SAT0546 SENSORINEURAL HEARING LOSS IN TAKAYASU'S ARTERITIS

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Background: Sensorineural hearing loss has been reported to be increased in several chronic autoimmune and non-autoimmune diseases such as systemic lupus erythematosus (SLE), progressive systemic sclerosis, rheumatoid arthritis, small vessel vasculitides, ankylosing spondylitis and Behçet's syndrome. We had sporadically noted several degrees of sensorineural hearing loss among our Takayasu's arteritis (TA) patients. While some revealed this in their past history, others had relapsing attacks of hearing loss independent of or associated with vascular relapses.

Objectives: We formally investigated the frequency and type of hearing loss among TA patients and suitable controls.

Methods: The study was done in two parts. In the first part, consecutive TA and SLE patients seen at outpatient clinic along with apparently healthy controls were administered a standardised questionnaire that assessed hearing loss, tinnitus, and episodic vertigo. In the second part, previously registered TA and SLE patients for another study were called to specifically for otological examination and audiometry tests that included pure-tone air and bone conduction, speech audiometry and acoustic reflex threshold test.

Results: In the first part, 73 patients with TA, 107 patients with SLE and 133 healthy controls were studied as shown in table 1. The frequency of those with hearing deficit/loss, tinnitus and vertigo were significantly more common among both TA and SLE patients (table 1). While the frequency of those with hearing deficit/loss was similar in TA and SLE, those with tinnitus and vertigo were significantly more common in TA.

In the second part, 50 patients with TA, 22 patients with SLE and 32 healthy controls were studied as shown in table 2. Audiometry tests revealed that several degrees of hearing loss were present in 36% of the patients with TA, 18% of the patients with SLE and 6.3% of the healthy controls ($p<0.05$). This was mostly due to sensorineural hearing loss in TA patients (13/18) and high-frequency type was the most common pattern. Moreover, those TA patients with sensorineural hearing loss did not show any specific vascular pattern.

Abstract SAT0546 – Table 1. Results of the questionnaire survey

	Takayasu's arteritis (n=73)	SLE (n=107)	Healthy controls (n=133)	p value
Mean age	43.7±11.0	44.0±10.3	42.5±8.3	
Disease duration, med [IQR]	7 ³⁻¹²	8 ³⁻¹¹	NA	
Hearing deficit/loss, n (%)	20 (27.4)	21 (19.6)	4 (3.0)	0.001
Tinnitus, n (%)	37 (50.7)	34 (31.8)	16 (12.0)	0.001
Vertigo, n (%)	39 (53.4)	38 (35.5)	19 (14.3)	0.001
	Takayasu's arteritis (n=50)	SLE (n=22)	Healthy controls (n=32)	p value
Mean (SD) age	41.3±8.9	42.1 ±10.3	41.1±0.8	NS
Disease duration, med [IQR]	6 ⁴⁻¹¹	7 ³⁻¹¹	-	NS
Hearing loss, n (%)	18 (36.0)	4 (18.1)	2 (6.3)	<0.05
Sensorineural type, n (%)	13 (26.0)	2 (9.0)	1 (3.1)	
Conductive type, n (%)	5 (10.0)	2 (9.0)	1 (3.1)	

Conclusions: We are unaware of previous surveys of sensorineural hearing loss in TA. Our study shows that audiovestibular system is considerably affected in TA, similar to that observed in SLE. The fact that there was no clear vascular pattern among patients with hearing loss, suggest that small vessel vasculitis was probably the cause of this hearing loss.

Disclosure of Interest: None declared

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SAT0547 PROGNOSTIC NUTRITIONAL INDEX FOR ESTIMATING BIRMINGHAM VASCULITIS ACTIVITY SCORE IN ANCA-ASSOCIATED VASCULITIS

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Background: The prognostic nutritional index (PNI), which was first introduced by Onodera et al., is calculated based on the serum albumin level and total lymphocyte count in the peripheral blood. It is proposed to be a parameter to reflect immunonutritional status, however, it is also known that albumin and lymphocyte count may decrease in proportion to inflammatory burdens in autoimmune diseases.

Objectives: We investigated whether PNI at diagnosis can be used for estimating BVAS at diagnosis in antineutrophil cytoplasmic antibody (ANCA) associated vasculitis (AAV) patients.

Methods: We retrospectively reviewed the medical records of 160 patients with AAV. We calculated Birmingham vasculitis activity score (BVAS). We collected laboratory results including erythrocyte sedimentation rate (ESR), C-reactive protein (CRP), while blood cell, lymphocyte and platelet counts and serum albumin. Prognostic nutritional index (PNI) was calculated by $(10 \times \text{serum albumin (g/dl)} + 0.005 \times \text{lymphocyte count (/mm}^3))$. The association was assessed linear regression analyses. The optimal cut-off of PNI for predicting relapse was set at 36.6. Comparison of cumulative relapse free survival was analysed by the Kaplan-Meier survival analysis.

Results: The mean age at diagnosis was 55.2 years and 48 patients were male. Eighty-five patients had MPA, 41 patients had GPA and 34 patients had EGPA. The mean BVAS and PNI at diagnosis were 11.9 and 43.4. Forty-three patients experienced relapse of AAV. In univariable linear regression analysis, BVAS was positively correlated with ESR and CRP and was negatively correlated with lymphocyte count, serum albumin level and PNI. In multivariable analysis, BVAS was the most significantly associated with only PNI (standardised $\beta = -0.296$). Patients having PNI at diagnosis ≤ 36.6 exhibited significantly lower cumulative relapse free survival rate than those having PNI at diagnosis > 36.6 ($p=0.002$).

Abstract SAT0547 – Table 3. Univariable linear regression analysis of BVAS and continuous variables in 160 patients with AAV

	Univariable analysis		
	Regression Coefficient (Crude B)	Correlation Coefficient (R=β)	p value
Demographic data at diagnosis			
Age (year old)	0.027	0.053	0.504
Acute reactants at diagnosis			
ESR (mm/hr)	0.044	0.218	0.006
CRP (mg/L)	0.023	0.169	0.033
Laboratory results reflecting inflammation at diagnosis			
White blood cell count (/mm ³)	0.000	0.104	0.189
Lymphocyte count (/mm ³)	-0.002	-0.214	0.007
Platelet count (x1,000/mm ³)	0.003	0.060	0.449
Serum albumin (g/dL)	-2.557	-0.262	0.001
PNI	-0.252	-0.307	<0.001

BVAS: Birmingham vasculitis activity score; ANCA: antineutrophil cytoplasmic antibody; AAV: ANCA associated vasculitis; ESR: erythrocyte sedimentation rate; CRP: C-reactive protein; PNI: prognostic nutritional index.

Abstract SAT0547 – Figure 2. The association of PNI at diagnosis with relapse of AAV during the follow-up. We selected the optimal cut-off of PNI at 36.6, divided patients into two groups according to the cut-off and conducted Kaplan-Meier survival analysis. Patients having PNI at diagnosis ≤ 36.6 exhibited significantly lower cumulative relapse free survival rate than those having PNI at diagnosis > 36.6 ($p=0.002$). PNI: prognostic nutritional index; AAV: antineutrophil cytoplasmic antibody.

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.

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SAT0548 PROGNOSTIC FACTORS FOR INTERSTITIAL LUNG DISEASE WITH MICROSCOPIC POLYANGIITIS

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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.

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SAT0549

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

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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.

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SAT0550

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

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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;