GIANT CELL ARTERITIS

Joint Ophthalmology and Rheumatology Referral and Treatment guidelines
University Hospitals Bristol NHS Foundation Trust (UHBristol)

EARLY RECOGNITION

Over 50 years old?
- Headache
- Jaw/tongue claudication (pain worsens on chewing), occasionally limb claudication
- Scalp tenderness
- Raised CRP or ESR>50
- Constitutional symptoms (fatigue, weight loss)
- Visual symptoms (loss of vision or diplopia, either transient or sustained)
- Temporal artery abnormalities (prominent artery/tender to palpation/absent pulse)

If over 50 and the above symptoms/signs are present even in the absence of headache…..

THINK GIANT CELL ARTERITIS (GCA)

General Principles:
GCA is almost never seen in the under-50 age group. Symptoms are often sudden onset. Over 90% of patients with GCA will describe constitutional symptoms, in particular fatigue. Malignancy must be excluded if marked weight loss is present or when concerning symptoms are detailed on systemic enquiry or examination. Although headache is present in two thirds of patients with GCA, it may be transient and is not always localised to the temporal regions. Jaw pain/claudication is the most specific symptom of GCA and may be associated with an increased risk of neuro-opthalmic complications.

Clinical Assessment:
1. Full clinical history including detailed systemic enquiry.
2. Cardiovascular examination to include peripheral pulses.
3. Temporal artery palpation
4. Cranial nerve examination (including ophthalmoscopy if available) but please do not delay referral of all patients with visual symptoms to ophthalmology for full assessment.
5. Full examination to exclude mimics (infection, malignancy or cervical spine pathology).

Baseline Investigations:
- CRP and ESR/PV, FBC, U&Es, LFTs.
- Urine Dipstick
- CXR (looking for mediastinal widening indicating aortic aneurysm)
## IMMEDIATE MANAGEMENT

All cases of suspected GCA should be discussed with the Ophthalmology or Rheumatology team, depending on the presence/absence of visual symptoms. See referral section for contacts.

**GCA with visual symptoms:**
Refer ALL cases for urgent ophthalmological assessment +/- admission to the Bristol Eye Hospital for IV methylprednisolone (IVMP). 24 hours service.

**GCA without visual symptoms:**
Discuss ALL cases with rheumatology and start:
- With jaw claudication: Prednisolone 60mg daily.
- Without jaw claudication: Prednisolone 40-60mg daily.
(Do not delay treatment out of office hours, but please still discuss to ensure appropriate follow-up)

Consider protein pump inhibitors for gastro-intestinal protection and bone protection.

### URGENT REFERRAL: (see also Figure 1)

**GCA with visual symptoms:**
Please contact the Bristol Eye Hospital casualty department during daytime hours (08:30 – 17:00 Monday to Saturday, 08:30 – 16:30 Sundays) on 0117 3424732. A triage nurse will arrange review. Outside these hours (or if no reply from eye casualty during daytime hours), contact the ophthalmology SpR on-call through the BRI Switchboard.

**GCA without visual symptoms:**
Please discuss all referrals with the rheumatology on-call, in order to ensure appropriate investigations are requested and follow-up arranged in a timely way, a written referral alone will not be accepted.

#### 9-5pm Monday to Friday:
- Contact the rheumatology team on bleep 7021 (via switchboard 01179 230000).

#### Out of hours:
- Start treatment and contact rheumatology team (bleep 7021) at 9-5pm on the next working weekday to arrange further follow up.

### TEMPORAL ARTERY BIOPSY (TAB)

- Urgent temporal artery biopsy will be arranged by rheumatology or ophthalmology team after discussion with the referring physician.
- TAB is the current gold standard but should not delay treatment
- False negatives can occur due to skip lesions or short biopsy sample (should be >1-2 cm).
- Aim to perform within 2 weeks of starting prednisolone to avoid treatment affecting result.
- If symptoms/response are typical, patients with negative biopsies should continue to be managed as GCA.
TEMPORAL ARTERY ULTRASOUND (TA U/S)

- Urgent temporal artery ultrasound will be arranged by the rheumatology or ophthalmology team after discussion with the referring physician.
- Temporal artery ultrasound is an emerging tool in the assessment of patients with suspected giant cell arteritis.
- Ultrasound is carried out by the vascular studies department, and will usually happen within 48 hours of referral.

Figure 1. Referral guidelines flowchart.
GCA: Giant Cell Arteritis; BEH: Bristol Eye Hospital; IVMP: Intravenous methylprednisolone; TAB: Temporal artery biopsy; TA U/S: Temporal artery ultrasound; BRI: Bristol Royal Infirmary. UKIVAS: UK and Ireland Vasculitis Registry.
**GENERAL MANAGEMENT PRINCIPLES**

The following guidance does not apply to the management of GCA with eye involvement. Patients remain under the care of Dr Catherine Guly, consultant ophthalmic physician at the Bristol Eye Hospital.

**Dose reduction for patients under Rheumatology care:**
This is a standard regimen, but will be tailored to the individual.
40-60mg od prednisolone continued for 2- 4 weeks (or until resolution of symptoms and lab abnormalities).
- then dose is reduced by 10mg every 2 weeks to 20mg od
- then by 2.5mg every 4 weeks to 10mg od
- then by 1mg every 1-2 months, provided there is no relapse

**Bone Protection:**
All patients should be commenced on supplemental calcium and vitamin D, and DEXA scanning arranged. High risk patients can be started on bisphosphonates (eg age>75, previous fracture).

**Monitoring of Treatment:**
Follow-up within Rheumatology:
Weeks 0, 2-3, and then months 3, 6, 12 in the first year, then tailored to individual patient.
- Rheumatology review - evidence of disease activity, adverse effects and complications.
- Chest radiograph to monitor for thoracic aortic aneurysm every 2 years
- Patients with visual involvement will be seen more frequently in ophthalmology.

In collaboration with general practice:
- Monthly repeat FBC, U&Es, Glucose, CRP and ESR/PV, - frequency reduced if stable.
- Screening and monitoring cardiovascular risk factors (increased due to GCA disease and treatment with prednisolone) - diabetes, BP and lipids.

**Relapses:**
Repeat inflammatory markers in all cases but do not delay referral if visual symptoms.

**Visual Symptoms:**
Discuss with eye-casualty or the ophthalmology SpR on-call (available 24/7, via BRI Switchboard, see above)

**Headache only:**
Treat with last effective prednisolone dose.

**Headache & Jaw Claudication:**
Treat with 40-60mg prednisolone daily and contact Rheum.

If patient is repeatedly relapsing on prednisolone dose reduction consider other causes, for example cervical spondylosis causing neck pain, urinary tract infections causing rise in inflammatory markers and general malaise.

**Steroid sparing agents:** Methotrexate, azathioprine and leflunomide may be considered in individual cases.

**Severe manifestations e.g. large vessel vasculitis or CVA.** Additional immunosuppressants +/- IV MP, Biologics or cyclophosphamide will be considered in individual cases.

**References**