THU0589

'KU FEVER': A CASE REPORT

N. Cernovschi-Feasey¹, D. Christidis¹, M. Lloyd¹, S. Melath¹, J. Wajed¹. ¹Frimley, Rheumatology, Frimley, United Kingdom

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.

Disclosure of Interests: None declared

DOI: 10.1136/annrheumdis-2020-eular.4833

THU0590

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

M. Di Cicco¹, O. M. Epis¹, C. Casu¹, A. Adinolfi¹, L. Alvaro¹, V. Campanella¹, M. Chevallard¹, M. Muscarà¹, M. Romano¹, E. Schito¹, N. Ughi¹, E. Verduci¹, D. A. Filippini¹. ¹Niguarda Hospital, Rheumatology, Milan, Italy

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.

Disclosure of Interests: MARIA DI CICCIO: None declared, oscar massimiliano epis Consultant of: yes, Speakers bureau: yes, Cinzia Casu: None declared, Antonella Adinolfi: None declared, Luisa Alvaro: None declared, Valeria Campanella: None declared, Michel Chevallard: None declared, Marina Muscarà: None declared, Mariaeva Romano: None declared, Emanuela Schito: None declared, Nicola Ughi: None declared, Elisa Verduci: None declared, Davide Antonio Filippini: None declared

DOI: 10.1136/annrheumdis-2020-eular.5663

THU0591

CENTRAL NERVOUS SYSTEM VASCULITIS IN WHIPPLE DISEASE: A CASE REPORT

A. Giollo¹, C. Zivelonghi², D. Cardellini², G. M. Schiavi², G. Vattemi², O. Viapiana¹, M. Rossini¹. ¹University of Verona, Rheumatology Section, Department of Medicine, Verona, Italy; ²University of Verona, Department of Neuroscience, Biomedicine and Movement Sciences, Verona, Italy

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

initiation of intravenous (IV) methylprednisolone followed by cyclophosphamide without significant improvement. Re-evaluation of the long-standing history of joint symptoms unresponsive to immunotherapy, along with recurrent fevers and chronic diarrhea raised the suspicion of unrecognized Whipple's disease. PCR for *Tropheryma Whipplei* was positive in stool, urine, blood and CSF, and duodenal mucosal biopsies confirmed the diagnosis. A combination of ceftriaxone, doxycycline, and hydroxychloroquine was initiated. Three days later the patient developed periorcular burning pain and cutaneous vesicles consistent with shingles. Varicella-zoster virus DNA was detected in CSF and IV acyclovir was started. At 3 months follow-up neurologic examination was unremarkable except for a slightly fatuous behavior.

Conclusion: Recognition of rare manifestations of WD is important to avoid diagnostic delay and inappropriate, potentially harmful treatments.

Figure. Brain MRI with FLAIR sequences revealing hyper intense lesions in the subcortical white matter of both hemispheres (A, B). C, DWI sequences showing several recent ischemic lesions bilaterally with prominent involvement of the left frontal lobe. D, black blood sequences documenting enhancement and thickening of the wall vessel of both MCAs (red circles).

Disclosure of Interests: Alessandro Giollo: None declared, Cecilia Zivelonghi: None declared, Davide Cardellini: None declared, Gian Marco Schiavi: None declared, Gaetano Vattermi: None declared, Ombretta Viapiana: None declared, Maurizio Rossini Speakers bureau: AbbVie, Abiogen, Amgen, BMS, Eli-Lilly, Novartis, Pfizer, Sanofi, Sandoz and UCB
DOI: 10.1136/annrheumdis-2020-eular.4985

THU0592 INSIDIOUS CORONARY ARTERY DISEASE IN A YOUNG PATIENT WITH POLYARTERITIS NODOSA: A CASE REPORT AND REVIEW OF LITERATURE

H. Huang¹, Z. Zhang¹. ¹Peking University First Hospital, Beijing, China

Background: Polyarteritis nodosa (PAN) is a systemic necrotizing vasculitis that typically affects medium-sized muscular arteries, with occasional involvement of small muscular arteries[1]. Although overt myocardial infarction is uncommon, myocardial ischemia may result from narrowing or occlusion of the coronary arteries[2].

Objectives: Herein, we report a case with 7-year's history of PAN and unstable angina pectoris due to coronary occlusions of the three main arteries. We also reviewed the literatures regarding coronary artery involvement in PAN.

Methods: A 22-year-old Chinese man who presented with chest pain lasting for a few minutes and then subsiding spontaneously for 1 month was admitted to our hospital. He was diagnosed as PAN 7 years ago and during 7-years' follow-up, he has been in stable condition, without any discomfort or abnormal laboratory findings. In December 2019, he suffered from chest distress accompanied by retrosternal pain, with frequency of about 2-3 times a week. His symptoms were gradually aggravating with dyspnea at night.

Results: Coronary computed tomography angiography showed diffuse coronary stenosis (Fig. 1). Further coronary angiography revealed a slight plaque infiltration of the left main coronary artery, and occlusion of all the three major coronary arteries, as well as multiple coronary aneurysms. 95% stenosis of the obtuse margin branch artery was also found and a stent was then implanted (Fig. 2). Prednisone 50mg/day and methotrexate 15mg/week were reinitiated, in combination with anti-anginal medications including aspirin and statin.

Fig. 1 Coronary computed tomography angiography found diffuse coronary stenosis.

Fig. 2 Coronary angiography. (a) A 50% stenosis followed by aneurysmal change of the proximal end of left anterior descending (LAD) artery, and totally occluded from the middle segment; A aneurysmal change of the initial part of left circumflex artery (LCX) and then totally occluded (dotted line); A 95% stenosis obtuse margin branch. (b) A totally occluded right coronary artery (dotted line). (c) Final appearance of the LCX after stent implantation.

After we reviewed all the English literatures reporting cardiac involvements in adults with PAN from 1990 to 2019, a total of 34 patients from 32 articles were identified. 25 (73.5%) patients were admitted to hospital due to acute coronary syndromes manifesting as chest pain or dyspnea. Coronary stenosis or occlusions were most common on imaging or autopsy. Most of the patients had more than one vessel involved, of whom 7 patients showed evidence of triple vessel lesions. Aneurysm was also common in these patients, especially multiple aneurysms. Spontaneous coronary artery dissections were rare in PAN patients. Most patients received glucocorticoid, and/or immunosuppressant therapy, including cyclophosphamide and azathioprine, with or without invasive operations. 15 patients died from cardiopulmonary arrest, the most frequent cause being death, and 15 patients were stable without symptoms after treatment.

Conclusion: We report a young PAN patient with insidious stenosis of three main coronary arteries under the circumstance of stable disease activity for years. This reminds us of the necessity of assessing heart, probably other organs as well, in PAN patients even though their acute phase reactants in serum are normal. But how often to do the screening and which screening examination should be done, remain to be further investigated.