

($p=0.3$). The median SUVmax was 3.0 [1.8–8.6] without significant difference between GCA and TA: 3.4 (2.1–8.6) versus 2.6 (1.8–7.1) ($p=0.4$), respectively. Eleven PET (61%) were performed under treatment, which consisted of steroids with a median dose at 30 mg/day [3–240]. Among 11 patients with active disease, 8 had inflammatory patterns and 3 had normal PET/MR, i.e. a sensibility of 73%, and the sensibility increased to 100% in patients with active TA disease. Median SUVmax were 4.7 [2.1–8.6] in patients with active disease versus 2 [1.8–2.6] in patients with remission ($p=0.003$).

Conclusions: PET/MR is a new hybrid modality of imaging which is interesting for the diagnosis and the follow-up of large-vessel vasculitis.

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AB0653 PREDICTION OF RELAPSES IN AUTOIMMUNE LARGE-VESSEL VASCULITIS – TOWARDS PERSONALISED IMMUNOSUPPRESSIVE TREATMENT STEWARDSHIP

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Background: Giant cell arteritis (GCA) is an autoimmune disease of the large arteries. Treatment consists in long-term immunosuppression with glucocorticoids (GC). About half of the patients have disease flares ('relapses') despite standard therapy. Tocilizumab (TCZ) – an anti-IL6 receptor antibody – is highly effective in reducing relapses, but has high costs.

Objectives: Here, we tested whether the initial clinical presentation and/or immunological findings might predict a GCA patient subset with poor response to GC. These might benefit from early TCZ therapy.

Methods: We performed a chart review on 113 patients from our prospective GCA cohort over the first two years after diagnosis. All had a follow-up of at least three months (median follow-up 24 months, IQR 12.6–24). Clinical findings at diagnosis, routine labs (at 0, 1, 3, 6, and 12 months) and therapy information (drug and dose) were extracted from the electronic database.

Relapses were defined as the presence of GCA-related symptoms (ischaemic pain, polymyalgia (PMR)) and/or elevated systemic inflammation parameters (CRP, ESR) that responded to an increase in GC-dose. GC receptor (GCR) expression levels in T cells were assessed using flow cytometry. Patients were genotyped for two polymorphisms in the Glucocorticoid receptor gene (NR3C1) that have been associated with steroid-responsiveness in other autoimmune disease (Systemic lupus, Pemphigus...).

Results: Over the first 12 months, 50,6% experienced at least one relapse. The majority of relapses occurred after three months of treatment (median time to relapse 102 days, range 19–312). This is when the GC dose is tapered below 20 mg/d. 'Relapsers' had an average of 1.66 (range 1–4) relapses in the first year. Patients with fever at initial presentation had a 2.2-fold (CI 1.1–5.05) higher risk to experience relapses ($p=0.02$). Other clinical findings were not associated with subsequent relapses. Low lymphocytes after the first month of therapy was the only lab value associated with relapse free follow up (698/ul vs 1021/ul, $p=0.02$). This was independent of the cumulative GC dose that 'non-relapsers' and

'relapsers' received in this period (1747 mg vs. 1710 mg mg, $p=0.4$). Relapsers had lower GCR expression levels, as assessed by flow cytometry.

Conclusions: Fever, lack of lymphocytopenia after one month of therapy and low GCR expression are risk factors for relapses in GCA. Low GCR expression combined with absence of lymphocytopenia during high dose GC therapy points at a constitutional steroid-resistance in relapsers. Whether patient stratification based on these parameters allows to safely adapt ('personalise') the intensity and/or duration of GCA treatment needs to be tested in a prospective clinical trial.

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AB0654 CLINICAL PROFILE AND RISK FACTORS OF INFECTIONS IN PATIENTS WITH ANCA-ASSOCIATED VASCULITIS (AAV) – 18-YEAR DATA FROM A SINGLE TERTIARY CENTRE

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Background: Despite advances in the treatment of AAV, there are still considerable morbidities related to treatment-related complications. Infection is one of the most commonly encountered problems in patients on immunosuppressive therapy.

Objectives: This single centre, retrospective study reviewed the clinical features and investigated the risk factors for infections among patients with AAV.

Methods: 104 patients with AAV diagnosed between January 2000 and December 2017 in a tertiary hospital were included. Demographic data and clinical parameters were reviewed. Logistic regression was performed to identify factors predicting infections.

Results: Around two-thirds of the 104 patients included were female (63.5% $n=66$). Mean age at diagnosis was 64.4-year-old. The majority (65.4%, $n=68$) had microscopic polyangiitis (MPA), 19.2% ($n=20$) had granulomatosis with polyangiitis (GPA) and 15.4% ($n=16$) had eosinophilic granulomatosis with polyangiitis (EGPA).

More than half of the patients (58%, $n=61$) experienced at least one episode of infection and 22% ($n=23$) had recurrent sepsis during their disease course. Infection was the leading cause of mortality of the 56 deceased patients in this series. Infections were less frequent in EGPA patients compared to their counterparts with MPA and GPA (37.5% vs 61.8%–65%).

Most infections were bacterial and multiple-drug resistant organisms were the causative agents in 8 patients. Two had neutropenic sepsis. Three had M. tuberculosis and five had herpes zoster. One had concomitant VZV and pneumocystis jiroveci pneumonitis.

Abstract AB0654 – Table 1.

Factors predicting infection	Odds Ratio ($p<0.05$)
– · Age	1.034
– · Renal insufficiency (serum creatinine ≥ 140 umol/L) at diagnosis	5.452
– · Disease-related organ failure	3.006
– · Dialysis support	6.504
– · Use of cyclophosphamide as induction agent	2.956

Conclusions: Infections were common and often led to significant morbidities and mortalities among AAV patients. Risk factors included age, renal insufficiency (serum creatinine >140 umol/L) on presentation, disease-related organ failure, need for renal supportive therapy, and the use of cyclophosphamide as induction agent.

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AB0655 AGREEMENT BETWEEN 18-FDG PET/CT AND CLINIMETRIC TAKAYASU ACTIVITY SCORES

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Background: The 18-FDG PET/CT is an objective tool employed for the diagnosis of Takayasu arteritis and also is used for the assessment of disease activity of this vasculitis.¹ There are few clinimetric scores developed for Takayasu clinical activity assessment such as the Indian Takayasu Clinical Activity Score (ITAS2010/ITAS.A),² the National Institutes of Health criteria (NIH score) from USA³ and the Mexican effort by Dabague-Reyes (DR score).⁴ The validity and

utility of 18-FDG PET/CT to measure the disease activity by studying wall enhancement compared to the clinimetric assessment has been slightly studied.

Objectives: To explore the agreement between 18-FDG PET/CT and the clinimetric tools for the estimation of Takayasu activity in one national reference centre.

Methods: The clinical records of patients that had performed an 18-FDG PET/CT were consecutively included. The required information to fulfil the ITAS2010, ITAS.A, NIH and DR were gathered from clinical charts. The cut-off points we used are the following: SUVmax ≥ 2.1 for 18-FDG PET/CT, for ITAS2010 ≥ 2 points, for ITAS.A ≥ 4 points, for NIH ≥ 2 points and for DR ≥ 5 points. Kappa index was calculated, comparing SUVmax with all the clinimetric measures. As an exploratory exercise, ROC curves were performed. A p value less than 0.05 was considered statistically significant.

Results: Thirty six clinical records were reviewed. There was enough information to score ITAS2010 in 31 patients, ITAS.A in 28 patients, NIH and DR in 35 patients each. In our patients, moderate agreement was observed between 18-FDG PET/CT and DR score (Kappa=0.542, p=0.001). A tendency of weak agreement was observed with the NIH score (Kappa=0.215, p=0.086) and ITAS.A (kappa=0.351, p=0.063). There was no agreement with ITAS2010 (Kappa=0.107, p=0.519). Significant AUC were observed with DR (AUC=0.817, p=0.005) and NIH (AUC=0.756, p=0.025); however, this results were not obtained with ITAS2010 (AUC=0.675, p=0.124) and ITAS.A (AUC=0.697, p=0.083).

Conclusions: There was no strong agreement between 18-FDG PET/CT and any of these activity indices. On the other hand, these data suggest that the best disease activity tool in Mexican patients were DR and the NIH scores. Comparative studies in other populations are warranted.

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AB0656 CRYOGLOBULIN EVALUATION: ANALYSIS OF INTRA-LABORATORY AND INTER-LABORATORY VARIABILITY

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Background: Cryoglobulins (CRG) are immunoglobulins that precipitate in serum at temperatures below 37°C and resolubilize upon warming. The main reasons of interest of a clinical pathologist in the study of cryoglobulinemia are: 1) lack of standardisation in the preanalytical, analytical and postanalytical phases of the process (classification and reporting); 2) peculiarities of physiopathological mechanism 3) important clinical consequences. Vermeersch et al. studied these issues in 2008. To assess current practice in the detection, analysis, and reporting of cryoglobulins, a questionnaire was sent to 140 laboratories. They showed that only 36% of laboratories used standard procedures of analysis. Consequently, they concluded that standardisation was needed for cryoglobulin detection to avoid missed diagnoses and improve the comparability of results. Sargur et al. in 2010 reviewed the classification and clinical features of cryoglobulins and suggested “best practice” guidelines for laboratory detection and identification of cryoglobulins. They particularly highlighted the relevance of preanalytical and analytical phases: maintenance of the sample at a stable temperature of 37°C, especially throughout the initial steps (collection and transportation); centrifugation and separation methods; cryoprecipitate quantification; cryoprecipitate washing techniques; immunocharacterization of cryoprecipitates especially through immunofixation techniques (considered the “gold standard”).

Objectives: To verify and assess the variability of laboratory processes of CRG.

Methods: We checked laboratory databases of Hospital and University (Lab A and B) of Modena with long tradition in the cryoglobulin analysis (more than 6000 tests from 2002 to 2017). Concerning CRG testing, 734 patient samples were studied in both laboratories. We compared our results according to Brouet classification into subgroups: type I, II and III. Therefore, we evaluated intra-laboratory variability, compared to previous or more frequent results. Finally, we studied inter-laboratory variability based on non-concordant laboratory reports.

Results: In the following table, we have represented the comparison between labs about the same patient cohort in 734 patient samples:

Abstract AB0656 – Table 1.

	I type (n)	II type (n)	III type (n)
Lab A	21	242	108
Lab B	42	270	108
Chi-quadro	p=0.0016	p=0.0004	Ns

Intra-laboratory variability: 14% Lab A, 16% Lab B (ns). Inter-laboratory variability: non-concordance in 25% of cases, considering 133 patients studied in both laboratories (Chi-square test).

Conclusions: No data about variability in CRG analysis are reported in literature. National and international guidelines are not explicative enough. Furthermore, many doubts about classifications are established. Our experience is unique but limited in two laboratories. Given the variability of testing conditions used in different laboratories and the lack of test standards and reference values, we confirm the need of further investigations into standardisation of CRG testing. New guidelines are fundamental, in order to optimise all phases of CRG research (pre and post analysis) and to ensure correct diagnosis and adequate treatments of the associated diseases.

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AB0657 CYCLOPHOSPHAMIDE-SPARING ROLE OF AN INTENSIFIED B-CELL DEPLETION PROTOCOL IN ANCA-ASSOCIATED VASCULITIS: A CASE-CONTROL STUDY

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Background: The management of ANCA-associated-vasculitis (AAV) requires the use of immunosuppressive drugs with potential toxicity. Recently, two trials demonstrated the efficacy of Rituximab (RTX) for the therapy of AAV.

Objectives: this case-control study aims to evaluate the immunosuppressive-sparing effect of a RTX-based protocol compared to the standard CYC treatment.

Methods: 26 patients with AAV and extracapillary glomerulonephritis were prospectively enrolled. Thirteen patients received an intensified protocol of B-cell depletion therapy (IBCDT) consisting of 4-weekly infusions of 375 mg/sm RTX followed by 2 infusions after 1 and 2 months, 3 pulses of methylprednisolone followed by prednisone tapered to 5 mg/day in three months and 2 pulses of 10 mg/kg CYC, without further maintenance therapy. Thirteen patients treated with 2 mg/kg/day CYC followed by azathioprine as a maintenance therapy served as controls.

Results: A significant improvement (p<0.05) of B-VAS, ESR, CRP and ANCA was observed in the IBCDT-group at 3, 6 and 12 months, with decrease of mean creatinine values from 4.81±6.4 mg/dl to 2.21±3.8 mg/dl.

When compared to controls, no difference was observed in terms of complete and partial response. However, the IBCDT regimen achieved a 1 g/month reduction of CYC cumulative dose (p<0.001).

Conclusions: in the treatment of this sample of severe AAV patients, the IBCDT protocol appeared to be noninferior to CYC-based regimen. Notably, the IBCDT regimen allowed a significant reduction of CYC cumulative dose.

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AB0658 DIFFERENT ORBITAL MANIFESTATIONS OF GRANULOMATOSIS WITH POLYANGIITIS. COMPARATIVE STUDY

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Background: Ophthalmic manifestations are typical for granulomatosis with polyangiitis (GPA), and occur in 28.6%–60% of patients. In 8% of cases they lead to permanent visual loss. According to different studies orbital lesion develops in 5%–30.6% of GPA patients and is considered to be the second most prevalent ophthalmic manifestation after conjunctivitis/episcleritis.

Objectives: to study clinical features of different orbital manifestations of GPA.

Methods: 74 GPA patients with orbital involvement were studied and compared. 3 types of orbital involvement were proposed: orbital mass (45 patients),