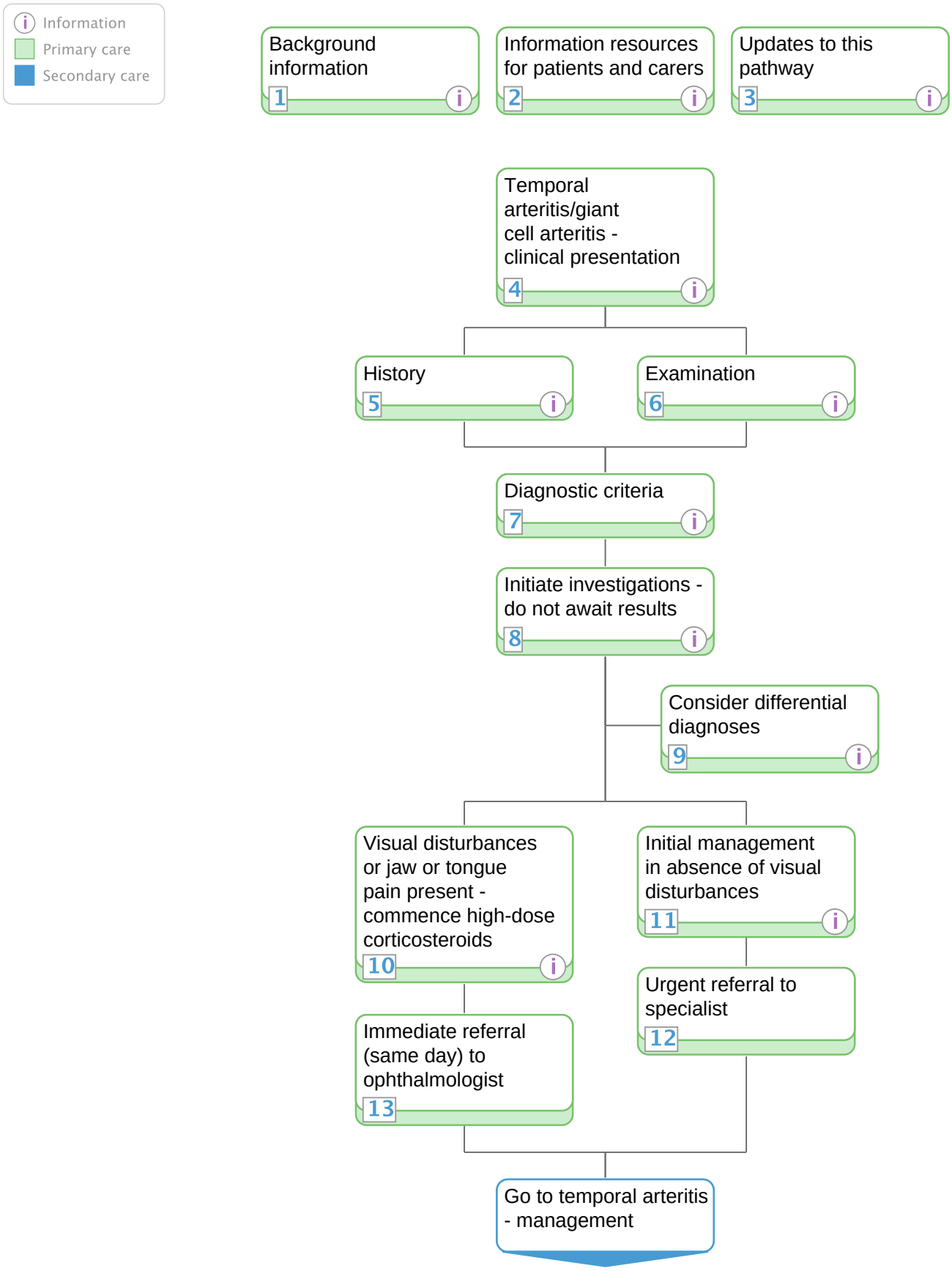
Temporal arteritis/giant cell arteritis - suspected



Published: 29-Jul-2010 Valid until: 31-May-2012

Temporal arteritis/giant cell arteritis - suspected

1 Background information

Quick info:

Scope:

- diagnosis, management, and complications of temporal arteritis in adults, across primary and secondary care settings
- consideration of temporal artery biopsy
- consideration of therapy risks associated with corticosteroids (especially initial high dose therapy)
- new imaging modalities for diagnosis of large vessel vasculitis

Out of scope:

- management and diagnosis of polymyalgia rheumatica (PMR) – see 'Polymyalgia rheumatica' pathway

Definition:

- known as temporal arteritis, giant cell arteritis (GCA), or cranial arteritis
- type of vasculitis that leads to granulomatous inflammation in wall of medium and large arteries and occlusion of lumen by intimal hyperplasia
- predominantly affects extracranial branches of carotid artery and branches of ophthalmic artery – occasionally affects the aorta itself
- can be associated with significant early irreversible neuro-ophthalmic complications (vision loss and stroke) and is therefore a medical emergency
- cause is unknown

Incidence:

- it is the most common systemic vasculitis in Western countries
- incidence is reported as 22/100,000 per year in the UK [1]

Risk factors:

- rare before age 50 years – mean onset is at age 70 years
- three times more common in females than males
- seven times more common in white people than black people

Prognosis:

- vision loss (total or partial) occurs in up to 20% of those with temporal arteritis – recovery of vision is rare [1]
- other significant complications include cardiovascular disease and large artery complications such as aortic aneurysm
- complications are usually prevented through high dose corticosteroid treatment
- relapses are common, especially if corticosteroid dose is reduced too quickly

NB: As up to 40% of people with temporal arteritis also have symptoms of PMR, there are suggestions that the underlying pathophysiology is the same, however there is no firm evidence at present to support this idea [1].

NB: This information appears on each page of this pathway.

References:

[1] Clinical Knowledge Summaries (CKS). Giant cell arteritis. Version 1.1. Newcastle upon Tyne: CKS; 2009.

[2] Dasgupta B, Borg FA, Hassan N et al on behalf of the BSR and BHPR Standards, Guidelines and Audit Working Group. BSR and BHPR Guidelines for the management of giant cell arteritis. Oxford: British Society for Rheumatology; 2010.

[3] Map of Medicine (MoM). London: MoM; 2010.

[4] Scottish Intercollegiate Guidelines Network (SIGN). Diagnosis and management of headache in adults. Edinburgh; SIGN: 2008.

2 Information resources for patients and carers

Quick info:

Patients and carers in England and Wales can access this pathway through NHS Choices at http://healthguides.mapofmedicine.com/choices/map/temporal_arteritis_giant_cell_arteritis1.html

The following resources have been produced by organisations certified by The Information Standard:

- Arthritis Care at http://www.arthritiscare.org.uk
- 'Giant cell arteritis' (URL) from eMC Medicine Guides at www.medguides.medicines.org.uk
- 'Giant cell arteritis (Temporal arteritis)' (PDF) from Patient UK at http://www.patient.co.uk
- 'Preventing steroid-induced osteoporosis' (PDF) from Patient UK at http://www.patient.co.uk
- 'Steroid tablets' (PDF) from Patient UK at http://www.patient.co.uk

The following resources have been written or recommended by national policy bodies or guideline producers whose content has informed this pathway:

- 'Corticosteroid (drugs)' patient leaflet (URL) from Clinical Knowledge Summaries (CKS) at http://www.cks.nhs.uk
- 'Osteoporosis' patient leaflet (URL) from CKS at http://www.cks.nhs.uk

Published: 29-Jul-2010 Valid until: 31-May-2012

Temporal arteritis/giant cell arteritis - suspected

- 'Polymyalgia rheumatica and giant cell arteritis' patient leaflet (URL) from CKS at http://www.cks.nhs.uk

Information for carers and people with disabilities is available at:

- 'Caring for someone' (URL) from Directgov at http://www.direct.gov.uk
- 'Disabled people' (URL) from Directgov at http://www.direct.gov.uk

Explanations of clinical laboratory tests used in diagnosis and treatment are available at 'Understanding Your Tests' (URL) from Lab Tests Online-UK at http://www.labtestsonline.org.uk

The Map of Medicine is committed to providing high quality health and social care information for patients and carers. For details on how these resources are identified, please see our Policy on Information Resources for Patients and Carers.

NB: This information appears on each page of this pathway.

3 Updates to this pathway

Quick info:

Date of publication: 30-Jul-2010

Interim update: A link node to the 'Venous thromboembolism (VTE) risk assessment' pathway has been included at relevant points to encourage secondary care providers to risk assess patients for VTE on admission to hospital.

Date of publication: 29-April-2010

Three floating nodes now appear at the top of each pathway page. These provide:

- easy access to scope and background information on each page of the pathway whilst reducing repetition between nodes
- easy access to patient resources/leaflets
- information on pathway updates

This pathway was updated in line with the following guidelines:

- [1] Clinical Knowledge Summaries (CKS). Giant cell arteritis. Version 1.1. Newcastle upon Tyne: CKS; 2009.
- [2] Dasgupta B, Borg FA, Hassan N et al on behalf of the BSR and BHPR Standards, Guidelines and Audit Working Group. BSR and BHPR Guidelines for the management of giant cell arteritis. Oxford: British Society for Rheumatology; 2010.
- [4] Scottish Intercollegiate Guidelines Network (SIGN). Diagnosis and management of headache in adults. Edinburgh; SIGN: 2008.

Further information was provided by the following references: [3,5,6]. For more information please see the pathway's Provenance certificate.

The pathway has been completely restructured and redrafted in line with the Map of Medicine's editorial methodology and to bring in line with current clinical practice.

NB: This information appears on each page of this pathway.

4 Temporal arteritis/giant cell arteritis - clinical presentation

Quick info:

Presentation of temporal arteritis can include any of the following:

- new-onset headache with severe pain:
  - usually unilateral and localised to the temporal area, occasionally diffuse or bilateral, generally of sudden onset [2]
  - typically localised to the temple but may be localised to the occipital area or may be diffuse [1]
  - can be diffuse [4]
- jaw and tongue claudication (high predictor)
- visual disturbances (often monocular) such as diplopia, blurring, amaurosis fugax, or vision loss
- scalp pain and tenderness
- systemic symptoms (may have been present for some time), eg:
  - fever (may be the only presenting feature)
  - weight loss
  - loss of appetite
  - depression
  - fatigue
- peripheral arthritis and distal swelling with pitting oedema
- respiratory tract symptoms are present in 10% of people and may be caused by arteritis, eg [1]:
  - cough
  - sore throat
  - hoarse voice
- symptoms of polymyalgia rheumatica (PMR), eg severe bilateral muscle pain:
  - symptoms of PMR are present in up to 40% of people with temporal arteritis [1]

Published: 29-Jul-2010 Valid until: 31-May-2012

Temporal arteritis/giant cell arteritis - suspected

- see 'Polymyalgia rheumatica' pathway

NB: Patient is usually over age 50 years and the condition is three times more common in females than males.

This information was drawn from the following references:

[1] Clinical Knowledge Summaries (CKS). Giant cell arteritis. Version 1.1. Newcastle upon Tyne: CKS; 2009.

[2] Dasgupta B, Borg FA, Hassan N et al on behalf of the BSR and BHPR Standards, Guidelines and Audit Working Group. BSR and BHPR Guidelines for the management of giant cell arteritis. Oxford: British Society for Rheumatology; 2010.

[4] Scottish Intercollegiate Guidelines Network (SIGN). Diagnosis and management of headache in adults. Edinburgh; SIGN: 2008.

5 History

Quick info:

- onset and duration of symptoms
- nature of headaches:
  - frequency and duration
  - severity
  - location
- other neurological features
- visual disturbances
- nature and duration of any systemic symptoms
- respiratory tract symptoms
- symptoms of arteritis elsewhere
- previous medical history, especially polymyalgia rheumatica (PMR)
- current medications

This information was drawn from the following references:

[1] Clinical Knowledge Summaries (CKS). Giant cell arteritis. Version 1.1. Newcastle upon Tyne: CKS; 2009.

[2] Dasgupta B, Borg FA, Hassan N et al on behalf of the BSR and BHPR Standards, Guidelines and Audit Working Group. BSR and BHPR Guidelines for the management of giant cell arteritis. Oxford: British Society for Rheumatology; 2010.

6 Examination

Quick info:

Examine for common signs of temporal arteritis:

- in up to 75% of people, temporal arteries are visibly thickened, tender, and nodular [1]:
  - occasionally the overlying skin is red, and pulsation may be reduced or absent
- bruits may be heard over the carotid, subclavian, axillary, and brachial arteries
- abnormal fundoscopy – the following features are usually seen following loss of vision:
  - small haemorrhages in the retina
  - pallor and oedema of the optic disc
  - 'cotton-wool' patches
- relative afferent papillary defect present on swinging flashlight test
- visual field defects
- neurological features occur in about 30% of people [1]:
  - mononeuropathy or polyneuropathy of arms or legs
  - transient ischaemic attacks in the distribution of the carotid or vertebrobasilar arteries
- in approximately 25% of people peripheral arthritis and pitting oedema of the extremities occurs [1]:
  - swelling is most prominent over the dorsum of the hands and wrists, the ankles, and the tops of the feet
- in approximately 40% of people symptoms of polymyalgia rheumatica (PMR) are present, eg [1]:
  - bilateral upper arm stiffness, aching, or tenderness
- different blood pressure measurements in the upper limbs and impaired peripheral pulses may indicate large vessel involvement

Other complications include:

- later development of thoracic aortic aneurysm
- aortic dissection

This information was drawn from the following references:

[1] Clinical Knowledge Summaries (CKS). Giant cell arteritis. Version 1.1. Newcastle upon Tyne: CKS; 2009.

[2] Dasgupta B, Borg FA, Hassan N et al on behalf of the BSR and BHPR Standards, Guidelines and Audit Working Group. BSR and BHPR Guidelines for the management of giant cell arteritis. Oxford: British Society for Rheumatology; 2010.

Published: 29-Jul-2010 Valid until: 31-May-2012

Temporal arteritis/giant cell arteritis - suspected

7 Diagnostic criteria

Quick info:

Several sets of criteria for diagnosing temporal arteritis have been proposed – the set suggested by the American College of Rheumatology may be the easiest to apply, requiring the presence of three or more of the following [1,2]:

- onset at or over age 50 years
- new type of headache
- thickened, tender, or nodular temporal artery with decreased pulsation
- erythrocyte sedimentation rate (ESR) 50mm per hour or greater
- abnormal arterial biopsy showing necrotising arteritis with mononuclear infiltrate or granulomatous inflammation, usually with multinucleated giant cells

The presence of any three of more of the above criteria is reported as having sensitivity and specificity above 90% for temporal arteritis [1].

If patient has symptoms or signs suggestive of temporal arteritis, start treatment while awaiting results of investigations [3].

References:

[1] Clinical Knowledge Summaries (CKS). Giant cell arteritis. Version 1.1. Newcastle upon Tyne: CKS; 2009.

[2] Dasgupta B, Borg FA, Hassan N et al on behalf of the BSR and BHPR Standards, Guidelines and Audit Working Group. BSR and BHPR Guidelines for the management of giant cell arteritis. Oxford: British Society for Rheumatology; 2010.

[3] Map of Medicine (MoM). London: MoM; 2010.

8 Initiate investigations - do not await results

Quick info:

If patient has symptoms or signs suggestive of temporal arteritis, start treatment while awaiting results of investigations [3].

In primary care, the initial diagnosis of temporal arteritis is based mainly on symptoms and signs although the following investigations may support the diagnosis [1]:

- erythrocyte sedimentation rate (ESR) – often greater than 50 mm per hour but up to 10% of people present with a rate lower than this [1]
- C-reactive protein (CRP) – usually elevated [1]
- low levels of ESR or CRP do not exclude diagnosis of giant cell arteritis (GCA) within a suggestive clinical context [1]
- combining the two tests gives higher specificity (97%) than CRP alone (83% in males, 79% in females) [4]
- plasma viscosity is sometimes used in diagnosing temporal arteritis, although there is no evidence of effectiveness [4]

Other abnormal tests may include:

- normochromic normocytic anaemia [1]
- raised platelet count [1]
- abnormal liver function tests, particularly alkaline phosphatase [1]
- urinalysis [2]
- chest X-ray [2]

Ultrasonography may help in diagnosing temporal arteritis, but requires cautious interpretation based on clinical presentation – suggestive findings are [5]:

- halo around temporal artery lumen
- stenosis and occlusion of the temporal arteries

References:

[1] Clinical Knowledge Summaries (CKS). Giant cell arteritis. Version 1.1. Newcastle upon Tyne: CKS; 2009.

[2] Dasgupta B, Borg FA, Hassan N et al on behalf of the BSR and BHPR Standards, Guidelines and Audit Working Group. BSR and BHPR Guidelines for the management of giant cell arteritis. Oxford: British Society for Rheumatology; 2010.

[3] Map of Medicine (MoM). London: MoM; 2010.

[4] Scottish Intercollegiate Guidelines Network (SIGN). Diagnosis and management of headache in adults. Edinburgh; SIGN: 2008.

[5] Karassa FB, Matsagas MI, Schmidt WA et al. Meta-analysis: test performance of ultrasonography for giant-cell arteritis. Ann Intern Med 2005;142: 359-69.

9 Consider differential diagnoses

Quick info:

Published: 29-Jul-2010 Valid until: 31-May-2012

Temporal arteritis/giant cell arteritis - suspected

Temporal arteritis/giant cell arteritis (GCA) should be considered and excluded in all patients over age 50 years who present with a new headache or change in headache [4]:

- any patient with jaw claudication and headache should be considered to have temporal arteritis until proven otherwise

Temporal arteritis may atypically present without headaches, prominent jaw/tongue pain only, low acute phase markers, severe polymyalgia and occipital pain – these atypical presentations should be carefully considered while excluding differential diagnoses [2].

Differential diagnoses include:

- herpes zoster [1,2]
- migraine [1,2]
- cluster headache [1,2]
- transient ischaemic attack [1,2]
- temporomandibular joint pain [1,2]
- cervical spondylosis [1,2]
- sinus disease [2]
- ear problems [2]
- cranial malignancy [1]
- ankylosing spondylitis [1]
- acute angle closure glaucoma [1]
- primary systemic vasculitis [3]

NB: There is usually little difficulty in distinguishing temporal arteritis from other forms of arteritis [1].

References:

[1] Clinical Knowledge Summaries (CKS). Giant cell arteritis. Version 1.1. Newcastle upon Tyne: CKS; 2009.

[2] Dasgupta B, Borg FA, Hassan N et al on behalf of the BSR and BHPR Standards, Guidelines and Audit Working Group. BSR and BHPR Guidelines for the management of giant cell arteritis. Oxford: British Society for Rheumatology; 2010.

[3] Map of Medicine (MoM). London: MoM; 2010.

[4] Scottish Intercollegiate Guidelines Network (SIGN). Diagnosis and management of headache in adults. Edinburgh; SIGN: 2008.

10 Visual disturbances or jaw or tongue pain present - commence high-dose corticosteroids

Quick info:

Lack of recognition and delay in treatment may lead to irreversible loss of vision – blindness in giant cell arteritis (GCA) almost always occurs prior to steroid therapy [1]:

- immediate referral to ophthalmologist is imperative [1]
- local referral variations may apply – direct access to ophthalmology may be available or the patient may be referred to A&E [3]

British Society for Rheumatology (BSR) guidelines recommend that for complicated temporal arteritis (jaw or tongue claudication or visual symptoms) [2]:

- for evolving visual loss or history of amaurosis fugax – intravenous (IV) methylprednisolone 500mg to 1g daily for 3 days, then oral corticosteroids
- for established vision loss – at least 60mg prednisolone daily

Clinical Knowledge Summaries (CKS) suggest [1]:

- initiate treatment immediately with high-dose corticosteroids – 60mg oral prednisolone daily

Consider giving low-dose daily aspirin to all people with temporal arteritis to protect against ischaemic cranial complications, if there are no contra-indications (eg active peptic ulceration or bleeding disorder) [1,2].

A proton pump inhibitor (PPI; eg omeprazole) is recommended for gastrointestinal protection (low-dose aspirin and high-dose prednisolone are both associated with significant gastrointestinal toxicity) [1,2].

References:

[1] Clinical Knowledge Summaries (CKS). Giant cell arteritis. Version 1.1. Newcastle upon Tyne: CKS; 2009.

[2] Dasgupta B, Borg FA, Hassan N et al on behalf of the BSR and BHPR Standards, Guidelines and Audit Working Group. BSR and BHPR Guidelines for the management of giant cell arteritis. Oxford: British Society for Rheumatology; 2010.

[3] Map of Medicine (MoM). London: MoM; 2010.

11 Initial management in absence of visual disturbances

Quick info:

Immediately initiate treatment [1,2]:

Published: 29-Jul-2010 Valid until: 31-May-2012

Temporal arteritis/giant cell arteritis - suspected

- high dose corticosteroids – 40-60mg oral prednisolone daily:
  - British Society of Rheumatology guidelines suggests a minimum of 0.75mg/kg [2]
  - Clinical Knowledge Summaries (CKS) guidance is a minimum of 0.7mg/kg [1]
- consider low-dose daily aspirin to protect against ischaemic cranial complications, if there are no contra-indications (eg active peptic ulceration or bleeding disorder) [1,2]
- start a proton pump inhibitor (PPI; eg omeprazole) for gastrointestinal protection (low-dose aspirin and high-dose prednisolone are both associated with significant gastrointestinal toxicity) [1,2]
- offer adequate analgesia for headache [3]

Establish a baseline against which to assess response to treatment by documenting [1]:

- current symptoms, signs, and any disability
- symptoms or disability present before the onset of the current problem
- erythrocyte sedimentation rate (ESR) and C-reactive protein level

Refer for urgent specialist assessment [1,2]:

- this may be to rheumatological, general medicine, or geriatric specialist, depending on local referral pathways [1]
- temporal artery biopsy should ideally be done in all patients within 1 week of starting corticosteroids [2]

While awaiting referral [1]:

- advise the person to seek immediate (same day) medical advice if they develop any visual disturbances
- assess response to prednisolone within 48 hours – if poor, seek specialist advice and consider an alternative diagnosis

NB: Lack of recognition and delay in treatment may lead to irreversible loss of vision – blindness in giant cell arteritis (GCA) almost always occurs prior to steroid therapy [1,2].

References:

[1] Clinical Knowledge Summaries (CKS). Giant cell arteritis. Version 1.1. Newcastle upon Tyne: CKS; 2009.

[2] Dasgupta B, Borg FA, Hassan N et al on behalf of the BSR and BHPR Standards, Guidelines and Audit Working Group. BSR and BHPR Guidelines for the management of giant cell arteritis. Oxford: British Society for Rheumatology; 2010.

[3] Map of Medicine (MoM). London: MoM; 2010.

Published: 29-Jul-2010 Valid until: 31-May-2012

Temporal arteritis/giant cell arteritis - suspected

Key Dates

Published: 29-Jul-2010, by International
Valid until: 31-May-2012

Accreditations

The care map is accredited by:

The Chief Knowledge Officer of the NHS:
Disclaimer

Evidence summary for Temporal arteritis/giant cell arteritis - suspected

This pathway has been developed according to the Map of Medicine editorial methodology (http://mapofmedicine.com/whatisthemap/editorialmethodology). The content of this pathway is based on high-quality guidelines [1,2,4], and critically appraised meta-analyses and systematic reviews [5,6]. Practice-based knowledge has been added by the Map of Medicine's Clinical Editorial Team and Fellows [3]. The evidence-based pathway has then been peer-reviewed by experts.

Search date: Feb-2010

References

This is a list of all the references that have passed critical appraisal for use in the care map Temporal arteritis/giant cell arteritis

ID	Reference
1	Clinical Knowledge Summaries (CKS). Giant cell arteritis. Version 1.1. Newcastle upon Tyne: CKS; 2009. http://www.cks.nhs.uk/giant_cell_arteritis
2	Dasgupta B, Borg FA, Hassan N et al on behalf of the BSR and BHPR Standards Guidelines and Audit Working Group. BSR and BHPR Guidelines for the management of giant cell arteritis. Oxford: British Society for Rheumatology; 2010. http://www.rheumatology.org.uk/includes/documents/cm_docs/2010/m/2_management_of_giant_cell_arteritis.pdf
3	Map of Medicine (MoM). London: MoM; 2010.
4	Scottish Intercollegiate Guidelines Network (SIGN). Diagnosis and management of headache in adults. Edinburgh: SIGN; 2008. http://www.sign.ac.uk/pdf/sign107.pdf
5	Karassa FB, Matsagas MI, Schmidt WA et al. Meta-analysis: test performance of ultrasonography for giant-cell arteritis. Ann Intern Med 2005; 142: 359-69. http://www.ncbi.nlm.nih.gov/pubmed/15738455
6	Mahr AD, Jover JA, Spiera RF et al. Adjunctive methotrexate for treatment of giant cell arteritis: an individual patient data meta-analysis. Arthritis Rheum 2007; 56: 2789-97. http://www.ncbi.nlm.nih.gov/pubmed/17665429

Disclaimers

The Chief Knowledge Officer of the NHS

It is not the function of the Chief Knowledge Officer of the NHS to substitute for the role of the clinician, but to support the clinician in enabling access to know-how and knowledge. Users of the Map of Medicine are therefore urged to use their own professional judgement to ensure that the patient receives the best possible care. Whilst reasonable efforts have been made to ensure the accuracy of the information on this online clinical knowledge resource, we cannot guarantee its correctness or completeness. The information on the Map of Medicine is subject to change and we cannot guarantee that it is up-to-date.