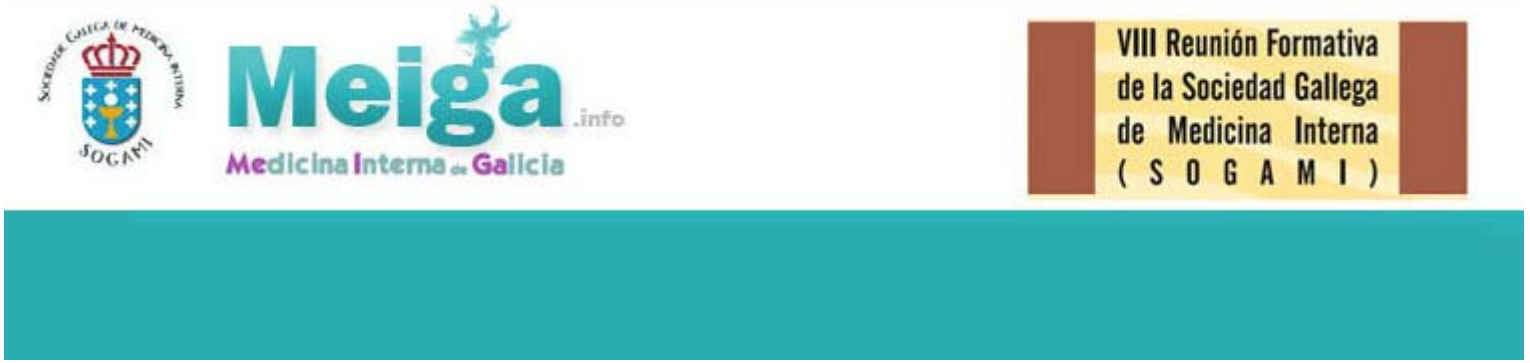
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Medicina Interna de Galicia
VIII Reunión Formativa
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(S O G A M I)

Caso 2

- Paciente de 60 años, sin hábitos tóxicos ni AMC

- AP:
  - Bronquiectasias pulmonares
  - HTA (hidroclortiacida)

- EA: Febrícula 37.5-38º + cefalea holocraneal + tos + sd. constitucional con pérdida 3 kg peso, de 1 mes evolución. Artromialgias y rigidez manos en las ultimas dos semanas. No claudicación mandibular. No alteraciones visuales.

Caso 2

- EXPLORACIÓN FISICA:
  - Ligera palidez piel y mucosas. No adenopatías. No lesiones cutáneas. Temp: 37.7º, TA: 150/95
  - ACR: TCR 80 pm, SS 2/6 Ao. Crepitantes gruesos ambas bases pulmonares. Sibilantes aislados.
  - Abdomen blando y depresible, sin megalias. PPL negativa
  - Expl. Neurológica sin focalidad
  - Ligera sinovitis ambas muñecas
  - A Temporales visibles, no induradas, pulsátiles

Caso 2

- ANALITICA: Hb 10.6, Hto 30.5, VCM 86, plaq 360.000, leucos 20.000 (90%S), urea 40, creat 0.9, GOT 50, GPT 70, FA 150, GGT 90, VSG 90, PCR 3, fib 4. ANA negativos.

- Hemocultivos y urocultivo: negativos
- Serología neumococo, mycoplasma, coxiella, lues, HVB, HVC: negativas

- Rx Tórax: aumento densidad bases pulmonares

- TAC torácico: bronquiectasias en ambos lóbulos inferiores y LM. No condensaciones ni adenopatías.

- TAC abdominal: no visceromegalias. No adenopatías ni masas abdominales.

Criterios diagnósticos ACG

- Edad > 50 años al inicio
- Cefalea de reciente aparición o de nuevas características
- Anormalidades en las arterias temporales
- VSG > 50 mm 1ª hora
- Biopsia de la AT: infiltrado mononuclear + granulomas

3 de 5 criterios: sensibilidad 93,5 % y especificidad 91%

Hunder GG, Bloch DA, Michel BA. The American College of Rheumatology criteria for the classification of giant cell arteritis. Arthritis Rheum 1990

respuesta a corticoides

Caso 2

- BAT: compatible con arteritis de la temporal

- Se inició tratamiento con Prednisona 60 mg/día, despareciendo la sintomatología

- EVOLUCION:
- A los 3 meses del diagnóstico, tras disminución dosis prednisona a 20 mg/día, reinició febrícula y artritis manos, que desaparecieron al aumentar de nuevo la dosis de corticoides a 40 mg/día.

- A los 6 meses del diagnóstico, tras disminución de prednisona a 15 mg/día, reinicia de nuevo sintomatología que remite tras aumento dosis de esteroides. Consulta a nuestro Centro.

Caso 2

1. Ante la evolución clínica de la paciente: ¿Cuál seria la mejor opción?

    1) Mantener dosis de prednisona 20 mg/día durante al menos 6 meses
    2) Añadir metotrexate
    3) Administrar rituximab
    4) Replantear el diagnóstico
    5) Respuestas 1 y 2

Caso 2

- Se inició tratamiento con metotrexate, pudiendo disminuir progresivamente la dosis de prednisona.
- A los 18 meses del diagnóstico tras inicio descenso dosis de metotrexate, reinicia febrícula, tos seca, astenia, artromialgias y parestesias en EEII de predominio en pie izquierdo
- Analitica: VSG 90, Hb 10, Hto 30, VCM 90, leucos 17.000 (90%S), fib 3.6, PCR 3, creat 0.9, p hepáticas normales. ANA negativos. FR negativo
- Rx Torax: no condensaciones
- EMG: mononeuritis múltiple

Caso 2

2. Ante la evolución clínica de la paciente: ¿Cuál sería la mejor opción?

1) Aumentar dosis de metotrexate
2) Sustituir metotrexate por ciclofosfamida
3) Replantear diagnóstico
4) Practicar biopsia nervio sural
5) Respuestas 3 y 4

Giant-Cell Arteritis and Polymyalgia Rheumatica

Cornelia M. Weyand, MD, and Jörg J. Goronzy, MD

Ann Intern Med. 2003;139:505-515.

Table 1. The Spectrum of Clinical Features in Giant-Cell Arteritis

Feature	Manifestation
Manifestations related to vascular injury	
Common features (30%–80% of patients)	
Headaches	No particular pattern; severe, sometimes throbbing; often localized
Scalp tenderness	Often temporal; elicited by touching, grooming, or wearing glasses; temporal artery can be thickened, tender, or nodular
Jaw claudication	Elicited by prolonged talking or chewing
Less common features (<20% of patients)	
Ocular symptoms	Partial or complete visual loss, amaurosis fugax, or ocular motor deficit
Blindness	Unilateral or bilateral; usually permanent
Painful dysphagia	Sore throat
Respiratory symptoms	Dry, nonproductive cough
Limb claudication	Elicited by use of arms; combined with paresthesias
Absent or asymmetrical pulses	
Asymmetrical blood pressure readings	
Infrequent features (<5% of patients)	
Ischemia of the central nervous system	Typically vertebrobasilar insufficiency; imbalance; cortical blindness; confusion
Tongue claudication	
Aortic regurgitation	Dilatation of the proximal aorta
Myocardial infarction	
Peripheral neuropathy	
Deafness	
Tissue gangrene	Scalp, tongue, or extremities
Manifestations related to systemic inflammation	
Common features (40%–100% of patients)	
Intense acute-phase response	Elevated erythrocyte sedimentation rate, C-reactive protein level, interleukin-6 level, levels of other acute-phase proteins; elevated liver function test results; thrombocytosis
Anemia	Normocytic, normochromic
Polymyalgia rheumatica	Pain and stiffness in neck, shoulders, and pelvic girdle
Wasting syndrome	Fever, anorexia or weight loss, malaise, night sweats, depression
Infrequent features (<20% of patients)	
Peripheral synovitis	Typically wrist

BSR and BHPR guidelines for the management of giant cell arteritis

Bhaskar Dasgupta[1], Frances A. Borg[1], Nada Hassan[1], Leslie Alexander[1], Kevin Barraclough[2], Brian Bourke[3], Joan Fulcher[4], Jane Hollywood[1], Andrew Hutchings[5], Pat James[4], Valerie Kyle[6], Jennifer Nott[7], Michael Power[8] and Ash Samanta[9] on behalf of the BSR and BHPR Standards, Guidelines and Audit Working Group

Rheumatology 2010;49:1594–1597

A patient >50 years of age presenting with the following features should raise suspicion of GCA:

- Abrupt-onset headache (usually unilateral in the temporal area).
- Scalp tenderness.
- Jaw and tongue claudication.
- Visual symptoms (including diplopia).
- Constitutional symptoms.
- Polymyalgic symptoms.
- Limb claudication.

Examination may show:

- Abnormal superficial temporal artery: tender, thickened with reduced or absent pulsation.
- Scalp tenderness.
- Transient or permanent visual loss.
- Visual field defect.
- Relative afferent papillary defect.
- Anterior ischaemic optic neuritis.
- Central retinal artery occlusion.

The symptoms of GCA should respond rapidly to high-dose glucocorticosteroid treatment, followed by resolution of the inflammatory response. Failure to do so should raise the question of an alternative diagnosis.

Suggested tapering regimen:

- 40–60 mg prednisolone continued until symptoms and laboratory abnormalities resolve (at least 3–4 weeks);
- then dose is reduced by 10 mg every 2 weeks to 20 mg;
- then by 2.5 mg every 2–4 weeks to 10 mg; and
- then by 1 mg every 1–2 months provided there is no relapse.

Caso 2

- Biopsia N sural: vasculitis necrotizante de vaso de mediano calibre

- Revisión BAT: hiperplasia intimal. No infiltrado inflamatorio ni células gigantes.

- OD: PAN

Clinical Features and Outcomes in 348 Patients With Polyarteritis Nodosa

Christian Pagnoux,[1] Raphaèle Seror,[1] Corneliu Henegar,[2] Alfred Mahr,[1] Pascal Cohen,[1] Véronique Le Guern,[1] Boris Bienvenu,[3] Luc Mouthon,[1] and Loïc Guillevin,[1] for the French Vasculitis Study Group

ARTHRITIS & RHEUMATISM
Vol. 62, No. 2, February 2010, pp 616–626

		ed PAN)	HBV-related PAN (n = 123)	P†
		83/40 (2.1)		0.22
		0.9 ±		0.66
		8.9 ±		0.05
		209 (92.9)	115 (93.5)	0.83
Fever	222 (63.8)	136 (60.4)	86 (69.9)	0.08
Weight loss	242 (69.5)	149 (66.2)	93 (75.6)	0.07
Weight lost, mean ± SD kg	6.3 ± 6.5	5.4 ± 5.9	7.9 ± 7.2	0.001
Myalgias	204 (58.6)	139 (61.8)	65 (52.8)	0.11
Arthralgias	170 (48.9)	106 (47.1)	64 (52.0)	0.38
Neurologic manifestations	275 (79.0)	167 (74.2)	108 (87.8)	0.003
Peripheral neuropathy	258 (74.1)	153 (68.0)	105 (85.4)	<0.001
Mononeuritis multiplex	246 (70.7)	145 (64.4)	101 (82.1)	<0.001
Central nervous system	16 (4.6)	11 (4.9)	5 (4.1)	0.73
Urologic and renal manifestations	176 (50.6)	100 (44.4)	76 (61.8)	0.002
Hematuria	53 (15.2)	34 (15.1)	19 (15.4)	0.93
Proteinuria (>0.4 gm/24 hours)	75 (21.6)	46 (20.4)	29 (23.6)	0.50
Recent-onset hypertension	121 (34.8)	61 (27.1)	60 (48.8)	<0.001
Severe hypertension	24 (6.9)	11 (4.9)	13 (10.6)	0.05
Orchitis or testicular tenderness	38 (17.3)	18 (13.1)	20 (24.1)	0.02
Cutaneous manifestations	173 (49.7)	130 (57.8)	43 (35)	<0.001
Nodules	60 (17.2)	53 (23.6)	7 (5.7)	<0.001
Purpura	77 (22.1)	55 (24.4)	22 (17.9)	0.16
Livedo	58 (16.7)	45 (20.0)	13 (10.6)	0.02
Peripheral limb edema	85 (24.4)	50 (22.2)	35 (28.5)	0.20
Gastrointestinal manifestations	132 (37.9)	70 (31.1)	62 (50.4)	<0.001
Abdominal pain	124 (35.6)	62 (27.6)	62 (50.4)	<0.001
Bleeding	12 (3.4)	8 (3.6)	4 (3.3)	0.99
Perforation(s)	15 (4.3)	8 (3.6)	7 (5.7)	0.35
Cholecystitis	13 (3.7)	5 (2.2)	8 (6.5)	0.07
Appendicitis	4 (1.1)	2 (0.9)	2 (1.6)	0.62
Pancreatitis	13 (3.7)	6 (2.7)	7 (5.7)	0.23
Gastrointestinal manifestations requiring surgery	48 (13.8)	24 (10.7)	24 (19.5)	0.02
Cardiac and vascular manifestations	78 (22.4)	46 (20.4)	32 (26.0)	0.23
Vasculitis-related cardiomyopathy	26 (7.5)	10 (4.4)	16 (13.0)	0.004
Pericarditis	19 (5.5)	11 (4.9)	8 (6.5)	0.53
Digital ischemia (without necrotic lesions)	21 (6.0)	12 (5.3)	9 (7.3)	0.46
Distal necrotic lesions and/or limb arterial claudication	22 (6.3)	14 (6.2)	8 (6.5)	0.92
Ophthalmologic manifestations	30 (8.6)	17 (7.6)	13 (10.6)	0.34
Retinal vasculitis/exudate	15 (4.3)	8 (3.6)	7 (5.7)	0.35
Pulmonary manifestations‡				
Cough	20 (5.7)	14 (6.2)	6 (4.9)	0.61
Lung infiltrates	12 (3.4)	10 (4.4)	2 (1.6)	0.23
Pleural effusions	12 (3.4)	7 (3.1)	5 (4.1)	0.76
Periostitis	3 (0.9)	3 (1.3)	0	0.55
FFS = 0	208 (59.8)	150 (66.7)	58 (47.1)	0.002
FFS = 1	78 (22.4)	41 (18.2)	37 (30.1)	
FFS ≥2	62 (17.8)	34 (15.1)	28 (22.8)	
BVAS at diagnosis, mean ± SD	16.5 ± 8.3	15.1 ± 7.6	19.1 ± 9.1	<0.001

Table 2. Main biologic and radiologic characteristics at diagnosis of the 348 patients with PAN, by HBV status*

Characteristic	All patients (n = 348)	Non–HBV-related PAN (n = 225)	HBV-related PAN (n = 123)	P†
Laboratory findings				
Creatinine level >140 μmoles/liter‡	53 (15.2)	34 (15.1)	19 (15.4)	0.93
Creatinine, mean ± SD μmoles/liter‡	111.6 ± 88	105.6 ± 90	122.5 ± 84	0.12
Creatinine, mean ± SD mg/dl‡	1.26 ± 1.00	1.19 ± 1.02	1.39 ± 0.95	
C-reactive protein, mean ± SD mg/liter	101.5 ± 95	110.7 ± 101	73.8 ± 68	0.03
ESR >30 mm/hour§	267 (82.4)	176 (83.0)	91 (81.3)	0.69
Elevated transaminase level (AST and/or ALT)	114 (32.8)	35 (15.6)	79 (64.2)	<0.001
Angiography				
Gastrointestinal microaneurysms and/or stenoses¶	86 (57.7)	45 (48.9)	41 (71.9)	0.006
Kidney microaneurysms and/or stenoses#	104 (66.2)	59 (62.8)	45 (71.4)	0.26

3. ¿Qué tratamiento seria más adecuado para inducir la remisión de la enfermedad?
    1) Corticoides
    2) Corticoides + metotrexate
    3) Corticoides + CF oral
    4) Corticoides + CF iv (bolus)
    5) Corticoides + azatioprina

Treatment of Polyarteritis Nodosa and Microscopic Polyangiitis Without Poor-Prognosis Factors

A Prospective Randomized Study of One Hundred Twenty-Four Patients

Camillo Ribi,[1] Pascal Cohen,[1] Christian Pagnoux,[1] Alfred Mahr,[1] Jean-Pierre Arène,[1] Xavier Puéchal,[2] Philippe Carli,[3] Xavier Kyndt,[4] Claire Le Hello,[5] Philippe Letellier,[5] Jean-François Cordier,[6] and Loïc Guillevin,[1] for the French Vasculitis Study Group

ARTHRITIS & RHEUMATISM
Vol. 62, No. 4, April 2010, pp 1186–1197

Prospectivo, multicéntrico

Conclusion. **For patients with PAN or MPA with an FFS of 0, overall 5-year survival was good, but first-line corticosteroid treatment was able to achieve and maintain remission in only about half of the patients, and 40% of the patients required additional immunosuppressive therapy. Azathioprine or pulse cyclophosphamide was fairly effective for treating corticosteroid-resistant disease or major relapses.**

Table 2. Comparison of the Chapel Hill Consensus Conference (CHCC) Nomenclature and American College of Rheumatology (ACR) criteria for the classification of vasculitis. The ACR criteria are represented in an abbreviated form and are a combination of criteria in the traditional format and the classification tree, presented separately in the original papers.[44,45].

CHCC definition	ACR criteria	Comments
Giant-cell (temporal) arteritis (GCA)		
Large vessel vasculitis Granulomatous arteritis of the aorta and its major branches with a predilection for the extracranial branches of the carotid artery Often involves the temporal artery Patients usually >50 years May be associated with polymyalgia rheumatica	1. Age >50 years at onset 2. New type of headache 3. Claudication of jaw or tongue or on deglutition 4. Abnormal temporal artery on clinical examination 5. Scalp tenderness or nodules 6. Temporal artery biopsy showing vasculitis, or item (5.) can be used as a surrogate if biopsy is not available Criteria classify GCA with sensitivity 95.3% and specificity 90.7%	The CHCC definition of GCA includes patients who have inflammation of subclavian and brachial arteries.[64] Such cases do not meet ACR criteria for GCA. Vasculitis of the temporal artery may occur in WG but may be misclassified as GCA using ACR criteria.[65] Both CHCC and ACR criteria recognise that age of onset >50 years is an important feature of GCA. The ESR is non-specific and is not included in either the CHCC definition or ACR criteria, but is obviously useful in the *diagnosis* of GCA
Takayasu's arteritis (TA)		
Large vessel vasculitis Granulomatous inflammation of the aorta and its major branches Patients usually >50 years	1. Age >40 years at onset 2. Limb claudication 3. Diminished pulses 4. BP >10 mm Hg difference between arms 5. Bruits 6. Arteriogram abnormal Three criteria classify as TA with sensitivity 90.5% and specificity 97.8%	Patients with early symptoms such as myalgia and fever satisfy neither the CHCC definition nor ACR criteria for TA. An ESR >50 mm/hour is typical, but is not included in either the CHCC definition or ACR criteria, although useful in the *diagnosis* of TA. Both the CHCC and ACR recognize that age <50 years differentiates TA from GCA
Polyarteritis nodosa (PAN)		
Medium vessel vasculitis Necrotizing inflammation of medium-sized or small arteries without glomerulonephritis (GN) or vasculitis in arterioles, capillaries, or venules	1. Weight loss 2. Livedo reticularis 3. Testicular pain or tenderness 4. Myalgias, myopathy or tenderness 5. Neuropathy 6. Hypertension 7. Renal impairment 8. Hepatitis B virus 9. Abnormal arteriography 10. Biopsy of artery showing PMN Three criteria classify PAN with sensitivity 82.2% and specificity 86.6%	The CHCC decided that the name 'PAN' should be restricted to disease involving medium or small arteries, excluding those with smaller vessel involvement. PAN may therefore be exceedingly rare, if not non-existent (even Kussmaul and Maier's original patient had evidence of small vessel involvement!).[46,66] Patients who have GN or small vessel vasculitis are defined as MPA rather than PAN using CHCC.[67] The clinical diagnosis of PAN hinges on the findings at biopsy and/or arteriography. Note also that the HBV-associated PAN is included within this group of *primary* vasculitis by the ACR criteria

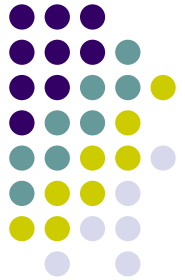
Table continued over page

Caso 2

- Tratamiento CF bolus (6) + cortis y posteriormente AZA hasta completar 18 meses, con remisión completa de la sintomatología. Stop tratamiento a los 24 meses.

- Durante seguimiento, 3 episodios de sobreinfección respiratoria + espectoración hemoptóica y un episodio de dolor abdominal periumbilical por los que se efectuaron TAC torácico + cultivos seriados de esputo, y angioTAC abdominal para descartar afección de la aorta abdominal y/o de la A Mesentérica

Respuestas correctas



- Pregunta 1: respuesta 2)
- Pregunta 2: respuesta 5)
- Pregunta 3: respuesta 4)