CLINICAL PRACTICE GUIDELINE: Emergency Department

Giant Cell Arteritis

Disclaimer: This Clinical Practice Guideline ('CPG') was written for use in The Royal Victorian Eye and Ear Hospital Emergency Department. It should be used under the guidance of an Ophthalmology or ENT registrar. If clinical advice is required, please contact the Eye and Ear Admitting Officer for assistance: EYE: +61 3 9929 8033; ENT: +61 3 9929 8032. Links to internal Eye and Ear documents cannot be accessed from the website CPG.

Description:

Giant Cell Arteritis (GCA), also known as Temporal Arteritis, is a systemic vasculitis affecting medium and large arteries with significant ocular and systemic morbidity.

Red Flags:

- This is an ocular emergency with risk of bilateral blindness if not diagnosed and treated immediately
- Patient can present with transient visual loss, diplopia or any cranial nerve palsy
- Do not wait for temporal artery biopsy (TAB) before initiating steroid treatment if suspicion of GCA high
- Rarely GCA is possible with a normal ESR and CRP
- GCA is possible without systemic symptoms (incidence of "occult GCA" can be up to 20%)
- Not every patient with an elevated ESR and/or CRP has GCA
- NOTE: COVID precautions: all suspected GCA patients given high dose steroids should consider self-isolation for 2 weeks with the exception of hospital attendance for TAB or neuro-ophthalmology appointments

Background:

- More common in Caucasian women
- Average age: 71 years old. Most patients are over 60, very rare younger than 50 years old

How to Assess:

History:

- Systemic symptoms
  - Signs and symptoms of systemic inflammation are present in vast majority of patients and often precede ophthalmic symptoms
  - Headache, malaise, fatigue, weight loss, fever, sweats, scalp tenderness*, jaw claudication*, muscle aches (GCA is on a spectrum with polymyalgia rheumatica)
    (* most specific symptoms)
  - In presence of jaw claudication-like symptoms, exclude denture/tooth pain and temporomandibular joint pain
  - History of transient ischemic attack, stroke, myocardial infarction

- Ocular symptoms
    - Acute loss of vision (arteritic anterior ischemic optic neuropathy (AAION)), central retinal artery occlusion (CRAO)
    - Amaurosis fugax (precedes AAION in 10-30% of patients)
    - Diplopia
    - Ocular pain

Examination:

- Complete ophthalmic examination must be performed
    - Loss of vision: usually less than 6/60
        - AAION: cause of permanent visual loss in 90% of patients with GCA.
            - Signs of optic nerve dysfunction: afferent pupil defect, loss of color vision. Optic nerve: chalky white oedema.
        - CRAO
        - Less common: posterior ischemic optic neuropathy, cilioretinal artery occlusion, anterior segment and ocular ischemia
    - Ocular motility deficit: any pattern possible. Due to cranial nerve palsy, brain stem or orbital ischemia. Can mimic myasthenia gravis.
- Palpate superficial temporal artery (STA). In GCA, STA can be tender, firm, nodular, pulseless.

Investigations:

- Immediate ESR, CRP, FBE.
    - ESR can be normal in up to 22% of patients with GCA, and can be elevated by anaemia and other inflammatory/infectious diseases.
    - An elevated CRP is probably more specific.
    - Thrombocytosis is associated with GCA.
- Fluorescein angiography for delayed choroidal perfusion can be helpful in discriminating AAION from non-arteritic AION.

Acute Management:

- Discuss with neuro-ophthalmology consultant if there is uncertainty as to requirement for TAB and for advice regarding steroid treatment prior to biopsy.
- Initiate treatment immediately when suspicion for GCA is high as second eye involved within 24 hours in 1/3 of patients and within 7 days in 2/3 of patients. Treatment usually prevents involvement of second eye.
- Systemic steroid treatment - see red flags for COVID precautions
    - Intravenous (IV) steroids indicated in patients with recent visual loss, myocardial infarction, cerebrovascular ischemia
        - Admit
        - Dosage: 1000 milligrams (mg) daily for 3 days.

High dose methylprednisolone can be given once daily. In high-risk patients consider divided doses (twice or three times daily) or reduced dose. High-risk patients include patients with diabetes, heart failure, uncontrolled hypertension, cardiac disease, renal failure, suspected peptic ulcer disease, acute, chronic or latent infections, concurrent immunosuppression. Discuss with consultant and, in some cases, St Vincent's medical registrar. Medically unstable or high-risk patients, may require transfer to a medical unit (e.g. at St Vincent's) for appropriate supervision during high dose IV steroid therapy. See Methylprednisolone Inpatient Procedure

    - Oral prednisolone indicated if no sight threatening symptoms or if IV methylprednisolone a relative contraindication, e.g., frail/diabetic patient.
        - Dosage: 1 mg/kg up to 70 mg daily.

- Organise Temporal Artery Biopsy (TAB)
    - See Temporal Artery Biopsy Procedure for booking details
        - Contact Neuro-ophthalmology Registrar with patient details. Complete consent form and forward to Surgical Booking Unit immediately. Indicate 'right or left' TAB on consent. Preferred side for TAB may be indicated on consent form.
        - TAB usually performed in Day Surgery Treatment Room on afternoon list every Monday.
    - See TAB teaching package for pre, intra, and post-operative information and learning resources
        - TAB must occur within seven days of assessment to minimise false negative histopathology due to steroid treatment.
        - It is not necessary to routinely discontinue anticoagulants or anti-platelets for TAB. If these medications have been stopped by a medical practitioner, they can be resumed post-operatively on the day of surgery.
        - If unable to find pulse of STA (and no palpable 'cord'), please organise for patient to have STA mapped (ultrasound) by St Vincent's radiology department. Book radiology to be done on morning before biopsy scheduled.

Follow up:

- Neuro-ophthalmology clinic on Wednesday pm 2 days after TAB to review TAB results and management (unless patient was referred by a private specialist and will be followed up by the private specialist)

Additional notes:

- Patients given systemic steroids need to be advised of potential side effects

Give the patient a copy of Giant Cell Arteritis Patient Information and Temporal Artery Biopsy Patient Information sheets available on the Policy Centre or Eye and Ear website patient information hub.

Evidence Table

Author(s)	Title	Source	Level of Evidence (I – VII)
W Rahman, FZ Rahman	Giant cell (temporal) arteritis: an overview and update	Surv Ophthalmol. 2005 Sep-Oct; 50(5):415-28	I
George Spaeth	Ophthalmic Surgery, Principles and Practice, Chapter 52, Temporal Artery Biopsy		VII
Nesher G	The diagnosis and classification of giant cell arteritis	J Autoimmun. 2014 Jan 21, epub	VII
Colin CK Chan, Mark Paine, Justin O'Day	Steroid management in giant cell arteritis	Br J Ophthalmol 2001:85:1061-1064	VI
Helen V. Danesh-Meyer, Peter J. Savino	Giant cell arteritis	Curr Opin Ophthalmol, 2007:18:443-447	VII
Thomas RG Poole, Elizabeth M Graham, Sebastian B Lucas	Giant cell arteritis with a normal ESR and CRP	Eye 2003;17:92-93	VII
SS Hayrey, PA Podhajsky, B Zimmerman	Occult Giant Cell Arteritis: Ocular Manifestations	Am J Ophthalmol 1998;125:521-526	IV
M Haq, et al	Is it Necessary to Hold Anticoagulation Prior to Temporal Artery Biopsy	Poster Abstract, Number 2782. 2018 American Rheumatology Annual Meeting, October 2018	VI

The Hierarchy of Evidence

The Hierarchy of evidence is based on summaries from the National Health and Medical Research Council (2009), the Oxford Centre for Evidence-based Medicine Levels of Evidence (2011) and Melynk and Fineout-Overholt (2011).

I) Evidence obtained from a systematic review of all relevant randomised control trials.
II) Evidence obtained from at least one well designed randomised control trial.
III) Evidence obtained from well-designed controlled trials without randomisation.
IV) Evidence obtained from well-designed cohort studies, case control studies, interrupted time series with a control group, historically controlled studies, interrupted time series without a control group or with case series.
V) Evidence obtained from systematic reviews of descriptive and qualitative studies.
VI) Evidence obtained from single descriptive and qualitative studies.
VII) Expert opinion from clinician, authorities and/or reports of expert committees or based on physiology.

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