Conclusions: PNI at diagnosis can be used to estimate BVAS at diagnosis and PNI at diagnosis ≤ 36.6 may predict relapse during the follow-up in AAV patients.

REFERENCES:

- [1] Nihon Geka Gakkai Zasshi 1984;85:1001–5.
- [2] J Cancer Res Clin Oncol 2014;140:1537–49.
- [3] Arthritis Rheum 2013;65:1–11.

Acknowledgements: None.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.4991

SAT0548 PROGNOSTIC FACTORS FOR INTERSTITIAL LUNG DISEASE WITH MICROSCOPIC POLYANGIITIS

T. Shoda^{1,2}, T. Takeuchi², K. Isoda^{1,2}, J. Komma^{1,2}, T. Kotani², S. Makino².

¹Clinical Immunology and Rheumatology, Yodogawa Christian Hospital;

²Department of Internal Medicine (IV), Osaka Medical College, Osaka, Japan

Background: Many cases of interstitial lung disease (ILD) complicated by microscopic polyangiitis (MPA) show the UIP pattern on chest HRCT, being similar to idiopathic pulmonary fibrosis (IPF/UIP), and the associated prognosis is poor compared with that of MPA without ILD.¹ However, the details have not been fully clarified.

Objectives: Prognostic factors of MPA-ILD sufficiently treated with immunosuppressive therapy were investigated.

Methods: Of consecutive patients with MPA who received inpatient treatment at our hospitals between 2001 and 2016, MPO-ANCA-positive patients who met the 2007 EMEA classification criteria and had concomitant ILD on HRCT were selected as the subjects. Using the clinical data and HRCT fibrosis score,² the outcome and prognostic factors were retrospectively investigated.

Results: The subjects were 65 patients with MPA-ILD, 31 and 34 patients were male and female, respectively, and the median age (interquartile range) was 73^{67–76} years old. At the time of treatment initiation, MPO-ANCA was 129 (50.9–359) EU; KL-6, 461 (289–665) U/mL; Aa-DO2, 25.1 (15.6–34.2); %FVC, 81.2 (67.8–93.9)%; and %DLco/VA, 62.7 (45.3–73.2)%. On HRCT, the UIP and non-UIP patterns were observed in 44 and 21 patients, respectively. In treatment, prednisolone was administered to 63 patients, immunosuppressants were used in 55 patients, and blood purification therapy was concomitantly administered to 9 patients. MPO-ANCA on the final follow-up was lower than the detection sensitivity in 56 patients. The outcome was death in 23 patients, and the 5- and 10 year survival rates after treatment initiation were 69.8% and 51.1%, respectively (acute exacerbation of interstitial pneumonia: 5 patients, infection and alveolar haemorrhage: 3, infection: 1, pneumothorax: 1, lung cancer: 2, pulmonary hypertension: 1, sudden death: 4, heart failure: 2, renal failure: 2, cerebral haemorrhage: 1, intestinal haemorrhage: 1). Regarding lung disease-related death, the age ($p=0.018$), %FVC ($p=0.026$), HRCT fibrosis score ($p<0.001$), CPFE ($p=0.004$), decreased lung volume ($p=0.041$), and honeycombing ($p=0.051$) were extracted on univariate analysis, and the HRCT fibrosis score was significant on multivariate analysis ($p=0.001$). The prognosis of patients with a fibrosis score of 19% or higher was significantly poor.

Conclusions: Many MPA-ILD patients showed the UIP pattern, but their prognosis was better than that of previously reported IPF/UIP patients, suggesting that early immunosuppressive therapy is effective. However, expansion of fibrosis was included in the factors indicating a poor prognosis, suggesting the limit of immunosuppressive therapy.

REFERENCES:

- [1] Fernandez Casares M, Gonzalez A, et al. Microscopic polyangiitis associated with pulmonary fibrosis. Clin Rheumatol. 2015;34(7):1273–7.
- [2] Wells AU, Hansell DM, Rubens MB, et al. Fibrosing alveolitis in systemic sclerosis: indices of lung function in relation to extent of disease on computed tomography. Arthritis Rheum 1997;40:1229–36.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.5298

SAT0549

AUTOLOGOUS HEMATOPOIETIC STEM CELL TRANSPLANTATION (HSCT) FOR ANTI-NEUTROPHIL CYTOPLASMIC ANTIBODY (ANCA)-ASSOCIATED VASCULITIS (AAV) – A EBMT RETROSPECTIVE SURVEY

T. Alexander¹, C. Samuelson², T. Daikeler³, J. Henes⁴, L. Skagerlind⁵, M. Akil², G. Ehninger⁶, E. Jantunen⁷, D. Farge⁸, J. Snowden⁹ on behalf of EBMT Autoimmune Diseases Working Party. ¹Rheumatology, University Medicine Berlin, Berlin, Germany; ²Childrens Department, Sheffield Teaching Hospitals NHS Foundation Trust, Sheffield, UK; ³Rheumatology, University Hospital, Basel, Switzerland; ⁴Rheumatology, Medizinische Universitätsklinik Abt. II, Tübingen, Germany; ⁵Hematological Department, University Hospital of Umeå, Umeå, Sweden; ⁶Hematology Department, Universitätsklinikums Dresden, Dresden, Germany; ⁷Department of Medicine, University of Eastern Finland, Kuopio, Finland; ⁸Internal Medicine, St. Louis Hospital, Paris, France; ⁹Hematology Department, Sheffield Teaching Hospitals NHS Foundation Trust, Sheffield, UK

Background: ANCA-associated vasculitides (AAV) are chronic autoimmune diseases, which can present with life-threatening multi-system involvement. Despite the use of rituximab and other available modern biologic therapies some patients with AAV develop severe and refractory courses of disease.

Objectives: The aim of this study was to analyse outcomes of autologous hematopoietic stem cell transplantation (HSCT) for refractory AAV

Methods: Adults receiving HSCT for AAV and whose data were registered within the EBMT Autoimmune Disease Working Party (ADWP) were identified retrospectively through the EBMT database. Treating physicians were surveyed to produce a retrospective evaluation of outcomes.

Results: 7 patients underwent HSCT primarily for AAV between 1999–2014 in 6 centres across Europe. 5 females and 2 males were transplanted; 6 had a diagnosis of granulomatosis with polyangiitis (GPA) and 1 eosinophilic granulomatosis with polyangiitis (EGPA). Median age was 39 years (range 32–55 years). Patients had received 4–6 prior lines of therapy, including cyclophosphamide (CYC, with median cumulative dose of 80 g) and steroids in every case, and rituximab in 4 cases. Stem cell source was peripheral blood in every case; CD34-selection was performed in 4 cases, mean CD34⁺ cell dose was $4.2 \times 10^6/\text{kg}$ (range $0.6\text{--}7.9 \times 10^6/\text{kg}$). Conditioning regimen was CYC/ATG in 5 patients and CYC in 2 patients. Median follow-up was 86 months (range 1–204 months). Transplant-related mortality (TRM) occurred in 2 causes. All but one patient went into remission but 3 later relapsed at 6, 12 and 36 months, respectively, and required further treatment for disease control. At time of last clinical follow-up, 3 patients had drug-dependent partial response; 1 had drug-dependent complete remission and 1 had drug-free complete remission.

Conclusions: Outcomes of HSCT for these heavily pre-treated AAV patients were variable. Only 1 patient achieved drug-free complete remission and TRM was observed in a quarter. Nevertheless, HSCT had the potential to stabilise AAV in patients who initially failed to respond to conventional therapies. These data do not support HSCT for advanced stage ANCA-positive vasculitis, although it may have a place as salvage treatment in otherwise refractory patients. As for other autoimmune diseases, HSCT may provide better outcomes when performed at early stage of disease. Overall, HSCT should only be performed in clinical trial settings in experienced centres.

Disclosure of Interest: None declared

DOI: 10.1136/annrheumdis-2018-eular.7404

SAT0550

HEALTH-RELATED QUALITY OF LIFE IN PATIENTS WITH GIANT CELL ARTERITIS TREATED WITH TOCILIZUMAB IN A PHASE 3 RANDOMISED CONTROLLED TRIAL

V. Strand¹, S. Dimonaco², K. Tuckwell³, M. Kleerman³, N. Collinson², J. H. Stone⁴ on behalf of GiACTA Investigators. ¹Division of Immunology/Rheumatology, Stanford University, Palo Alto, USA; ²Roche Products Ltd, Welwyn Garden City, UK; ³Genentech, South San Francisco; ⁴Massachusetts General Hospital Rheumatology Unit, Harvard Medical School, Boston, USA

Background: Superior rates of sustained glucocorticoid (GC)-free remission were shown in patients with giant cell arteritis (GCA) treated with weekly or every-other-week (wk) subcutaneous tocilizumab (TCZ) 162 mg +26 wk GC taper for 52 wks compared with placebo +26 wk or 52-wk GC taper (PBO +26 or PBO +52) in the GiACTA trial. Statistically significant improvements in SF-36 Physical Component Summary (PCS) scores were reported for weekly TCZ vs PBO +52 and in patient-reported global assessment of disease activity for both TCZ groups vs both PBO groups.¹

Objectives: To report further analysis of patient-reported outcomes (PROs) in GiACTA.

Methods: Analyses of SF-36 PCS and Mental Component Summary (MCS), SF-36 domains, and Functional Assessment of Chronic Illness Therapy (FACIT)-fatigue compared patients treated with weekly TCZ ($n=100$) vs PBO +26 ($n=50$;

not shown) or PBO +52 (n=51) for 52 wks based on reported data, including all responders as well as patients with post-escape data following flare.
Results: Improvements in SF-36 PCS and MCS scores, 6 of 8 SF-36 domains, and FACIT-Fatigue at wk 52 were significantly greater with weekly TCZ vs PBO +52 ($p<0.01$) (table 1, figure 1). At wk 52, mean scores met or exceeded age/gender (A/G)-matched normative scores in the weekly TCZ group; higher proportions of patients reported scores exceeding A/G norms in SF-36 PCS and MCS, all SF-36 domains, and FACIT-Fatigue (Table) compared with PBO groups. The median cumulative prednisone dose over 52 wks was lower with weekly TCZ (1862.0 mg) vs PBO +26 (3296.0 mg) or PBO +52 (3817.5 mg) ($p<0.01$).

Table. Change From Baseline to Wk 52; mean score (% $\geq$ A/G norms)

	Weekly TCZ+26 n=100			PBO+52 n=51		
	Baseline	Wk 52	LSM Δ	Baseline	Wk 52	LSM Δ
PROs (A/G norms)						
PtGA	43.61	24.36	-17.14	47.78	35.44	-7.56
FACIT-Fatigue (40.0)	36.05 (43.4)	42.08 (73.8)	5.30 ^a	31.42 (32.7)	32.62 (35.6)	-0.42
SF-36 PCS (50.0)	43.10 (23.7)	47.75 (43.5)	4.18 ^a	41.12 (20.4)	41.24 (22.2)	-0.40
SF-36 MCS (50.0)	42.77 (33.0)	51.64 (60.0)	8.10 ^a	40.45 (34.7)	44.86 (40.0)	1.89
SF-36 Domains (A/G norms)						
Physical function (67.56)	69.10 (60.0)	78.28 (78.8)	6.83	59.40 (42.0)	65.44 (55.6)	2.68
Role physical (69.44)	49.56 (25.0)	73.75 (56.5)	20.64 ^a	45.38 (28.0)	53.89 (33.3)	4.46
Bodily pain (64.52)	61.93 (41.0)	73.25 (65.9)	10.89 ^a	55.67 (34.7)	56.27 (31.1)	-2.87
General health (66.49)	55.00 (25.8)	65.81 (57.6)	9.06 ^a	55.69 (36.0)	52.29 (22.0)	-4.05
Vitality (58.65)	50.19 (33.3)	66.13 (68.2)	15.69 ^a	42.38 (28.0)	49.17 (33.3)	3.53
Social function (81.49)	64.25 (29.0)	84.71 (63.5)	17.35 ^a	63.00 (40.0)	67.50 (40.0)	2.34
Role emotional (82.08)	66.38 (43.0)	82.45 (62.4)	13.37	60.33 (36.0)	69.63 (46.7)	3.53
Mental health (77.16)	64.04 (32.3)	77.94 (52.9)	12.54 ^a	59.10 (24.0)	66.33 (31.1)	3.13

LSM Δ , least squares mean change from baseline to wk 52; PtGA, patient-reported global assessment.

All analyses based on observed data (post-escape data included).

^a $p<0.01$ vs PBO+52.

^a $p<0.01$ vs PBO+52.
A/G, age/gender; BL, baseline.

Abstract SAT0550 – Figure 1. SF-36 Domains at BL and Week 52. ^a $p<0.01$ vs PBO+52. A/G, age/gender; BL, baseline.

Conclusions: Patients with GCA treated with weekly TCZ 162 mg and a 26-wk GC taper reported statistically significantly greater improvements in health-related quality of life and fatigue that exceeded normative values compared with those receiving 52-wk GC taper alone, in part ascribed to lower steroid doses.

REFERENCE:

[1] Stone JH, et al. N Engl J Med 2017;377:317–328.

Disclosure of Interest: V. Strand Consultant for: AbbVie, Amgen, AstraZeneca, BMS, Boehringer Ingelheim, Celgene, Celltrion, CORRONA, Crescendo, EMD Serono, Genentech/Roche, GSK, Janssen, Lilly, Merck, Novartis, Pfizer, Protogen, Regeneron, Samsung, Sandoz, Sanofi, UCB, S. Dimonaco Employee of: Roche, K. Tuckwell Shareholder of: Roche, Employee of: Roche, M. Kleerman Employee of: Genentech, N. Collinson Employee of: Roche, J. H. Stone Grant/research support from: Roche, Genentech, Xencor, Consultant for: Roche, Genentech, Xencor

DOI: 10.1136/annrheumdis-2018-eular.2616

SAT0551

LEG ULCERS IN BEHCET'S SYNDROME: AN OBSERVATIONAL SURVEY IN 24 PATIENTS

Y. Ozguler¹, Z. Kutlubay², A.S. Dikici³, M. Melikoglu¹, C. Mat², H. Yazici¹, E. Seyahi¹. ¹Department of Internal Medicine, Division of Rheumatology; ²Department of Dermatology; ³Department of Radiology, Istanbul University Cerrahpasa Medical Faculty, Istanbul, Turkey

Background: Formal experience with leg ulcers in Behçet's syndrome (BS) is limited. It is a rare complication that can be seen during the course of mainly pts with the post-thrombotic syndrome. They can be difficult to manage and may cause disability leading to unemployment and severe impairment in quality of life.

Objectives: In this observational survey, we aimed to describe clinical characteristics of pts with leg ulcers.

Methods: A total of 24 pts (23 M/1 F) that were seen at our multidisciplinary BS clinic between May 2016-Jan 2018 were evaluated with the help of a standardised questionnaire. Venous Doppler US and if necessary abdominal CT were used to evaluate localization of venous involvement. Biopsies were done if needed. Medical and other interventional treatments were recorded.

Results: The mean age at disease onset was 27.5 $\pm$ 7.1 years. The median duration between the disease onset and ulcer development was 4.0 [2.5–11.5] years. The median follow-up was 7.8 [IQR: 2.9–14.2] years. Eleven (46%) were unemployed due to the leg ulcer. Venous involvement was present in 20 pts (83%). Lower extremity vein thrombosis was shown in all these 20 pts. It was mostly bilateral (15/20). Femoral veins were the most commonly involved (18/20), followed by popliteal veins (14/20), v. saphena magna or v. saphena parva (7/20), iliac veins or inferior vena cava (8/20). The same 9/20 pts had other large vessel involvement such as pulmonary artery (n=5) or dural sinus thrombosis (n=3). Four pts did not have any venous thrombosis or insufficiency. Histopathologic studies could be done in 3 and showed necrotizing vasculitis in 2 and venous stasis in the 3rd. Twelve pts (50%) had solitary ulcers and 12 had 2 or more. We observed a total of 34 ulcers in 24 pts. They were mostly found around the medial malleolus (15/34) and anterior surface of the tibia (14/34). Five pts had leg ulcers at unusual places such as lateral malleolus (n=2), popliteal fossa (n=1) and posterior surface of the tibia (n=2). Immunosuppressive agents including azathioprine, cyclophosphamide, interferon alpha, infliximab, and corticosteroids were used. Bed rest was advised to almost all. Iloprost infusions were given in 13 (54%) for a median duration of 6 mo. Additionally, larvae of *Lucilia sericata* were tried in 9 pts. Skin graft insertion was used in 2 pts, however, was successful in 1. We observed that in 11 (46%) pts 17 ulcers healed in median 24 mo [IQR:9–78]. In the remaining 13 (54%) pts 17 ulcers stayed as unhealed for a median 7 years [IQR:5–11], despite all interventions and medical treatment. We saw the mean age at BS onset was significantly younger (24.4 $\pm$ 5.4 vs 30.4 $\pm$ 7.3 years; $p<0.05$) and the median time between disease onset and ulcer development was significantly shorter (3 [IQR:1–4.5] vs 10 [IQR:4–14] years, $p<0.05$) in pts with healed ulcers compared to those with unhealed ulcers. There were no apparent associations with the type of management and the severity of venous involvement.