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GIANT CELL ARTERITIS WITH NORMAL INFLAMMATORY MARKERS

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Background:

Giant Cell is a granulomatous medium to large vessel vasculitis. It typically occurs in older people and is associated with polymyalgia rheumatica.[1] The condition usually manifests with involvement of the extracranial branches of the carotid artery. This results in the classical symptoms of a headache, temporal tenderness, jaw claudication and associated constitutional symptoms such as fever, lethargy, and malaise. The most serious manifestation is permanent visual loss which occurs due to optic nerve ischaemia.[2]

Case Presentation:

We present a case of an 86-year-old gentleman presented to hospital with sudden onset visual loss in his right eye. His past medical history included Chronic lymphocytic leukaemia, Benign Prostate Hypertrophy and Macular Degeneration. Initial blood tests revealed a CRP which was less than 5 with an ESR of 30 with a platelet count of 260.

He did not have any of the typical features GCA i.e. headache, scalp tenderness, jaw claudication or constitutional symptoms. There were no associated PMR symptoms. An ophthalmology review was sought. Ophthalmological assessment revealed right eye anterior ischaemic optic neuropathy. This was followed by a Temporal artery ultrasound scan showed widespread bilateral halo sign consistent with inflammation. This raised strong suspicion of GCA and the patient was subsequently treated with 3 days of IV Methylprednisolone followed by Prednisolone. This was given to protect the left eye from optic nerve ischemia.

Temporal artery biopsy performed at a later stage showed adventitial chronic inflammation of uncertain significance.

The combination of Ischemic neuropathy and Halo sign on ultrasound sufficed for a diagnosis of GCA to be made.[5]

Discussion:

The Inflammatory nature of GCA typically results in raised inflammatory markers. Therefore, the American College of Rheumatology include an erythrocyte sedimentation rate (ESR) of >50 mm/hr as one of its five classification criteria.[6] CRP is a more sensitive marker than ESR for a positive temporal artery biopsy, which is diagnostic of GCA. In clinical practise, both tests are performed to evaluate for GCA.[7]

This case is unusual as at the time of presentation, the inflammatory markers were normal. Furthermore, the patient did not exhibit the classical symptoms of headache, scalp tenderness, jaw claudication or PMR related symptoms. Rather, the patient presented directly with visual loss.

Epidemiology:

In the UK, the incidence of giant cell arteritis is approximately 2.2 per 10,000 person years.[3] A full time GP is likely to see a new case every 1–2 years.[4] GCA typically affects people in middle age and older. Its incidence rises with increasing age.[2]

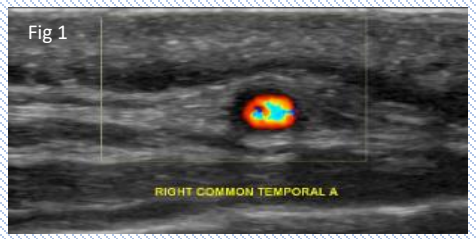

Fig 1:
GCA is characterized by oedematous infiltration of the intima, media and adventitia with lymphocytes and macrophages with or without giant cells. This results in artery wall thickening, which is most commonly concentric. On ultrasound, this phenomenon is termed 'Halo sign' as depicted in this image. [9]

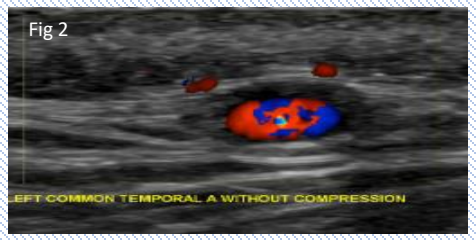

Fig2 :
The ultrasound scans of the right and left common temporal arteries demonstrates a 'halo sign' with intimal thickening noted of 0.5 mm. The vessels remain patent. The arteries demonstrates partial compression under probe pressure. The scan findings demonstrated here are compatible with GCA.

Learning Points:

1. Notable learning points drawn from this case are that normal Inflammatory markers should not exclude the diagnosis of GCA. If suspicion remains high, inflammatory markers should be repeated. Ultrasound of temporal arteries should be considered early followed by temporal artery biopsy.
2. It should also be noted that GCA may not present with the typical symptoms of headache, temporal tenderness or jaw claudication. Instead, it may present directly with visual symptoms.
3. Early treatment with steroids is essential to prevent progression of visual symptoms and to protect the contralateral eye.[8]

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