THU0589 'KU FEVER': A CASE REPORT

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Background: Anti-Ku antibodies have been associated with various connective tissue diseases, including myositis, arthritis, interstitial lung disease and glomerulonephritis¹.

Objectives: We present a case of a woman initially diagnosed with biopsy proven Kikuchi-Fujimoto disease who later developed a rapidly progressive myositis in association with anti-Ku antibodies.

Methods: A 47 year-old woman, originally from Myanmar, presented with lymphadenopathy, myalgia, fatigue, livedo reticularis and low-grade fever for the previous 6 months. This was initially diagnosed as a viral infection. Her myalgia progressed with proximal muscle weakness in both legs and associated rise in creatine kinase levels to 349U/L (normal range 25-200). She also developed dyspnoea, an erythematous rash, mouth ulcers and unintentional weight loss.

Blood tests show a lymphopenia and progressively rising CK, with a maximum level of 516 U/L. MRI whole body confirmed a widespread diffuse myositis in her upper and lower limbs, with an unusual 'speckled' pattern. High resolution CT Chest was normal. C3 0.40g/l (normal range 0.75-1.65) and C4 0.12 g/l (0.14-0.54) were low, with a positive ANA (1:160) and Ro-60 antibody. dsDNA, antiphospholipid screen and virology screens were all negative. Extended myositis panel revealed positive anti-Ku antibodies. Axillary lymph node biopsy confirmed necrotising lymphadenitis, consistent with Kikuchi-Fujimoto disease.

Results: She was initially treated with low dose Prednisolone and Hydroxychloroquine, with a limited response. Due to progressive myositis, pulsed IV Methylprednisolone 1g was provided over 3 days and mycophenolate mofetil (MMF) was started.

An inpatient stay was needed after developing an axillary node abscess and a chest infection. This was treated with intravenous antibiotics and repeated aspirations. Due to progressive myositis on a background of sepsis, intravenous immunoglobulin (IVIG) was administered over 5 days. Our patient made a good recovery, with normalization of CK levels and resolution of the myositis noted on repeat MRI scan. She remains on MMF as maintenance therapy.

Conclusion: Anti Ku antibodies appear to be associated with 2 spectrums of disease – elevated CK levels with interstitial lung disease and renal disease associated with anti-dsDNA antibodies¹. To our knowledge this is the first report in association with Kikuchi-Fujimoto disease. The combination of MMF and IVIG appears to have been effective treatment and her renal function remains stable, although we are monitoring the patient carefully for the possible development of interstitial lung disease.

References:

- [1] Spielmann L, Nespola B, Séverac F, et al. Anti-Ku syndrome with elevated CK and anti-Ku syndrome with anti-dsDNA are two distinct entities with different outcomes. *Annals of the Rheumatic Diseases* 2019;78:1101-1106.

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THU0590 WHEN RARE IS EVEN RARER: A COMPLEX CASE OF BEHCET DISEASE

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Background: Behcet disease is a rare inflammatory disorder with the unique ability to affect vessels of any size. The disease could be associated to thrombosis in both the venous and arterial compartment, and often aneurysms. In particular, the presence of aneurysms of the pulmonary artery is rarely, if ever, seen in conditions other than Behcet. Cardiac involvement, albeit uncommon, is also described and associated to a severe prognosis. The treatment is based on immunosuppressants, meanwhile the use of anticoagulants -especially when aneurysms are present- is debated.

Objectives: To describe a complex case of Behcet disease.

Methods: We report the case of a 45 years old man of Chinese origin who presented to A&E with fever and acute dyspnea. Blood test revealed raised ESR and CRP and raised neutrophil count. Chest X rays showed bilateral opacities suggesting pneumonia. The patient did not improve over the course of antibiotics. Later on, he presented with an episode of hemoptysis and worsening dyspnea, so he was admitted to the Intensive Care Unit. CT showed bilateral pulmonary thromboembolism and aneurysm of the pulmonary artery. Echocardiogram and cardio-MRI revealed a large, mobile thrombus within the right atrium. Extensive work-up for infections and cancer was unrevealing. ANA, ENA and ANCA antibodies were negative. On the basis of a past medical history of recurrent oral ulcers and papulopustular skin lesions that patient admitted on questioning, a diagnosis of Behcet disease was suspected. In keeping with that, HLA-B51 turned out positive. The patient was promptly started on IV steroid pulses followed by Cyclophosphamide 1 gr IV monthly for six months, then on IV anti-TNF alpha Infliximab. He was also commenced on low molecular weight heparin (LMWH) and subsequently direct factor Xa inhibitor Apixaban.

Results: The patient improved significantly with progressive regression of the pulmonary CT changes. He was discharged and able to get back to his daily life activities. After 2 years and a half of treatment, the aneurysm was stable and the intracardiac thrombus completely cleared.

Conclusion: This case is of particular interest because of the concomitant presence of two rare vascular complications of Behcet disease-intracardiac thrombosis (<1-2%, less than 100 cases described worldwide) and pulmonary artery aneurysm (1-2%). Prompt introduction of immunosuppressant therapy was associated with a favorable outcome with no recurrence. We could speculate that, to some extent, the concomitant use of anticoagulants may have contributed to the complete resolution of the intracardiac thrombosis.

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THU0591 CENTRAL NERVOUS SYSTEM VASCULITIS IN WHIPPLE DISEASE: A CASE REPORT

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Background: Whipple disease (WD) is a rare systemic infection with possible involvement of the central nervous system (CNS). The neurological manifestations of the disease are various and can mimic any neurologic condition.

Objectives: To describe the severe neurological complications occurred in a patient with WD misdiagnosed as a chronic inflammatory immune-mediated disorder.

Methods: Case report

Results: A 46-year-old woman developed acute right-sided hemiparesis and dysarthria. She had a 10-year history of ill-defined rheumatic condition (defined as seronegative rheumatoid arthritis, spondyloarthritis, and adult onset Still's disease) presenting with polyarthritis, episodic fever > 38°C and rash of unclear etiology poorly responsive to different immunotherapies including methotrexate, anti-IL6 and anti-IL1 inhibitors. Brain MRI demonstrated multiple anterior circulation infarctions and stenosis of the bilateral M1 segments of the middle cerebral artery on MR angiography. Black blood sequences revealed contrast enhancement of the vessel walls consistent with vasculitis (Figure 1). Cerebrospinal fluid (CSF) analysis was unrevealing, including PCR for viruses and bacteria. A working diagnosis of primary CNS vasculitis and progressive neurologic deterioration with abnormal behavior and altered mental status prompted the