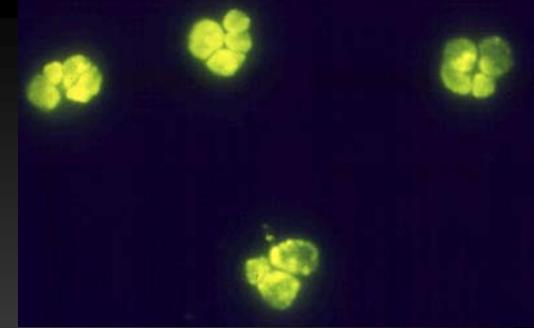
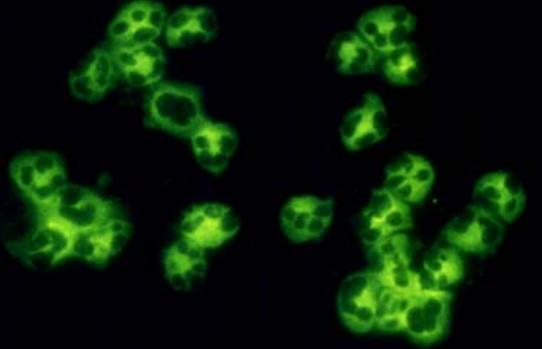


ARA criteria for diagnosis of systemic lupus erythematosus

Criterion	Definition
Malar rash	Fixed erythema, flat or raised, over the malar eminences, tending to spare the nasolabial folds
Discoid rash	Erythematous raised patches with adherent keratotic scaling and follicular plugging; atrophic scarring may occur in older lesions
Photosensitivity	Skin rash as a result of unusual reaction to sunlight, by patient history or physician observation
Oral ulcers	Oral or nasopharyngeal ulceration, usually painless, observed by a physician
Arthritis	Nonerosive arthritis involving 2 or more peripheral joints, characterized by tenderness, swelling, or effusion
Serositis	Pleuritis - convincing history of pleuritic pain or rub heard by a physician or evidence of pleural effusion OR
	Pericarditis - documented by EKG, rub or evidence of pericardial effusion
Renal disorder	Persistent proteinuria greater than 0.5 grams per day or greater than 3+ if quantitation not performed OR
	Cellular casts - may be red cell, hemoglobin, granular, tubular, or mixed
Neurologic disorder	Seizures OR psychosis - in the absence of offending drugs or known metabolic derangements (uremia, ketoacidosis, or electrolyte
Hematologic disorder	ANA -
Immunologic disorders	Positive antiphospholipid antibody OR
	Anti-DNA - antibody to native DNA in abnormal titer OR
	Anti-Sm - presence of antibody to Sm nuclear antigen OR
	False positive serologic test for syphilis known to be positive for at least six months and confirmed by Treponema pallidum immobilization or fluorescent treponemal antibody absorption test
Antinuclear antibody	An abnormal titer of antinuclear antibody by immunofluorescence or an equivalent assay at any point in time and in the absence of drugs known to be associated with "drug-induced lupus" syndrome



s **OR**
R



Autoinmunes

Granulomatosis de Wegener Panarteritis microscópica

Vasculitis cerebral

Fiebre
MEG
Sudoración
Nocturna
Pérdida de peso

Vías resp altas
Vías resp bajas
Hemorragia alveolar
Opacidades alveolares
Opacidades pleurales
Nódulos pulmonares
Adenopatía hiliar

1. Inflamación nasal/oral (úlceras, secreción purulenta)
2. Rx de tórax: nódulos, infiltrados fijos, cavidades
3. Sedimento urinario: microhematuria± cilindros hemáticos
4. Biopsia de arteria o área perivascular: inflamación granulomatosa

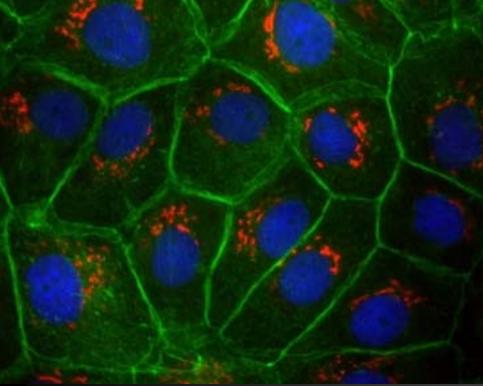
>2 criterios: **S 88%, E 92%**

Leavitt RY, Arthritis Rheum 1990

ANCA-

10% formas sistémicas
40% formas localizadas

Yamashita Y, J Comput Tomogr 1986



Autoinmunes

Enfermedad de Behçet



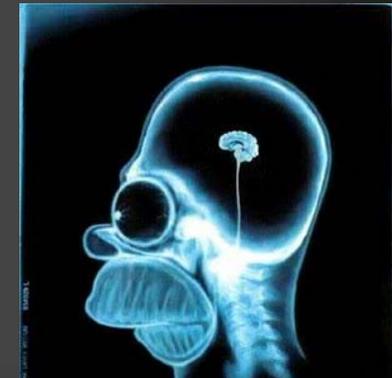
Vasculitis cerebral

Vasculitis
Pequeño
Mediano gran
vaso

Fiebre
MEG
Sudoración
Nocturna
Pérdida de peso

Infartos pulmonares
Hemorragia alveolar
COP
Neumonía eosinofílica

Úlceras Orales Recurrentes



20%

ACV isquémicos
Meningitis aséptica
Encefalitis

Serdaroglu P, J Neurol 1998

Autoinmunes Colagenovasculares



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Servicios de ¹Neurología y ²Neurorradiología
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Murcia

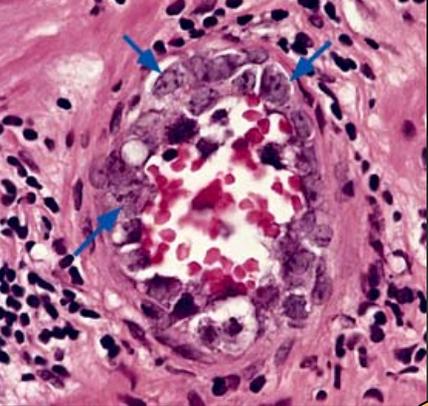
Vasculopatía cerebral fulminante
en el síndrome de Sjögren

Neurología 2009;24(7):498-510

Patrón pulmonar intersticial

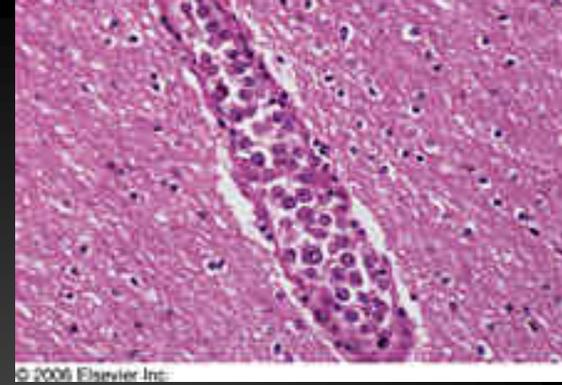
Xerostomía
Xeroftalmia
antiRo, antiLa





Neoplásicas

Linfoma intravascular de célula grande



© 2006 Elsevier Inc.

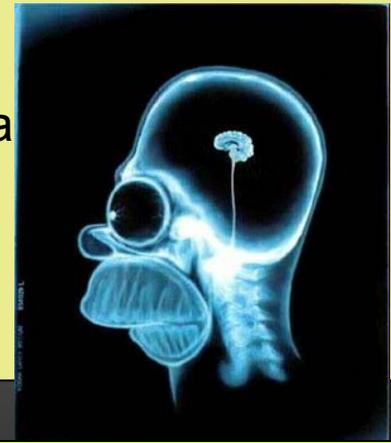
Linfoma intravascular

Lesiones cutáneas
Síndrome hemofagocítico
Hepatoesplenomegalia
Infiltrados pulmonares
fijos

Linfoma de célula grande, Angioendoteliomatosis maligna

Asociados con
alteración de los vasos

(ausencia)



80%
Síntomas B
Fiebre
MEG
Sudoración
Nocturna
Pérdida de peso

Aumento de:
LDH
B2-microglobulina
VSG
(90%)
Trombopenia (75%)
Transaminasas (20%)
Creatinina (20%)

40%
Afectación neurológica
grave y progresiva (ACVs),
demencia

Agradecimientos...

CASE RECORDS of the MASSACHUSETTS GENERAL HOSPITAL

Founded by Richard C. Cabot

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Case 9-2010: A 37-Year-Old Woman with Paresthesias and Ataxia

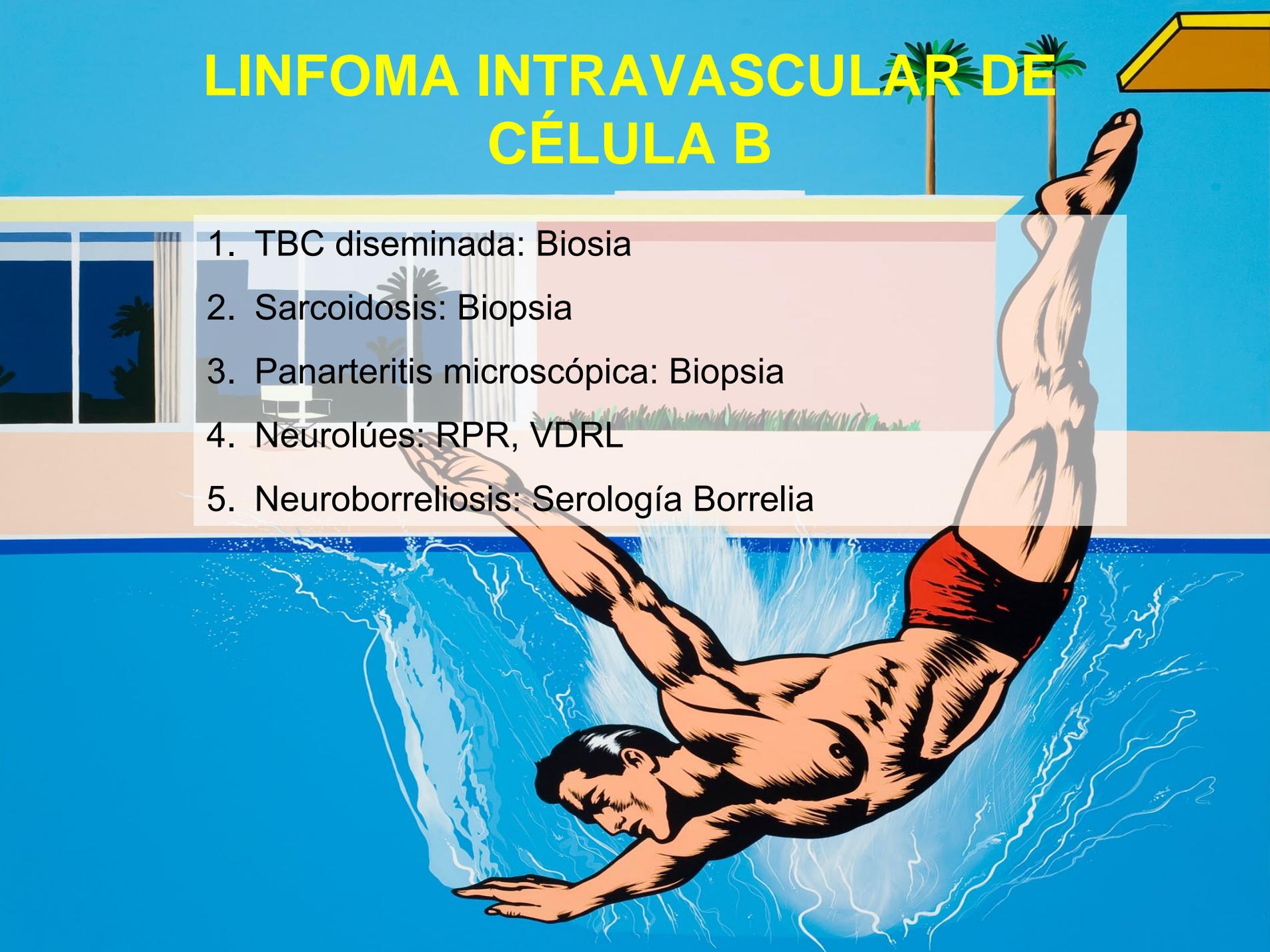
Misha L. Pless, M.D., Yi-Bin Chen, M.D., William A. Copen, M.D.,
and Matthew P. Frosch, M.D., Ph.D.

N ENGL J MED 362:12 NEJM.ORG MARCH 25, 2010

*“Es bueno leer el NEJM,
Si tienes FR para discutir un caso cerrado en público”*

LINFOMA INTRAVASCULAR DE CÉLULA B

1. TBC diseminada: Biopsia
2. Sarcoidosis: Biopsia
3. Panarteritis microscópica: Biopsia
4. Neurolúes: RPR, VDRL
5. Neuroborreliosis: Serología Borrelia



THE CASE OF WILLIE SUTTON

SUTTON'S RULE

Willie "The Actor" Sutton

FBI Ten Most Wanted Fugitives



Willie "The Actor" Sutton

Information

Born: June 30, 1901

Died: November 2, 1980
(aged 79)
Spring Hill, Florida

Charges: Bank robbery

Added: March 20, 1950

Caught: February 1952

Number: 11

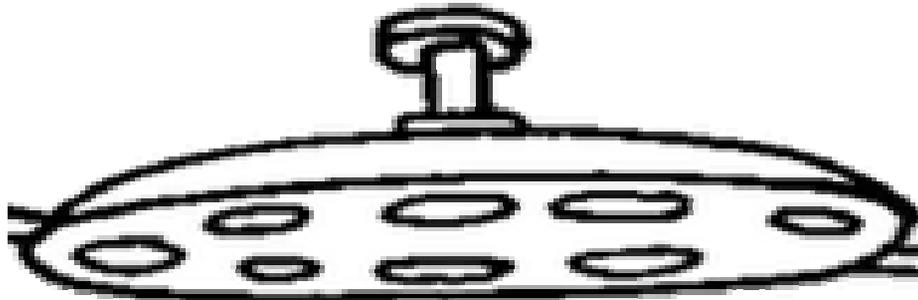
Captured

- William "Willie" Sutton (Brooklyn, 1901)
- Carrera criminal de 40 años
- Willie "el actor" y Willie "el hábil"
- "You can't rob a bank on charm and personality"
- Más de 2 millones \$ durante su carrera
- Consultor de bancos sobre técnicas antirrobo

"-Hey Willie, why do you still keep robbing banks?"

- Well, there's where the money is!"

HACEDOS
A UN LADO...



BIOPSIA HEPÁTICA

VS

CEREBRAL



... QUE HOY
TENGO UN
QA TENSO





MUCHAS GRACIAS