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**AB0572 EFFICACY AND SAFETY OF RITUXIMAB FOR ANCA ASSOCIATED VASCULITIS**

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**Background:** In Japan, rituximab (RTX) has become one of the dominant alternatives for ANCA associated vasculitis (AAV) even for elderly patients.

**Objectives:** The efficacy and safety of RTX for AAV patients was determined at our hospital.

**Methods:** Nineteen patients with AAV, including 14 newly diagnosed patients and 6 relapsed patients, had been treated with RTX. There were 10 males and 9 females. They were all MPO-ANCA positive; 16 were diagnosed with microscopic polyangiitis (MPA), 2 with granulomatosis with polyangiitis (GPA), and 1 with undifferentiated, according to the EMA classification of AAV. The mean age at RTX induction was 71.3 years (range: 40–82 years). The efficacy was evaluated by the BVAS score at the time of first induction and after 6 month treatment. Adverse events were recorded during the 6-month treatment.

**Results:** The mean of BVAS decreased from 17.3 (range: 7–35) at the first induction to 1.2 (0–4) at 6 month of RTX treatment. Of the 10 patients who could be followed-up for over 6 months, 7 patients achieved remission (BVAS=0) (remission rate: 70.0%). The titer of mean MPO-ANCA decreased 136.1 IU/mL (9.9–300 IU/mL) to 44.4 IU/mL (1.0–114.0 IU/mL) at 6 month. The dose of prednisolone decreased from 34.4mg/day (5–60mg/day) at baseline to 5.5mg/day (0–10 mg/day) at 6 month. The adverse events were as follows: 3 patients experienced reactivation of cytomegalovirus (CMV) with a CMV colitis, one patient with sepsis following urinary-tract infection, and one patient with bacterial pneumonia. One patient with PMA died of exacerbation of the disease itself.

**Conclusions:** In Japan, AAV, mostly MPO-ANCA-positive PMA, affects elderly persons. The results suggest that RTX may provide a therapeutic option for elderly patients at the age of over 70 years with severe MPA, with an acceptable safety profile and rapid glucocorticoid tapering. However, careful monitoring for infectious diseases will be needed.

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**AB0573 AUTOLOGOUS STEM CELL TRANSPLANTATION IN THE MANAGEMENT OF REFRACTORY BEHÇET'S SYNDROME: A CASE SERIES FROM A SINGLE CENTRE OF EXCELLENCE**

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**Background:** Evidence regarding haematopoietic stem cell transplantation (HSCT) in refractory Behçet's Syndrome (BS) is limited. A systematic review in 2014 identified 20 patients with BS who had undergone HSCT, from which 9 patients underwent HSCT with refractory BS as the primary indication. Of these 20 patients: 15 achieved complete remission, 1 patient achieved partial remission, 2 patients relapsed, 1 patient died and 1 had no outcome reported. Nine patients had reported complications, 5 patients had no complications and 6 patients were missing data. All data was sourced from single case studies (1).

**Objectives:** Outline the indications, outcomes and complications of HSCT in a case series of 4 patients with BS managed at a single centre of excellence.

**Methods:** Case notes were reviewed for 4 patients from the Liverpool Behçet's Centre of Excellence (LBCE) who have undergone HSCT.

**Results:** The primary indication for 3 of the 4 patients who have undergone autologous HSCT at LBCE was refractory BS. Patient 1 underwent HSCT for multiple myeloma, but suffered from severe BS with neurological and venous involvement and is now in partial remission with occasional mild muco-cutaneous ulceration, not requiring systemic immunosuppression. Patient 2 underwent HSCT for severe refractory neuro-Behçet's. This patient is now in complete remission after commencing azathioprine for oral ulceration. The indications for HSCT in patients 3 and 4 were severe muco-cutaneous ulceration and dermatological involvement refractory to numerous biologic agents with high ongoing steroid dependency. Patient 3 also had previous thrombophlebitis and patient 4 had previous gastro-intestinal ulceration. They are both currently in complete remission but follow up time is limited. Patient 3 had a post-HSCT complication of pneumonia and mucositis which resolved without persistent morbidity, patient 4 had an upper limb deep vein thrombosis (DVT) as an inpatient.

**Conclusions:** HSCT is an effective treatment modality for severe refractory BS and can result in medication free remission. However, the associated risks should be considered and alternative treatment options deliberated or exhausted before opting for this treatment modality.

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**AB0574 VENOUS INVOLVEMENT IN BEHÇET'S DISEASE: A RETROSPECTIVE ANALYSIS OF 75 CASES**

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**Objectives:** Behçet's disease (BD) is a chronic multisystem inflammatory disease characterized by recurrent mouth and genital ulcerations. The vascular involvement varies from 7, 7 to 43% depending on ethnicity. Venous lesions are

**Abstract AB0573 – Table 1**

Patient	Age at HSCT (years)	Sex	Disease duration at HSCT (years)	BS manifestations	Treatment pre-HSCT	HSCT Indication	Follow up (months)	Complications	Outcome	Treatment post-HSCT
1	47	M	6	OU GU S MSK V N	Tacrolimus	Multiple Myeloma	135	Nil	OU GU (minor)	Pred 4mg
2	29	M	5	OU GU S Ocular N	MMF Tacrolimus	Neuro-Behçets	80	Nil	1x flare 2013 (OU)	AZA
3	51	M	4	OU GU S V N	AZA MMF ETN ADA Certolizumab Interferon	Refractory, severe OU & GU	1	DVT	Remission	nil
4	42	F	Unknown	OU GU S MSK Gastro	AZA Leflunomide MTX Thalidomide IFX ADA ETN ALN	Refractory, severe OU & GU	8	Pneumonia & Mucositis	Remission	Pred 40mg

OU = oral ulceration, GU = genital ulceration, MSK = musculoskeletal, S = skin, N = neurological, V = vascular. AZA = azathioprine, CYCP = cyclophosphamide, MMF = mycophenolate mofetil, MTX = methotrexate. ADA = adalimumab, IFX = Infliximab, ETN = Etanercept, ALN = Alemtuzumab.

the most frequent vascular manifestation in BD. In this study, we looked at the pattern and outcome of venous events in BD.

**Methods:** Seventy five patients, who fulfilled the criteria of the International Study Group for diagnosis of BD, were recruited. We studied the characteristics of patients with thrombotic venous events. Clinical data parameters were recorded, including age at onset, the vascular and extra-vascular manifestations of the disease.

**Results:** Twenty six from 75 patients had vascular event. Twenty three of these patients had a venous event. The mean age of the patients at the first venous event was 32 years. There were 22 males and 4 females. The first venous event occurred before BD diagnosis in one case and in the same time of onset of the disease in two cases. In the other cases, venous event occurred in patients followed for BD and the mean disease duration was 5, 82 years. The mean number of recurrence of venous events was 1, 46. Deep vein thrombosis was the most frequent single vascular event (76, 92%). The most frequent localizations were in legs (23 cases). Four patients had cerebral vein thrombosis. A pulmonary venous involvement, a Budd-Chiari syndrome, an inferior and superior vena cava syndrome and arms thrombosis were found in only one case each one. An arterial event was associated in 2 cases. An association with ocular manifestations was observed in 26, 9% patients and neurologic manifestations in 11, 53%.

**Conclusions:** Although there is no agreement on the frequency rate of the vascular lesions in the literature, most of the vascular lesions in the literature, most of the reported series indicate that the venous lesions are by far, more common than the arterial lesions. Kabbaj et al. reported 85% venous, 10% arterial and 5% mixed arterial and venous involvement which is similar to the reported frequency in our patients.

In conclusion, the frequency of vascular complications of BD in our patients is similar to those reported around the world.

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#### AB0575 RETROSPECTIVE SURVEY OF CONCOMITANT AUTOIMMUNE DISEASES AND AUTOANTIBODIES IN A COHORT OF PATIENTS WITH ANCA-ASSOCIATED VASCULITIS (AAV)

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**Background:** Anti-neutrophil cytoplasmic antibodies (ANCA) associated vasculitis (AAV) – granulomatosis with polyangiitis (GPA), microscopic polyangiitis (MPA) and eosinophilic granulomatosis with polyangiitis (EGPA) have heterogeneous clinic and biologic phenotypes. Sporadic reports indicate that some AAV patients may have other autoimmune diseases but the frequency of the association is unknown.

**Objectives:** Our purpose was to assess the frequency of other autoimmune diseases or autoantibodies in a well-defined cohort of AAV patients.

**Methods:** Retrospective survey of a cohort of patients regularly controlled at outpatient facility of an Autoimmune Disease Department in a 5-year period (2011–2016). Clinical and immunologic data were retrieved from electronic records. All patients were diagnosed and treated by the authors.

**Results:** We included 110 AAV patients – 36 (32.7%) GPA, 45 (40.9%) MPA and 29 (26.4%) EGPA. Regarding to ANCA specificity – 23 (20.9%) against proteinase 3 (PR3)-ANCA, 68 (61.8%) against myeloperoxidase (MPO)-ANCA and 19 (17.3%) were ANCA negative. Organ-specific auto-immune diseases were actively searched in 73 (81.1%) patients and was present in 43 (58.9%) patients. More prevalent autoimmune diseases diagnosed were: hypothyroidism (15,34.9%), pernicious anemia (12,27.9%), Sjögren syndrome (8,18.6%), hyperthyroidism (4,9.3%) and primary biliary cholangitis (3,7.0%). Autoantibodies more frequently positive were: ANAs 29 (67.4%), ASMA 27 (62.8%), anti-parietal cells 20 (46.5%), anti-thyroglobulin 6 (14.0%), anti-peroxidase 12 (27.9%), anti-SSa 5 (11.6%), anti-SSb 3 (7.0%) and RF 14 (32.6%). Prevalence of organ-specific autoimmune disease was higher in MPA (67.4vs.32.6%,p=0.001) and ANCA-MPO carriers (81.4vs.18.6%,p<0.001). Pernicious anemia was more prevalent in MPA (41.4vs.0%,p=0.027) and in ANCA-MPO (34.3vs.0%,p=0.054) and myositis was more prevalent in ANCA-PR3 (12.5vs.0%,p=0.037). ANAs positivity was more prevalent in ANCA-MPO carriers (74.3%vs.36.5%,p=0.048). Predictive factors for development of organ-specific autoimmune disease identified were: female

gender (OR 2.710,95% CI 1.034–7.099,p=0.043), MPA syndrome (OR 3.578, 95% CI 1.334–9.523,p=0.011) and MPO carriers (OR 3.346, 95% CI 1.166–9.601,p=0.025).

**Conclusions:** A substantial percentage of AAV patients, particularly MPA and anti MPO carriers, have associated autoimmune diseases and autoantibodies. The limitations of our study (retrospective assessment and lack of comparator) do not allow accurate estimation of prevalence. The severity of AAV and difficulties in management, may lead to overlooking of associated autoimmune diseases which appear to be frequent. Associated autoimmune disease may contribute to additional burden in AAV patients. (Supported by SAF 2014 57708-R).

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#### AB0576 ANTINEUTROPHILIC CYTOPLASMIC ANTIBODY-ASSOCIATED VASCULITIS: IS HYPOCOMPLEMENTEMIA A PREDICTOR OF POOR PROGNOSIS?

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**Background:** Antineutrophil cytoplasmic antibodies (ANCA) associated vasculitis (AAV) is a necrotizing vasculitis that predominantly affects small vessels and is associated with ANCA specific for myeloperoxidase (MPO-ANCA) or proteinase 3 (PR3-ANCA). The major clinicopathologic variants of AAV include microscopic polyangiitis (MPA), granulomatosis with polyangiitis (GPA) and eosinophilic granulomatosis with polyangiitis (EGPA). Recent studies have demonstrated the crucial role of complement activation in the pathogenesis of AAV. However, the clinical characteristics of AAV with hypocomplementemia (HC) still remain unclear. **Objectives:** The aim of our study was to analyze the demographic, laboratory, treatment and clinical characteristics of AAV using medical records. To compare the AAV patients with and without HC.

**Methods:** Retrospective study of patients with AAV diagnosed and followed in a specific Systemic Autoimmune Diseases and Thrombosis Unit. We defined HC as the state in which at least one of the following was lower than the lower limit of the normal range: Complement 3 (C3), Complement 4 (C4). Demographic, clinical, treatment and evolution data were recorded and analyzed using SPSS 22.0.

**Results:** Thirty-six patients with AAV were included (94,4% MPO-ANCA, 5,6% PR3-ANCA). 8 patients were diagnosed of GPA, 21 of MPA and 3 of EGPA. 61,1% of the patients were males and the mean age at the onset of the disease was 63,11±14,38 years [30–85]. 75% of patients had any vascular risk factors. Renal involvement occurred in 32 patients (88,9%): hematuria in 96,9%, proteinuria in 90,6% (>1 gr 68,7%) and glomerular filtration decreased in 81,25%. Biopsy was performed in 22 patients and a focal and segmental necrotizing glomerulonephritis with extracapillary proliferation was the finding more common. Pulmonary disease (61,1%) included interstitial disease 40,9%, alveolar hemorrhage 22,7% and nodules 18,2%. Other clinical manifestations were constitutional syndrome 36,1% (the main symptom was asthenia); ear, nose, and throat manifestations 33,3%; neurologic involvement 27,8% (the main finding was polyneuropathy); articular manifestations 33,3% and skin lesions 16,7%. All the patients received prednisone in combination with cyclophosphamide (69,4%) or rituximab (13,9%). Azathioprine was used as glucocorticoid-sparing agent (41,7%). 83,3% of the patients needed hospital admission and 6 died (16,7%). Eleven patients (30,6%) had HC at their diagnosis of AAV. Compared to the AAV patients without HC (n=25), we found no significant differences in the outcome or severity of the manifestations (evaluated by Five factor score). The small sample size could in part explain these results.

**Conclusions:** Our patients with HC at diagnosis of AAV did not have different characteristics than those without HC. More studies are needed to determine if HC is a predictor of poor prognosis in AAV patients.

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#### AB0577 SYSTEMIC AUTOIMMUNE VASCULITIS IN RHEUMATOID ARTHRITIS – A POSTMORTEM CLINICOPATHOLOGIC STUDY OF 161 PATIENTS

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**Background:** Systemic vasculitis of autoimmune origin (A-SV) may be regarded as a basic histological manifestation of rheumatoid arthritis (RA) [1,2].

**Objectives:** The aim of this study was to characterize the severity of systemic autoimmune vasculitis at death in RA patients.

**Methods:** A randomized autopsy population of 161 in-patients with RA was studied.

A-SV complicated RA in 32 (19.88%) patients [3].

RA was confirmed clinically according to the criteria of the ACR [4].

The severity of vasculitis was evaluated by semi-quantitative, visual estimation on a 0 to 3 plus scale in 12 organs (heart, lung, liver, spleen, kidney, pancreas, gastrointestinal tract, adrenal gland, skeletal muscle, peripheral nerve, skin and brain) at death [1]. ("0": no vasculitis, "1": sporadically (scattered) located