Varón de 82 años con neuritis óptica y lesiones isquémicas en cuero cabelludo

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VII CURSO DE ACTUALIZACIÓN EN PATOLOGÍA AUTOINMUNE

Marbella, 7- 8 de Noviembre 2014

Antecedentes personales:

- Diabetes mellitus 2:
  - Retinopatía
  - Microalbuminuria
- HTA
- Hipertrofia benigna de próstata.

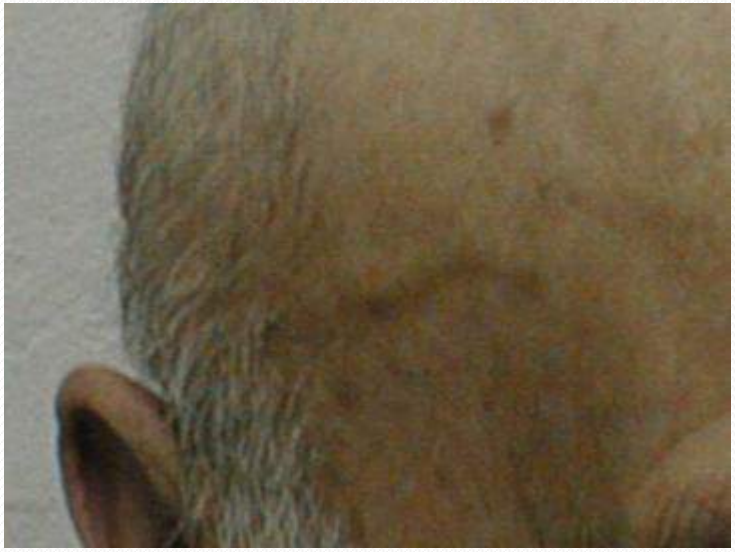
- TRATAMIENTO:
  - ARA II
  - Insulina
  - Estatina

Motivo de consulta:

- Pérdida súbita de agudeza visual ojo izquierdo.
- Oftalomólogo de guardia:
  - **NEURITIS ÓPTICA ISQUÉMICA**
- Cefalea occipital irradiada a región temporal derecha.
- Hiperalgesia cutánea.
- Dolor hemilengua derecha.
- Dolor y debilidad cinturas escapular y pelviana.

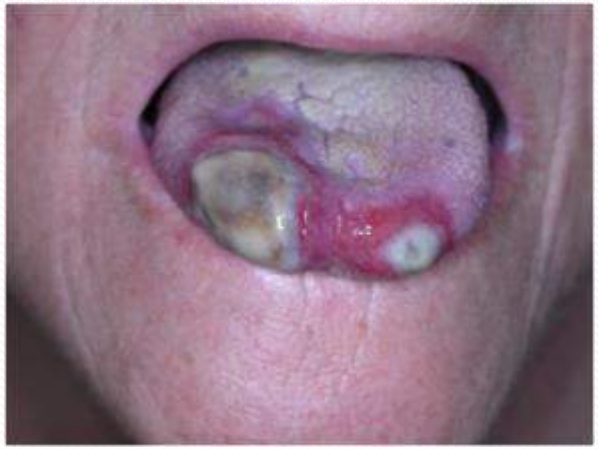
Exploración física:

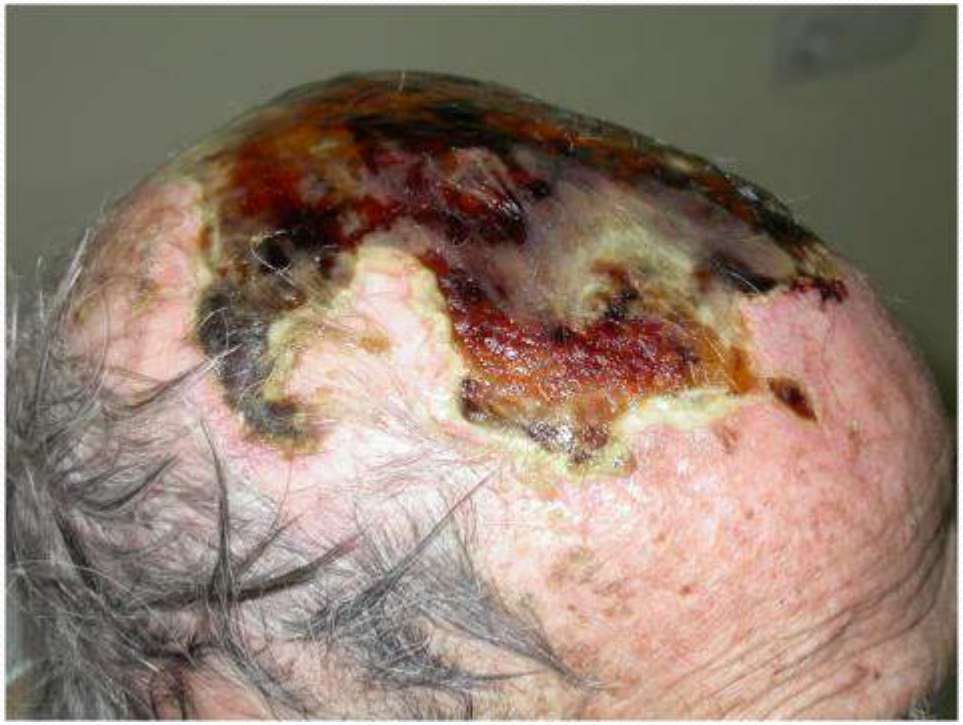
- Leve claudicación proximal de MMSS y MMII.
- Art. temporal dcha: engrosada, dolorosa y menor pulso que contralateral.
- Hiperestesia cutánea.



Exploración física:

- Lesiones isquémicas en región lateral derecha de la cabeza.





Pruebas complementarias:

LEUCOCITOS		10	10e3 µl	[4.8 - 10.8]
HEMATIES	*	3.53	10e6 µl	[4.7 - 6.1]
HEMOGLOBINA	*	10.8	gr/dl	[14 - 18]
HEMATOCRITO	*	32.3	%	[42 - 52]
VCM		91.3	fl	[80 - 94]
HCM		30.6	pg	[27 - 31]
CHCM		33.5	gr/dl	[33 - 37]
ADE		13.6	%	[11.5 - 14.5]
PLAQUETAS		258	10e3 µl	[130 - 400]
VPM		8.4	fl	[7.2 - 11.1]
NEUTROFILOS	*	89.5	%	[40 - 74]
LINFOCITOS	*	8.3	%	[19 - 48]
MONOCITOS		1.8	%	[1 - 9]
EOSINOFILOS		0	%	[0 - 7]
BASOFILOS		0.4	%	[0 - 2]

Pruebas complementarias:

	Resultado	Unidades	Valores de Referencia
BIOQUIMICA GENERAL			
GLUCOSA	* 234	mg/dl	[70 - 100]
CREATININA	1	mg/dl	[0.5 - 1.2]
BILIRRUBINA TOTAL	0.19	mg/dl	[0 - 1.2]
GPT	11	U/l	[10 - 40]
AMILASA TOTAL	76	U/L	[28 - 100]
SODIO	135	mEq/l	[135 - 148]
POTASIO	5	mEq/l	[3.8 - 5.2]
CLORO	98	mEq/l	[95 - 110]
CALCIO	10.1	mg/dl	[8.6 - 10.2]
PROTEINA C REACTIVA	* 40.35	mg/L	[0 - 5]

Juicio Clínico:

- **Neuritis óptica isquémica arterítica.**
- **Arteritis de la temporal**

Tratamiento:

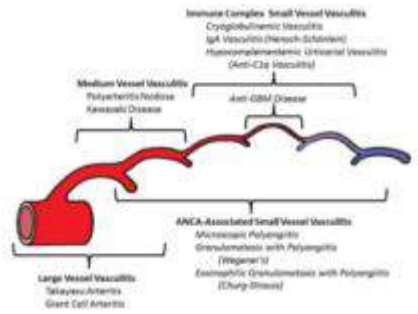
- **3 bolus de 6 metilprednisolona 500mg IV.**

1. ¿Cuál es la mejor denominación para esta vasculitis?

1. ¿Cuál es la mejor denominación para esta vasculitis?

2012 Revised International Chapel Hill Consensus Conference Nomenclature of Vasculitides

J. C. Jennette,[1] R. J. Falk,[1] P. A. Bacon,[2] N. Basu,[3] M. C. Cid,[4] F. Ferrario,[5] L. F. Flores-Suarez,[6] W. L. Gross,[7] L. Guillevin,[8] E. C. Hagen,[9] G. S. Hoffman,[10] D. R. Jayne,[11] C. G. M. Kallenberg,[12] P. Lamprecht,[13] C. A. Langford,[10] R. A. Luqmani,[14] A. D. Mahr,[15] E. L. Matteson,[16] P. A. Merkel,[17] S. Ozen,[18] C. D. Pusey,[19] N. Rasmussen,[20] A. J. Rees,[21] D. G. I. Scott,[22] U. Specks,[16] J. H. Stone,[23] K. Takahashi,[24] and R. A. Watts[25]



ARTHRITIS & RHEUMATISM
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Table 2. Names for vasculitides adopted by the 2012 International Chapel Hill Consensus Conference on the Nomenclature of Vasculitides

Large vessel vasculitis (LVV)
Takayasu arteritis (TAK)
Giant cell arteritis (GCA)

Giant cell arteritis (GCA). GCA is arteritis, often granulomatous and usually affecting the aorta and/or its major branches, with a predilection for the branches of the carotid and vertebral arteries. Giant cells are frequently but not always observed in biopsy specimens from patients with active GCA. The term "temporal arteritis" is not a suitable alternative for GCA because not all patients have temporal artery involvement, and other categories of vasculitis can affect the temporal arteries.

2. ¿Es posible distinguir una neuritis óptica isquémica de origen vasculítico de una no vasculítica en un paciente con múltiples factores de riesgo cardiovascular?

	Optic neuritis	Non-arteritic ischemic optic neuropathy	Arteritic ischemic optic neuropathy
Age	20 to 50 years	>50 years	>70 years
Gender	2:1 female	Equal	3.5:1 female
Pain	Present in >90 percent	Present in <10 percent	Headache, scalp tenderness, jaw claudication
Onset	Hours to days	Sudden	Sudden
Unilateral or bilateral	Usually unilateral	Usually unilateral; low chance may recur in other eye years later	May occur in both eyes in rapid sequence
Funduscopic examination	Papillitis present in one-third	Papillitis present in most	Pale swelling of disc; fundus may also be normal (posterior ischemic optic neuropathy - indicates giant cell arteritis)
Visual field defect	Central scotoma	Altitudinal (usually inferior) defect	Altitudinal or generalized constriction
Magnetic resonance imaging: optic nerve	Inflammation of optic nerve in most (one-third to one-half will have other demyelinating lesions)	Often normal	May show enhancement
Prognosis	Begins within two to four weeks, most achieve 20/40 or better	Over several months, only 40 percent improve by three or more lines	Poor once vision loss has occurred; may cause rapid blindness untreated

3. Y si la PCR hubiera sido normal ¿El planteamiento diagnóstico-terapéutico debería haber sido el mismo?

3. Y si la PCR hubiera sido normal ¿El planteamiento diagnóstico-terapéutico debería haber sido el mismo?

How common is inflammatory marker-negative disease in giant cell arteritis?

are commonly used to aid diagnosis of GCA. CRP has been reported to be a more sensitive predictor of the disease than ESR (97.5–100% for CRP vs 76–92% for ESR).[2,3] Our case is unique in that it belongs to the rarer

Table 1 Summary of current literature

	ESR-negative disease (%)	*CRP-negative disease (%)*	*ESR- and CRP-negative disease (%)*
Parikh *et al*[3]	14.3	1.7	0.8
Poole *et al*[5]			1 Case report
Levy *et al* (current study)		1 Case report	

4. ¿Qué pasa si es arterítica y no usamos corticoides? ¿Y si no lo es y los usamos?

The NEW ENGLAND JOURNAL of MEDICINE

CLINICAL PRACTICE

Caren G. Solomon, M.D., M.P.H., *Editor*

Giant-Cell Arteritis and Polymyalgia Rheumatica

Cornelia M. Weyand, M.D., Ph.D., and Jörg J. Goronzy, M.D., Ph.D.

N ENGL J MED 371;1 NEJM.ORG JULY 3, 2014

1 mg per kilogram of body weight per day. Given the risk of irreversible ischemic complications, new-onset clinical manifestations of disease indicating an unstable supply of blood to the eyes or the central nervous system (e.g., arteritic optic neuropathy) are typically managed with intravenous pulse therapy (e.g., 1000 mg of methylprednisolone per day for 3 consecutive days) to optimize immunosuppression and suppress tissue edema. Once tissue necrosis occurs (e.g., optic-nerve isch
with blindness for several hours), it is irrevers

Visual outcome of mega-dose intravenous corticosteroid treatment in non-arteritic anterior ischemic optic neuropathy – retrospective analysis

Michael Kinori[1,2*], Iris Ben-Bassat[1], Yael Wasserzug[1], Angela Chetrit[3] and Ruth Huna-Baron[1]

Table 1 Main demographic and clinical data of the treated and control group

	Treated group	Control group	P value
Male: female	14:10	16:8	0.55
Age (years)	54.4 ± 12.3	55.4 ± 9.6	0.78
Follow-up (months)	22.7 ± 23.4	36.2 ± 24.2	0.04
Number vascular risk factors*	1.4 ± 1.1	1.6 ± 1.1	0.60
Crowded disc	10 (42%)	13 (54%)	0.31
Mean VA (LogMAR)	0.54 ± 0.67	0.54 ± 0.49	0.80
Visual field parameters			
Mean quadrant involvement	2.4 ± 0.8	2.0 ± 0.6	0.007
Mean MD	9.7 ± 10.4	9.3 ± 10.5	0.9

VA = Visual acuity, MD = Mean deviation.
*Includes: Hypertention, diabetes mellitus, ischemic heart disease, dyslipidemia and smoking.

Conclusions: Our results suggest that IV corticosteroids may not improve the visual outcome of NAION patients. Since intravenous corticosteroids could potentially cause serious adverse effects, this treatment for NAION is questionable.

5. ¿Tendría que antiagregarlo? ¿Siempre o solo si bolus de corticoides?

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Cochrane Database Syst Rev. 2014 Aug

Conclusions

At the present time there is not enough data to make a comment on whether aspirin is of benefit in GCA. More research is needed.

6. Los médicos que asistieron a este paciente el lunes, solicitaron una ecografía de arterias temporales que se hizo 2 días más tarde. ¿Qué hallazgos esperaríamos encontrar?

7. ¿Modificarían los hallazgos de la eco el procedimiento diagnóstico tanto a la hora de indicar la realización de una biopsia, como del sitio de su realización?

Meta-analysis

British Journal of Surgery 2010; **97**: 1765–1771

Role of ultrasonography in the diagnosis of temporal arteritis

E. L. Ball, S. R. Walsh, T. Y. Tang, R. Gohil and J. M. F. Clarke

Results: There were 17 eligible studies containing 998 patients. When the halo sign on duplex imaging was compared with TA biopsy, the sensitivity was 75 (95 per cent confidence interval 67 to 82) per cent and the specificity was 83 (78 to 88) per cent. There was no heterogeneity across the eligible studies.

Conclusion: Duplex ultrasonography was relatively accurate for diagnosing temporal arteritis. It should become the first-line investigation, with biopsy reserved for patients with a negative scan.

Pathological Analysis

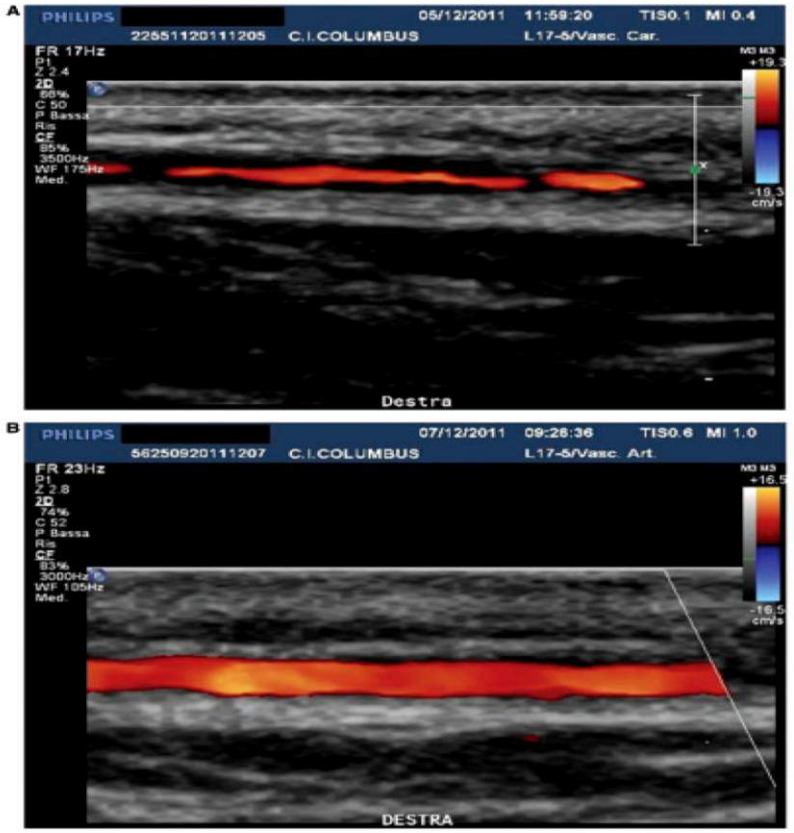
In cases of suspected giant-cell arteritis, histologic verification of vasculitis should be sought by means of a temporal-artery biopsy with assessment of a vascular segment that is 1.5 to 2.0 cm in length. Histologic analysis is the standard for diagnosis; it

Clinical vignette

Temporal ultrasonography findings in temporal arteritis: early disappearance of halo sign after only 2 days of steroid treatment

Rheumatology 2013;52:622

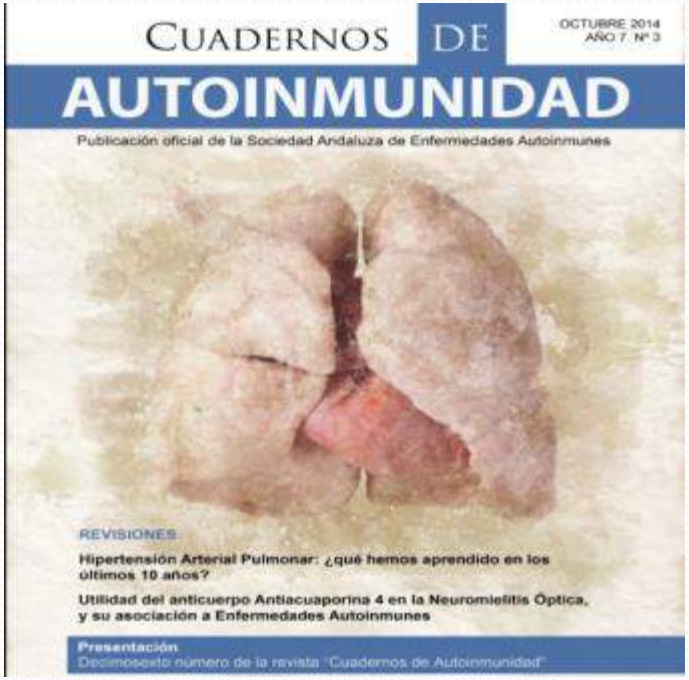
FIG. 1 Right temporal artery CDU features before (**A**) and after (**B**) 2 days of steroid treatment.



Steroids induced early disappearance of the halo sign, leading to normalization of CDU.

Is colour duplex sonography-guided temporal artery biopsy useful in the diagnosis of giant cell arteritis? A randomized study

Giuseppe Germanò[1], Francesco Muratore[1], Luca Cimino[2], Alberto Lo Gullo[3], Niccolò Possemato[1], Pierluigi Macchioni[1], Alberto Cavazza[4], Nicolò Pipitone[1], Luigi Boiardi[1] and Carlo Salvarani[1]



VASCULITIS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Manuela Moreno Higueras

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ECO-DOPPLER COLOR VS. PALPACIÓN DE LA ARTERIA TEMPORAL PARA ELECCIÓN DE LA BIOPSIA: "EL PODER ESTÁ EN TUS DEDOS"

Germano G, Muratore F, Cimino L, et al. Rheumatology (Oxford). 2014 Jun 17. pii: keu241.

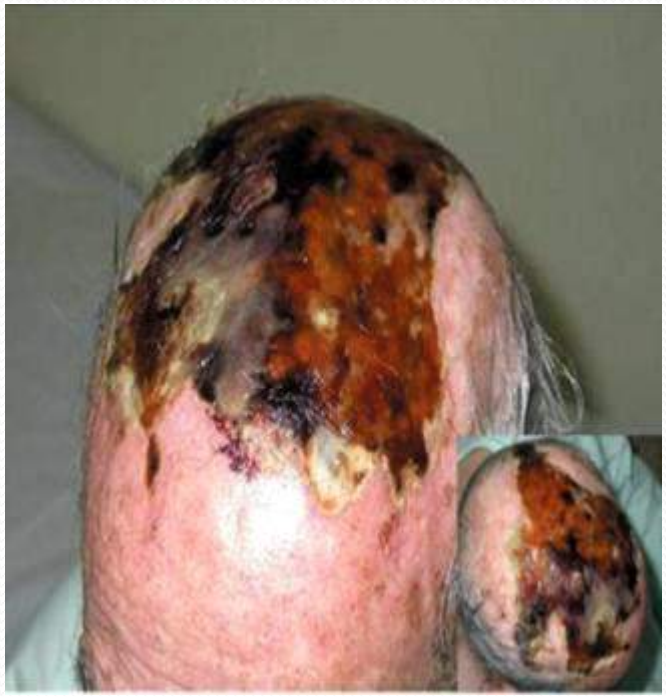
In conclusion, our study shows that CDS-guided TAB does not improve the sensitivity of TAB for diagnosing GCA. In the presence of the halo sign on CDS, the probability of a positive TAB is high, regardless of whether the biopsy in guided by CDS.

8. ¿A qué atribuirías las lesiones de la cabeza y la lengua?

Scalp necrosis as a manifestation of temporal arteritis

International Journal of Dermatology 2010, **49**, 466–474

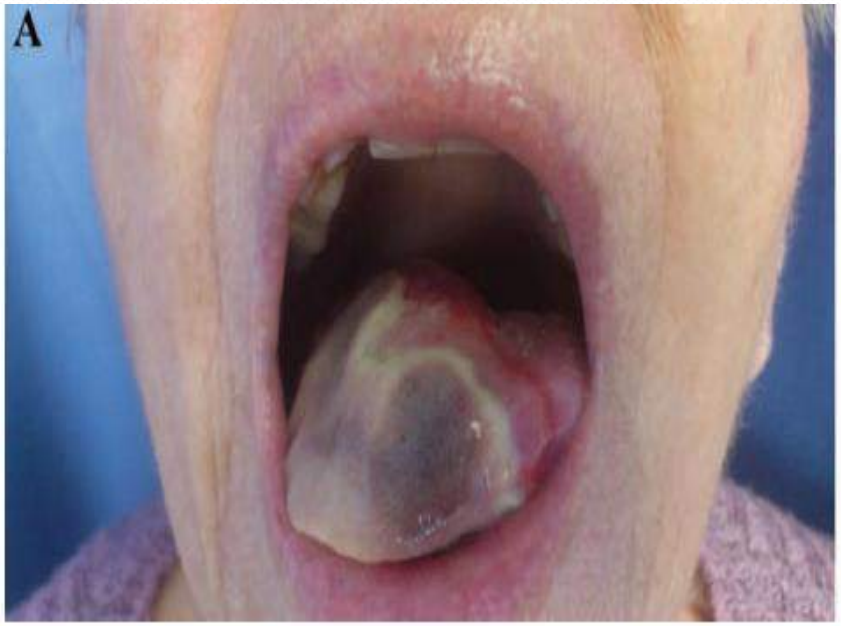
Cristina Serrano-Falcón, MD
Maria del Mar Serrano-Falcón, MD
Jose Luis Callejas-Rubio, MD
Salvio Serrano-Ortega, MD
Granada, Spain

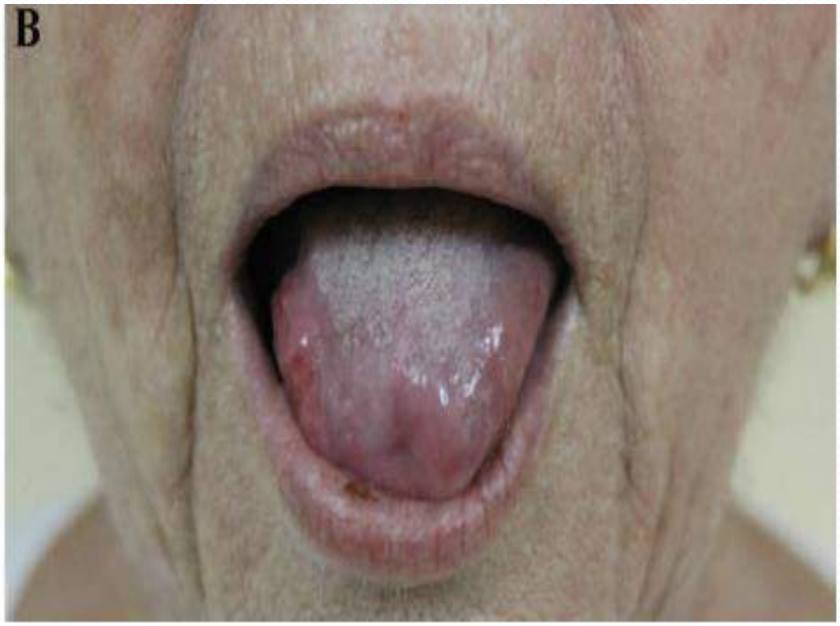


8. ¿A qué atribuirías las lesiones de la cabeza y la lengua?

Tongue necrosis: an unusual clinical presentation of giant cell arteritis

Arthritis Rheumatol. 2014 Oct;66(10):2803.





9. Dado los FRCV, ¿Usarías metotrexate como ahorrador de esteroides?

ARTHRITIS & RHEUMATISM
Vol. 56, No. 8, August 2007, pp 2789–2797

Adjunctive Methotrexate for Treatment of Giant Cell Arteritis

An Individual Patient Data Meta-Analysis

Alfred D. Mahr,[1] Juan A. Jover,[2] Robert F. Spiera,[3] César Hernández-García,[2] Benjamin Fernández-Gutiérrez,[2] Michael P. LaValley,[4] and Peter A. Merkel[1]

Conclusion. **In GCA, adjunctive treatment with MTX lowers the risk of relapse and reduces exposure to corticosteroids. These findings indicate that MTX could be considered as a therapeutic option in addition to standard-of-care treatment with corticosteroids for patients with GCA.**

Prednisolone combined with adjunctive immunosuppression is not superior to prednisolone alone in terms of efficacy and safety in giant cell arteritis: meta-analysis.

Clin Rheumatol 2014 Feb;33(2):227-36.

Yates M, Loke YK, Watts RA, MacGregor AJ.

- Meta-analysis of published data of the effectiveness of drug treatment in giant cell arteritis (GCA) to provide evidence to support the optimal use of glucocorticoids (GCs) and adjunct therapy.
- Together these comprised 638 participants of which 72 % were female.
- Three studies compared various GCs regimens, with two comparing IV GCs, the latter showing a marginal benefit with respect to relapse but a greater risk of infection
- Three used methotrexate as an adjunctive agent and showed marginal benefit with respect to relapse
- The remaining four trials compared prednisolone to dapsone, infliximab, adalimumab and hydroxychloroquine, respectively.

The results from this meta-analysis show that the use of adjunct agents is not associated with improved outcome.

10. ¿Sería necesario realizar estudio de imagen aórtico en este paciente?

Clinical and epidemiological research

EXTENDED REPORT

Should I send my patient with previous giant cell arteritis for imaging of the thoracic aorta? A systematic literature review and meta-analysis

Sarah Louise Mackie,[1] Elizabeth M A Hensor,[1] Ann W Morgan,[1] Colin T Pease[2]

Results Two analyses of routinely collected administrative data suggested a threefold risk of TAA/dissection in GCA compared with controls. In GCA cohorts without systematic imaging, 2–8% had TAA. In the two best-reported studies, aneurysm dissection/rupture occurred in 1% and 6% of GCA cases. Aortic imaging studies had a variety of TAA/TAD definitions, imaging methods and time points. There were limited data on age-matched controls. Three studies suggested that male sex may be a risk factor for TAA/TAD in GCA. On average, five to ten patients with GCA would need aortic imaging to detect one previously unknown TAA/TAD.

Conclusions The data support an association between GCA and TAA/TAD compared with age-matched controls, but the true relative risk, and the time course of that risk, remains unclear. It is also unclear whether chest radiography is a sufficiently sensitive screening tool. Clinicians should retain a high index of suspicion for aortic pathology in patients with GCA. Before ordering imaging, clinicians should consider whether, and how, detecting aortic pathology would affect a patient's management.

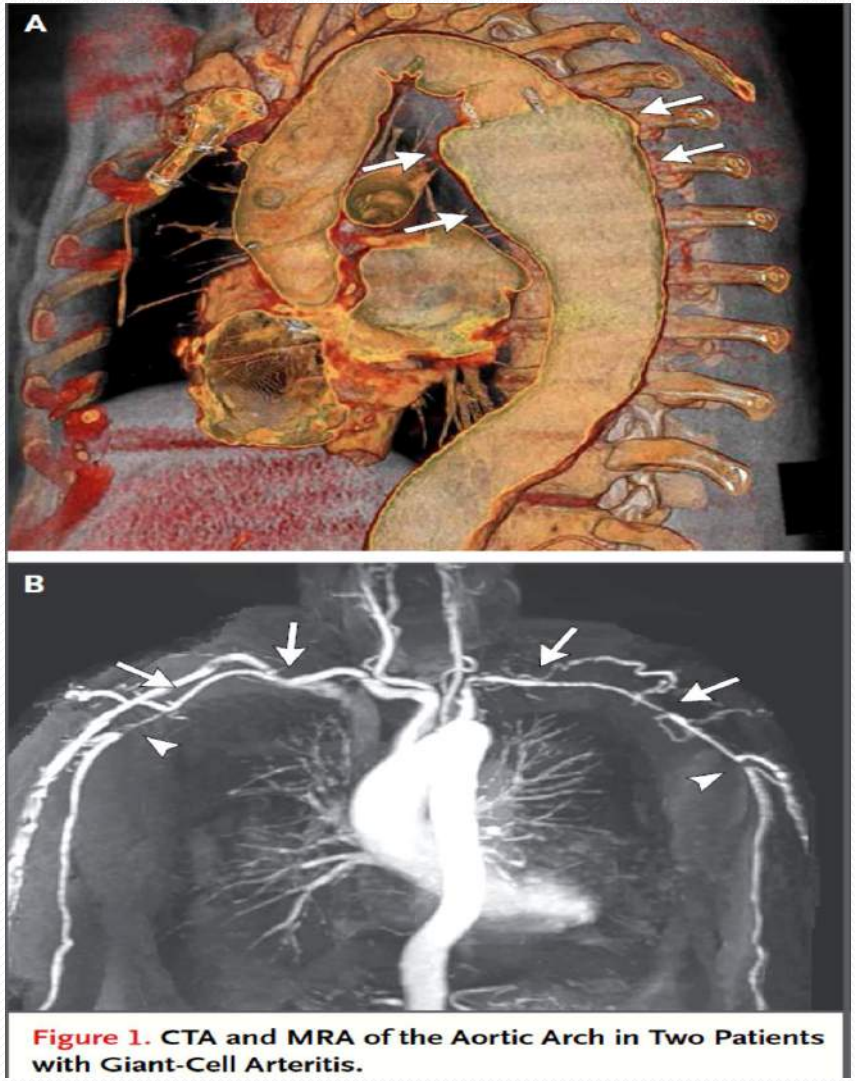


Figure 1. CTA and MRA of the Aortic Arch in Two Patients with Giant-Cell Arteritis.

Review Article

Giant Cell Arteritis: A Systematic Review of the Qualitative and Semiquantitative Methods to Assess Vasculitis with 18F-Fluorodeoxyglucose Positron Emission Tomography

Cristina Puppo,[1] Michela Massollo,[2] Francesco Paparo,[1] Dario Camellino,[3] Arnoldo Piccardo,[2] Mehrdad Shoushtari Zadeh Naseri,[2] Giampiero Villavecchia,[2] Gian Andrea Rollandi,[1] and Marco Amedeo Cimmino[3]

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5. Conclusion

In conclusion, 18F-FDG PET has been shown to have an important role in the diagnosis of extracranial vascular involvement in patients with GCA/PMR.

Qualitative methods are more specific than semiquantitative ones, but they have lower sensitivity. The aortic-to-blood pool uptake ratio is a promising semiquantitative method of analysis for the detection and grading of arterial inflammation. The normalization of the arterial wall uptake to the background activity of venous blood pool provides a good reference to assess vascular inflammation. Further prospective studies involving larger cohorts of GCA/PMR patients are required to better define the role of aortic-to-blood pool ratio as a reference method for the assessment of vasculitis in GCA patients.