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POS0831

EARLY INFECTIOUS RISK IN PATIENTS WITH NEWLY-DIAGNOSED ANTI-NEUTROPHILCYTOPLASMIC ANTIBODY-ASSOCIATED VASCULITIS ACCORDING TO THE REMISSION-INDUCTION THERAPY: A FRENCH MONOCENTRIC RETROSPECTIVE STUDY INCLUDING 145 PATIENTS

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Background: Few comparative data exist on early infections secondary to remission-induction therapy (RIT) with rituximab versus cyclophosphamide in newly-diagnosed ANCA-associated vasculitis (AAV) patients.

Objectives: We compared and analyzed the rate and predictors of severe infections in such patients within the first six months following RIT.

Methods: We included, from the databases of Caen University Hospital, all consecutive adults newly-diagnosed with granulomatosis with polyangiitis or microscopic polyangiitis between January 2006 and December 2019. We compared the survival without severe infections (WSI) and the survival without infection of any severity (WIOAS) within 6 months from the RIT, and used a multivariate cox analysis to identify predictors of infection.

Results: We included 145 patients, 27 in rituximab group and 118 in cyclophosphamide group. Patients in the rituximab group more frequently had pneumococcal vaccination ($p < 0.01$) and creatinine level $< 150 \mu\text{mol/L}$, while other characteristics, including Birmingham Vasculitis Activity Score, were comparable between both groups.

Overall, 37 severe infections and 65 infections of any severity were recorded. The survival WSI was similar in both groups ($p = 0.69$), but survival WIOAS was lower in rituximab group ($p = 0.005$).

In multivariate analysis, risk factors at diagnosis for severe infections were chronic urinary tract disease, dialysis and absence of prophylaxis with trimethoprim-sulfamethoxazole ($p < 0.01$ each).

Conclusion: The survival WIS within the 6 months following RIT was similar in patients with newly-diagnosed AAV treated by rituximab or cyclophosphamide, but survival WIOAS appeared to be lower within the 6 months following rituximab despite a better pneumococcal vaccination coverage.

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Scleroderma, myositis and related syndromes

POS0832

A NOVEL GREY SCALE AND POWER DOPPLER ULTRASONOGRAPHIC SCORE FOR IDIOPATHIC INFLAMMATORY MYOPATHIES: SIENA MYOSITIS ULTRASOUND GRADING SCALE

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Background: No clear-cut guidelines exist about the use of diagnostic procedures for idiopathic inflammatory myopathies (IIM) and only scanty and conflicting data report the use of ultrasound (US).

Objectives: We aimed to assess if grey-scale (GS) and Power Doppler (PD) US, graded with a 0-3-points-scale, may be a reliable tool in a cohort of patients affected by IIM.

Methods: We prospectively collected, since July to October 2020, all patients referred to Vasculitis and Myositis clinic, Rheumatology Unit, University of Siena, for suspected IIM, as well as patients with a previous, definite diagnosis of IIM and evaluated during follow-up or referred from other centers for a second opinion. All patients underwent US examination of both thighs in axial and longitudinal scans. Edema and atrophy, both assessed in GS, and PD, were graded with a 0-3-points-scale. Spearman test was used to identify the correlations between US and clinical and serological variables.

Results: A total of 18 patients was included. Four of them were evaluated twice, at baseline and within 3 months of therapy. Muscle edema was found to be directly correlated with physician global assessment (PhGA), serum myoglobin and PD and negatively with disease duration. PD score was positively correlated to PhGA and negatively to disease duration. Muscle atrophy directly correlated with Myositis Damage Index and patients' age. The single-thigh sub-analysis evidenced a direct correlation between PD score and Manual Muscle Test.

Conclusion: In our cohort, we found that edema and PD are strictly related to early, active myositis, suggesting that an inflamed muscle should appear swollen, thickened and with Doppler signal. Conversely, muscle atrophy reflects the age of the patient and the overall severity of the disease. Such findings shed a new, promising, light in the role of US in diagnosis and monitoring of IIMs.

Figure 1. Comparison of the 6-month survival without severe infection (A) or without infection of any severity (B) in patients with newly-diagnosed ANCA-associated vasculitis treated by rituximab or cyclophosphamide

Table 1. Siena Myositis Ultrasound Grading Scale (SMUGS).

	Grey-scale edema	Grey-scale atrophy	Power Doppler
0	Normal muscle echotexture with hyperechoic septa and hypoechoic muscle fibers, conserved thickness.	Normal muscle echotexture, with hyperechoic septa and hypoechoic muscle fibers, conserved thickness.	No PD signal.
1	Focal hypoechoic areas, where septa are less evident. Conserved thickness.	Focal heterogeneously hyperechoic areas, where septa are thicker and more evident, and muscle fibers are thinner. Conserved muscle thickness.	One or two PD signals in at least one muscle (PD vascular spots, small vessels of homogenous diameters, vessel diameters approximately not superior to fibrous intramuscular septa)
2	Diffuse and heterogeneous hypoechogenicity (rectus femoris as hypoechoic or more than vastus intermedius), septa diffusely less evident. Conserved thickness.	Diffuse and heterogeneously hyperechoic muscle, with thicker septa and thinner muscle fibers. Conserved muscle thickness.	More than 2 PD signals for each muscle (as vascular spots, small vessels of homogenous diameters, vessel diameters approximately not superior to fibrous intramuscular septa).
3	Diffuse and heterogeneous hypoechogenicity (rectus femoris as hypoechoic or more than vastus intermedius), septa diffusely less evident. Increased thickness (rectus femoris became thicker than vastus intermedius).	Diffuse and heterogeneously hyperechoic muscle, with thicker septa and thinner muscle fibers. Reduced muscle thickness.	More than 2 PD signals for each muscle with larger diameter of the vessel (at least superior to fibrous intramuscular septa), or vessels with different diameters or branched vessels.

Figure 1. Different PD findings (clockwise) in longitudinal anterior scans of the thigh: PD 3 in a patient with a recent diagnosis of anti-Mi2 DM; PD 2 in the same patient after one month of treatment with steroids and Methotrexate; PD 1 in a patient affected by anti-SAE DM, with a suspected disease flare; PD 0 in a patient affected by an advanced polymyositis diagnosed in 2000, currently not in treatment.

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POS0833

A RETROSPECTIVE COHORT STUDY IN CHINESE PATIENTS WITH ADULT POLYMYOSITIS AND DERMATOMYOSITIS: RISK OF COMORBIDITIES AND SUBCLASSIFICATION USING MACHINE LEARNING

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Background: Idiopathic inflammatory myopathy (IIM), also known as myositis, refers to a group of heterogeneous disorders including polymyositis (PM), dermatomyositis (DM), inclusion body myositis and immune-mediated necrotising myopathy. Phenotype, pathogenesis, and prognosis vary due to multi-organ involvement and comorbidities. With the clinical application of MSAs, a new classification system for myositis was explored to reduce confusion between subgroups. But it is far from showing the full picture of myositis due to high heterogeneity. Therefore, it is necessary to systematically evaluate the relevant risk factors of myositis for ILD, other rheumatic diseases, and malignancy for better clinical vigilance. And further exploring the subclassification of myositis is critical.

Objectives: To identify the risk factors in Chinese patients with adult polymyositis and dermatomyositis for their comorbidities and explore a subclassification system.

Methods: Clinical records of 397 patients with idiopathic inflammatory myopathies were retrospectively reviewed. Logistic regression was used to identify potential risk factors for interstitial lung disease (ILD), other rheumatic diseases, and malignancy after bivariate analysis. Hierarchical clustering and decisional tree were utilized to identify subgroups and explore a subclassification system.

Results: A total of 119 polymyositis and 191 dermatomyositis patients were included. Anti-PM/ScI, anti-Ro52, anti-aminoacyl-tRNA synthetase and anti-MDA5 (adjusted odds ratios (AOR)=4.779, 1.917, 5.092 and 7.714 respectively) antibodies were risks ($p<0.05$), whereas overlapping malignancy was protective (AOR=0.107; $p=0.002$) for ILD across polymyositis, dermatomyositis and the total group. In subgroup models, Raynaud's phenomenon, arthralgia and semi-quantitative anti-nuclear antibody (AOR=51.233, 4.261, 3.047 respectively) were risks for other overlapping rheumatic diseases ($p<0.05$). For overlapping malignancy, male and anti-TIF1 γ antibodies (AOR=2.533, 16.949) were risks ($p<0.05$), whereas disease duration and combination of ILD (AOR=0.954, 0.106) were protective in the total group ($p<0.05$); while anti-NXP2 antibodies were identified as risk factors (AOR=73.152; $p=0.038$) in polymyositis. Hierarchical clustering suggested a subclassification with 6 subgroups: malignancy overlapping dermatomyositis, classical dermatomyositis, polymyositis with severe muscle involvement, dermatomyositis with ILD, polymyositis with ILD, and overlapping of myositis with other rheumatic diseases according to the characteristics of grouped patients. Accuracy of the classification and regression trees model was 0.768 (95% CI 0.711 to 0.819) on training set and 0.633 (95%CI 0.499 to 0.754) on test set.

Conclusion: Accompanying ILD, other rheumatic diseases and malignancy are strongly associated with clinical manifestation and myositis-specific or myositis-associated autoantibodies among Chinese polymyositis and dermatomyositis patients. The subclassification system proposed a more precise phenotype defining toward stratified treatments.

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POS0834

LONG-TERM OUTCOME OF SSC ASSOCIATED ILD: IMPROVED SURVIVAL IN PPI TREATED PATIENTS

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