41. A COMMON RHEUMATOLOGY REFERRAL OF VAGUE CONSTITUTIONAL SYMPTOMS AND RAISED INFLAMMATORY MARKERS

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Introduction: Giant cell arteritis (GCA) is an inflammatory process affecting the medium/large vessels. Initially, the symptoms are vague constitutional symptoms caused by systemic inflammation. It is a chronic, granulomatous inflammation which most classically causes headache (in the temporal/occipital region), jaw claudication, scalp tenderness and visual loss/disturbance. There is now thought to be subgroups of GCA, one of which causes inflammation without temporal involvement (LV-GCA). If left untreated, this can have serious complications including ischemia (including cerebrovascular accident) and aortic aneurysm/dissection. Diagnosis of this subgroup can be challenging and in depth scanning is often required to support this.

Case description: A 75-year-old female presents with symptomatic anaemia (dizziness and shortness of breath) with haemoglobin of 72. This improves to 110 with 2 units blood transfusion but CRP is elevated at 250-290 during admission. Initially, this is treated as a UTI but CRP does not improve. She describes vague symptoms of fatigue for "weeks" and generally feeling unwell but no specific symptoms. She denies weight loss and there are no obvious infective sounding symptoms.

She has a past medical history of left-sided lung adenocarcinoma 2 years previously which was fully treated with lobectomy, recently diagnosed as Iron deficient anaemia, vague history of seronegative rheumatoid arthritis (on no treatment and no Rheumatology review), Diverticular disease and hypothyroid. Non-smoker and lives with husband and has full independence.

Her LFT's were deranged with mildly raised ALP/AST/ALT and there was also a description of nose bleed from the family 2 weeks prior to admission. She had a CT CAP which showed nil remarkable except trace fluid in pelvis. ESR is also raised >100 and albumin is low.

On further discussion when I reviewed her she described stiffness and pain in axial skeleton in keeping with PMR. There was no synovitis or tenderness in the joints, no rash, no sinusitis and no muscle tenderness. No features to suggest CTD or CREST. No headache or visual disturbance.

Plan was to assess ANCA, myeloma screen and arrange a PET scan for large vessel vasculitis given the CRP/ESR.

PET scan confirmed large vessel vasculitis - Marked FDG activity identified within the arterial walls of the large vessels throughout (carotids, subclavian arteries, aorta, iliacs and femoral arteries).

She was commenced on prednisolone with markedly good effect 2 weeks post treatment and is now on a reducing course of this with rheumatology follow up.

Discussion: GCA commonly affects people over the age of 50 and predominantly females. It causes inflammation in any of the proximal branches of the aorta and including the aorta itself. The diagnosis relies on the clinical history and inflammatory marker response. On occasions there will be overlap with polymyalgia rheumatica (PMR). A rational approach to diagnosis in GCA is required, based on a combination of clinical features together with relevant investigations.

Extra-cranial GCA (or LV-GCA) is a specific subgroup of GCA which will quite often not show the typical clinical features attributed to GCA. Importantly, it is now recognised that large vessel involvement is very common in all GCA patients and can lead to marked complications.

PET-CT scanning is extremely useful in diagnosing LV-GCA but delays occur in diagnosis secondary to waiting times and resources to carry out the scan and there is some information which suggests that these scans may lead to over-estimation of inflammation. Other imaging modalities include USS of some of the proximal branches of the aorta and CT/MRI angiography.

The concern of missing this diagnosis can be severe including ischaemic events, aneurysm of the aorta and possible CVA leading to significant morbidity and mortality. It is now wondered whether some of the patients who develop these complications could have underlying LVV.

Differential diagnoses must also be sought when making a diagnosis of LV-GCA including multiple myeloma, endocarditis/infections, other Rheumatological conditions and atherosclerosis. This can again lead to uncertainties when making a diagnosis.

It is important to commence steroid therapy at the earliest opportunity but awareness is required surrounding investigations and organising this as steroids can mask inflammation. Therefore it is important to correctly make the diagnosis with certainty and obtain a scan to prove LV-GCA as soon as possible.

Key learning points: One area of difficulty remains on the course of treatment for the extra-cranial GCA subgroup (or LV-GCA) and at present this is treated in the same way as cranial GCA. However, obviously with the advancements in tocilizumab as a steroid sparing therapy in cranial GCA this leads to a difficulty in understanding its role for the LV-GCA. Given the issue of not understanding biologics role in LV-GCA, this causes difficulty in utilising steroid sparing agents.

There is no diagnostic criteria for GCA but there is classification criteria which remains useful for the diagnosis of cranial GCA; however, it is less helpful for LV-GCA and therefore a large amount of the diagnosis depends on clinical judgement and imaging, as described above, to clarify the diagnosis. The importance of making a correct diagnosis is key given this will commit someone to a very long course of steroid therapy which obviously can lead to a great deal of side effects long term.

In addition, imaging modalities are now more plentiful for diagnosis and there is a large amount of information on this in the EULAR and BSR guidelines. Key areas of discussion still lie in which imaging techniques are best to use and the roles of using imaging in monitoring disease activity is still unknown.

Awareness is improving around this disease as there is obviously huge links to high morbidity and mortality complications including, as mentioned, stroke, ischaemic symptoms/claudication due to narrowing of the vessels from inflammation and aortic aneurysm/dissections too. Diagnosing all subgroups of GCA early is key to preventing these outcomes. In addition to this, it is important to obtain the correct diagnosis to prevent needless use of steroids and from missing potential differential diagnoses.

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