49. GIANT CELL ARTERITIS AND TAKAYASU ARTERITIS: TWO SIDES OF THE SAME COIN IN AN ELDERLY FEMALE?

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Introduction: Large vessel vasculitis (LVV) is a heterogeneous group of conditions, comprising of giant cell arteritis (GCA) and Takayasu arteritis (TAK). The American College of Rheumatologists (ACR) criteria provides a scoring system based on age, symptoms, and clinical examination findings, investigations to reach a diagnosis of either GCA or TAK. Similarly, European League Against Rheumatism (EULAR) recommendations discuss various imaging modalities in visualising vascular inflammation. Management is typically glucocorticoids, but for relapsing disease the challenge is finding suitable glucocorticoid-sparing agents. Here we describe a case of LVV presenting with great diagnostic complexity, discussing diagnostic modalities, treatment options and complications of therapy.

Case description: A 64-year-old female was referred to rheumatology clinic following extensive negative investigations, apart from on-going inflammatory response under gastroenterology for unexplained weight loss for over a year.

Further history revealed lower limb cramps, fatigue and frontal headaches. She did not report any jaw claudication or visual disturbances but had atypical neck pain. As the main carer for her mother with dementia all these symptoms led to significant functional impairment and a great deal of anxiety for the patient.

Under the gastroenterologists, she had had negative upper and lower GI endoscopies, CT thorax, abdomen and pelvis, labelled white cell scan and isotope bone scan in addition to a comprehensive septic screen including spinal imaging.

A diagnosis of systemic vasculitis was suspected. ANCA screen was negative. Patient declined a superficial temporal artery biopsy. A FDG PET scan confirmed increased activity in medium and large sized vessels, such as the common carotids, descending thoracic aorta, vertebral arteries, and femoral arteries. Sites of metabolic hypersignal were consistent with large vessel vasculitis. Previous CT angiography was reviewed once again. No evidence of TAK was identified. GCA-LV was decided as the most likely diagnosis.

The patient responded well to induction oral steroids with marked and rapid improvement in the inflammatory response. During the weaning down of steroids, the weight loss and the inflammatory response recurred.

Steroid sparing therapy was added on. Azathioprine was shortly discontinued owing to deranged liver function tests despite normal TPMT levels. Methotrexate was stopped as it caused suspected acute pneumonitis.

Oral bisphosphonates were not tolerated and she was listed for IV. Shortly after, she suffered a low trauma fracture neck of femur which caused fat emboli leading to a dense stroke. She recovered gradually with anticoagulation. Currently she is being screened for SC tocilizumab.

Discussion: Diagnosing LVV can be challenging owing to non-specific symptoms at presentation. Although our patient’s presentation of fatigue, weight loss and worsening inflammatory markers were reflective of a vasculitis-like pathology from the onset, confirming the exact diagnosis was far from straightforward. LVV has countless clinical manifestations with new onset headaches being the most predominant. Limb claudication, another presenting symptom, is described in literature as a clinical manifestation of LVV in 5-15% of cases. The patient’s complaint of neck pain is also a feature associated with external carotid artery disease.

Various imaging modalities, including MRI, CT angiogram, and FDG PET scans have been implicated in the diagnosis of LVV. Currently, no...
diagnostic threshold exists beyond which uptake is defining of vasculitis. One study (n = 40) evaluating diagnostic accuracy of PET scans in LVV cases against control patients, calculated 80% specificity and 65% sensitivity. Whereas, another study (n = 24) reported 100% specificity in use of PET scans for diagnosis of extracranial GCA.

A second dilemma encountered in this case following LVV diagnosis, was concluding the subtype. The patient’s diminishing pulses, a characteristic feature of TAK, was evaluated using angiography techniques. Results were inconsistent with the expected characteristic aortic stenosis or occlusions described by TAK diagnostic criteria. Despite no histological assessment from superficial temporal arteries, due to lack of scalp tenderness and jaw claudication, diagnosis was confirmed and managed as GCA-LVV.

This case was further complicated by finding a management regime both effective at tackling GCA-LVV activity, but also suitable for the patient. Disease which follows a relapsing course is indication for glucocorticoid-sparing agents. Published data is increasingly favouring use of tocilizumab, with a randomised control trial (n = 251) showing sustained remission in around 56% of patients. The aim with our patient remains finding a balance between adverse side effects and medication efficacy.

Key learning points: LVV is frequently the least obvious diagnosis, but nonetheless an important differential in a rheumatology clinic. The journey to diagnosing LVV is convoluted, either taking the route of temporal artery biopsy or various radiological imaging to gain an answer. This case highlights the broad extent of clinical manifestations of LVV, such as limb claudication.

In atypical presentations, with limited feasibility for histological studies, radiological imaging of arterial morphology has proven pivotal in reaching a diagnosis of LVV. A key finding for GCA on ultrasound is the ‘halo’ sign. The role of MRI has been evaluated in identifying changes in arterial wall thickness and enhancement of only temporal and occipital arteries. CT angiogram can demonstrate signs of active vessel disease more centrally. PET scans, although utilise ionising radiation, image the aorta and its branches in detail—therefore, guiding diagnosis.

This case would benefit the audience by demonstrating the diagnostic and treatment dilemmas in LVV. It will also highlight the disease burden of vasculitis especially due to the rare adverse effects this patient suffered due medication.

The conference would be a platform for the authors to learn from the experiences of various clinicians, around the diagnosis of LVV and to share with the department. We would very much like to have feedback from the experts in the field and also learn more about advances in biomarkers, like interleukins, now linked to LVV.

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