Giant cell [temporal] arteritis

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Giant cell arteritis (GCA) is a systemic vasculitis of unknown aetiology affecting medium and large arteries. New-onset headache in any patient over 50 years prompts consideration of GCA, as early recognition and treatment is critical to prevent blindness. Superficial temporal artery biopsy is most helpful for diagnosis (but false negatives occur).

Epidemiology

Primarily affects Caucasians, especially northern European. Gene for HLA-DRB1-04 has been identified as a risk factor. Female:male ratio is 4-6 : 1. Rare before the age of 50.

Pathophysiology

Cause unknown. Characterised by granulomatous inflammation, mostly along the internal elastic lamina of arterial walls. Inflammation may be followed by intimal proliferation and eventual stenosis or occlusion of the arterial segment. It is a T-cell dependent and antigen driven process, perhaps involving infectious agents (?HSV).

Clinical presentation

- Headache is most universal (85% cases), usually unilateral and over the temporal or occipital area, often worse at night, and gradually worsens over several days.
- Visual loss varies: sudden painless loss or transient initially (but blindness eventuates in 20-50% of those untreated). Bilateral visual loss within 2-3 weeks in >50% of untreated patients.
- Jaw/tongue claudication
- Diplopia
- Constitutional symptoms: anorexia and weight loss; fever and night sweats; malaise; and depression.

Diagnostic criteria (3 of the following)

- Age over 50 years
- New onset of headache or localised head pain
- Temporal artery tenderness or decreased temporal arterial pulse
- Westergren ESR >50 mm/h
- Arterial biopsy positive

Ophthalmic findings

Arteritic ischaemic optic neuropathy (AION) with eventual optic atrophy is the most common cause of visual loss (15% of patients). A relative afferent pupillary defect is present with monocular AION. The disc is oedematous and pale. Commonly, flame haemorrhages are located adjacent to the disc and the peripapillary retinal arterioles are often narrowed (Fig. 1)

Other ophthalmic manifestations include: branch retinal vein occlusion; central retinal artery occlusion; choroidal infarction; anterior segment ischaemic, cotton wool spots; and cranial nerve palsies (3rd most common).

Systemic complications include CVA and increased risk of thoracic aortic aneurysm/dissection.

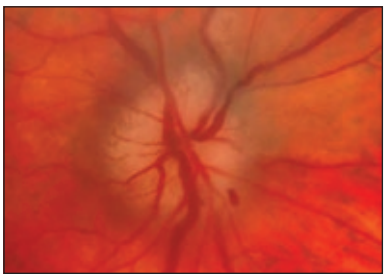

Fig 1. Pale disc; disc haemorrhage

Investigations

Superficial temporal artery biopsy remains the criterion standard for diagnosis, and should be performed promptly, within two weeks of commencing steroids.

Raised ESR (but up to 10% may have ESR <30 or normal) and CRP together have the greatest predictive value – sensitivity 100% and specificity 97%.

IV Fundus Fluorescein Angiogram (FFA) checks for choroidal hypoperfusion.

Management

Symptoms usually improve within days of steroid treatment. Arteritic ischaemic optic neuropathy is a medical emergency requiring IV methylprednisolone (1gm / day) for the first 3 days followed by high dose oral steroids (60-100 mg), tapered according to clinical response. The aim is to prevent visual loss, including the fellow eye. Some cases progress to visual loss despite therapy. ■