Introduction: Despite being the most common form of primary systemic vasculitis in adults, giant cell arteritis (GCA) remains a difficult diagnosis to make due to the breadth and versatility of presenting symptoms and signs. Many patients present with a myriad of symptoms which don’t fit neatly into the American College of Rheumatology criteria (1990) often with a negative or absent temporal artery biopsy. Here we describe an atypical and aggressive presentation of a patient with GCA.

Case description: A 77-year old male with a background of pertussis was initially referred to the medical take with 2 weeks of lethargy, raised C-reactive protein (343) and white cell count (27.8). He admitted to fevers, myalgia and sweats but denied any GCA symptoms. Examination revealed no abnormalities. He was treated for an infection of unknown source with broad spectrum antibiotics. A CT chest, abdomen and pelvis showed bibasal consolidation in the lungs with small effusions. He went on to have negative blood, sputum and urine cultures. Despite antibiotic escalations, his inflammatory markers remained high. An echocardiogram showed no vegetations. His pleural effusions were aspirated and revealed a transudative lymphocytic fluid with no acid-fast bacilli.

He was then referred to rheumatology. He denied GCA or connective tissue disease symptoms. He had a persistently raised CRP (>150) and WCC (>15) with no fevers. Examination revealed present temporal artery pulses and no abnormalities. Immunology was negative (ANA, ENA, ANCA and CCP). APET scan showed no changes consistent with a malignancy, large vessel vasculitis or infection.

Discussion:

Endovascular surgery is increasingly being preferred to open surgery, especially in patients presenting with potential vascular involvement. The role of MRI has been evaluated in identifying changes in arterial wall thickness and enhancement of only temporal and occipital arteries. The possibility of BD needs to be considered, especially in patients with vascular BD. Biochemically and symptomatically the patient has been well since infliximab was started, but we are currently awaiting a repeat CT scan. This case has also proved challenging as the patient has struggled with the diagnosis and need for frequent antibiotice scalations, his inflammatory markers remained high. An echocardiogram showed no vegetations. His pleural effusions were aspirated and revealed a transudative lymphocytic fluid with no acid-fast bacilli.

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He was labelled as an undifferentiated autoimmune disorder by the medical team and given a trial of prednisolone 30mg to which his CRP fell from 265 to 41. He was discharged with general medical follow-up and a prednisolone weaning regime.

After 1 month he re-presented to the medical take with acute bilateral visual loss (on 15mg prednisolone). This was preceded by a 2-week history of bitemporal headaches. There was no history of polymyalgia, jaw claudication or scalp tenderness. Ocular examination revealed bilateral swollen optic discs. He was admitted and treated with pulsed intravenous methylprednisolone.

An MRA of his head and neck showed intracranial irregularities consistent with a cerebral vasculitis. He was started on methotrexate alongside prednisolone and referred to the regional GCA centre for tocilizumab therapy.

Discussion: The frequency of ophthalmic complications in GCA is quoted to be as high as 30%. The case above presented with non-specific symptoms and extremely raised inflammatory markers. Despite numerous investigations including a PET scan, a definitive diagnosis was not made without the final presentation of permanent bilateral visual loss. The patient had appropriate and timely investigations for his symptoms at initial admission (blood tests, cultures, CT and PET imaging). Positive findings were appropriately investigated further. Ultimately, he was trialled with prednisolone when infection and malignancy were excluded by the medical team and discharged with follow up. Unfortunately, he was then readmitted with a dramatic relapse.

The British Society for Rheumatology recommends starting methotrexate following recurrent relapses in GCA, but we decided to start methotrexate due to the aggressive nature of his disease (sudden onset visual loss and cerebral vasculitis on MRA).

Few randomised controlled studies, case-control studies and meta-analyses have looked at the efficacy and steroid-sparing effects of methotrexate introduction at diagnosis of GCA to conflicting results. Jover et al. (2001) showed that low dose methotrexate (10mg) was effective in controlling GCA disease activity. This was supported by a meta-analysis (Mahr et al., 2007) and most recently in a prospective case-control study (Koster et al., 2019). The larger and well-designed Hoffman et al. (2002) trial showed a small benefit from methotrexate introduction but no statistical significance. There has been no large-trial convincing evidence of methotrexate efficacy in GCA at introduction or for relapses and this is reflected in the national guidance. However, the trials above used relatively low doses of methotrexate. Some of the trial also had short follow-up times and with slow steroid weans possibly masking a positive methotrexate effect.

Key learning points: Despite the vague and nondescript presentation of this patient, focusing on subtleties from the history and having a high index of suspicion are vital to making a diagnosis of GCA.

The patient had a negative PET scan despite progression to severe disease within 1 month. PET studies have shown high sensitivity and specificity values for the diagnosis of large vessel inflammation in GCA (90% and 98% respectively – Soussan et al., 2015). However, the lack of large vessel involvement on this scan was falsely reassuring.

Conflicts of interest: The authors have declared no conflicts of interest.