AB1151

POLYMYALGIA RHEUMATICA. NEW THERAPEUTIC STRATEGY BASED ON LOW DOSE OF METROTEXATE PLUS LOCAL INFILTRATION WITH CORTICOSTEROIDS

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Background: The polymyalgia rheumatica is a rheumatic inflammatory disease more frequent in the eldest population. The classic therapy is based on medium doses of corticosteroids followed by a maintenance phase of low doses, generally lasting from 1–2 years. Recurrences frequently require an escalation of dose, thus lengthening the treatment time, and that entails important comorbidity. Methotrexate (MTX) has been tested in 3 randomised clinical trials, showing in two of them the efficacy as a steroid sparing agent, but it has never been tested as monotherapy.

Objectives: To analyse the results of an alternative therapy in order to avoid the oral corticotherapy through the use of low dose of MTX and joint infiltration with a total duration of 24 months.

Methods: A prospective observational study in which patients that had been diagnosed with 2012 EULAR/ACR criteria were evaluated in outpatient medical consultations of Rheumatology at "Doce de Octubre" Hospital between 2015–2017, with the restriction of not having received previous steroid treatment. Right after diagnosis, the treatment with MTX 5–7.5 mg/w plus the infiltration of triamcinolone acetone in both shoulders begins, being repeated if necessary after 15 or 30 days, or in case of subsequent relapse.

Results: 26 patients were included, with an average follow-up 19±5 months.^{12–24} The age average at diagnosis was 74±7 years,^{55–83} being 56% women. 73% had symmetrical hand arthritis and 27% structural pathology of the rotator cuff. 96% had moderate-severe pain (VAS) in shoulders, 73% in hips and 54% in hands. These percentages after a month of treatment were 15%, 11.5% and 7.7%. From the onset of symptoms until the start of treatment 104±54 days passed,^{30–180} applying an initial dose of 5 mg of MTX in half of cases and 7.5 mg in the other half. There are no significant differences between precocity of the treatment or initial dose regarding a faster remission. The average of infiltrations in the shoulder per year is 2±1.4 (1–4). 25% of patients showed reactivation with good response to the MTX dose increase [maximum dose 8±1.7 (5–12.5)] plus/or joint infiltration. The average time until the revision (subjective clinical evaluation, HAQ and APR) was 2.7±1 months,^{1–6} being significantly higher ($p<0.05$) in patients with peripheral arthritis 3.3±1.9 vs 1.3±0.7. The change of HAQ, CRP and other variables are indicated in the table.

The treatment was stopped in 12% because of adverse effects (digestive intolerance, alopecia and respiratory infection in a patient with COPD). A patient showed mild plaquetopenia in which not necessary to suspend the treatment with MTX.

8 patients were monitored during ≥2 years, in 87% of them MTX has been removed with an average follow-up after the suspension of 3±1.7 months^{1–6} with no need to restart it in any case.

Abstract AB1151 – Table 1

	Start	1 Month	3 Months	6 Months	12 Months
Morning stiffness>1 hour	69%	38%	4%	4%	4%
HAQ	2	0.750	0.300	0.500	0.125
ESR	43,2	20	20	15	13
CRP mg/dl (0–0,5)	4,1	0,81	0,44	0,48	0,47
Peripheral arthritis	77%	23%	7%	11%	7%
Shoulder limitation	100%	69%	26%	23%	20%

Conclusions: The use of low dose of MTX and local joint infiltration with corticosteroids (initial and on demand) is an efficient therapeutic strategy with a low complication rate in PMR. All these results must be confirmed with controlled studies and a longer period of follow-up after the suspension of treatment.

Disclosure of Interest: None declared

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AB1152

MUSCULOSKELETAL MANIFESTATION OF DIABETES MELLITUS IS HIGHLY PREVALENT AND IS ASSOCIATED WITH POOR DIABETIC CONTROL

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Background: Diabetes mellitus is one of the most common medical conditions all over the world. A variety of musculoskeletal (MSK) conditions have been associated with diabetes mellitus (DM). These MSK symptoms are important to recognise because these can respond to treatment. Estimates of the prevalence of MSK problems in patients with DM vary depending upon the definitions used for the problems and the study population with diabetes, which can range from a primary care cohort to patients with severe diabetes in a specialised referral setting.

Objectives: The objectives of our study were: 1) to investigate the prevalence of musculoskeletal complications of diabetes in a consecutive cohort of patients attending a secondary care endocrinology clinic; and 2) to identify clinical associations of such musculoskeletal complications by examining likely contributing features including patient characteristics, life style factors and features of underlying diabetic disease.

Methods: The study participants were all consecutive patients attending endocrinology clinics of University Hospital Kerry for the management of their DM. Patients with chronic inflammatory arthritis or chronic autoimmune diseases were also excluded. This was a questionnaire-based study and was carried out in two steps. The first step involved having a short interview and assessment of the patient, and in the second step their clinical records were reviewed to populate clinical parameters. The clinical variables studied were gender, smoking habits, body mass index (BMI), units of alcohol intake per week, smoking habits, type of DM, duration of DM, medications used for DM, employment status and also their current HbA1c blood results. End organ complications of DM were also included. Moreover, personal history of cardiovascular risk factors and diseases were also recorded. Since there is a long list of rheumatologic manifestations which are potentially associated with DM and many of them are less sensitive and specific for DM, we choose to study the 4 common DM-associated rheumatologic presentations (Stiff hands, Carpal Tunnel Syndrome, Charcot joint, bilateral shoulder rotator cuff tendinopathy). Moreover, importantly, only patients with the musculoskeletal symptoms lasting for >3 months were included in the analysis. These 4 DM-associated musculoskeletal diseases are labelled as DM-MSK diseases.

Results: A total of 250 patients [mean age 66±16 years; 58% male; mean duration of DM 13±10 years; mean BMI of 27.5±6] were evaluated. DM-MSK diseases were present in 37.6% (n=94) of the entire cohort.

On univariate analysis, patients with older age, type-2 DM, using hypoglycaemic agents, hypertension, ischaemic heart disease, peripheral vascular disease, cerebrovascular accident, congestive heart disease, and patients with renal involvement had significantly higher prevalence of DM-MSK symptoms. However, on multivariate analysis (table-3), poor diabetic control as reflected by higher HbA1c levels, presence of diabetic kidney disease and advancing age were associated with the presence of DM-MSK symptoms.

Conclusions: We conclude that MSK manifestations of DM are very common but unfortunately these remain poorly recognised and continue to cause significant disabilities. Similar to the microvascular and macrovascular complications, MSK manifestations are associated with poor diabetic control.

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AB1153

IDIOPATIC LOBULAR PANNICULITIS (DISEASE WEBER-CHRISTIAN): CURRENT ASPECTS

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Background: Idiopathic lobular panniculitis (ILP) (syn. Weber-Christian disease) is the least studied condition from the group of systemic connective tissue diseases, which is characterised by predominant involvement of subcutaneous fat (SCF) tissue.

Objectives: To evaluate the interrelation between clinical signs and lab parameters in ILP patients.

Methods: The study evaluated 67 patients (9 males and 58 females) aged 20–76 years with verified ILP diagnosis and mean disease duration of 78.91^{48;540} months who were on the record at V. A. Nasonova Research Institute of Rheumatology during 2007–2017 yy. Alfa-1 antitrypsin, liver fractions, amylase, lipase, trypsin, ferritin, creatine phosphokinase (CPK), leptin and TNF- α levels were measured, chest CT and histopathological study of skin and SCF lesions biopsy specimens were made in addition to conventional clinical examination.

Results: ILP was found to affect all age groups, with 57% of cases falling on able-bodied adults aged 45–60 yy. Based on clinical manifestations including location, distribution, spatial extent of lesions, and clinical course of the disease, the following 4 clinical forms of ILP were identified: nodular (30 patients), plaque-like,¹⁰ infiltrative¹⁵ and mesenteric.¹² ILP population demonstrated significant ESR ($p=0,01$) and CRP ($p<0,0001$) elevation. ESR elevation correlated with palpatory nodular pain intensity, assessed by visual analogue scale (VAS ($p<0,05$, $r=0,29$), with the amount of affected body surface area (BSA) measured using the hand area surface (HAS) to equal 1% BSA ($p<0,05$, $r=0,50$), with elevation of body temperature ($p<0,05$, $r=0,68$) and CRP ($p<0,05$, $r=0,68$). CRP elevation correlated with pain intensity measured by VAS ($p<0,05$, $r=0,46$), affected BSA ($p<0,05$, $r=0,61$) and presence of stage II nodules ($p<0,05$, $r=0,41$). Histopathologic features of skin and SCF biopsy specimens were studied in 65 patients (97,01%), including antero – and retroperitoneal fat tissue biopsy specimens from 3 patients out of 5 (5,97%)

with mesenteric panniculitis without skin and SCF involvement; the remaining 2 were not biopsied in hard to access areas. Histopathological study ruled out the probability of neoplasms and confirmed the diagnosis of lobular panniculitis in all specimens.

Therapy included such common in rheumatology practice agents as glucocorticosteroids, NSAIDs, cytotoxic drugs, hydroxychloroquine, and oths). Therapeutic success was documented in 62.68% cases, therapeutic failure and disease progression – in 17.91% (12 patients), requiring dose escalation and modification of therapeutic regimen.

Conclusions: Identified correlation between clinical features and lab parameters measuring disease activity confirms ILP as a systemic inflammatory disease of the connective tissue. There's a flagrant necessity to improve physicians awareness of ILP, as well as need in future studies to enable earlier ILP diagnosis and identify more effective treatment of the disease.

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AB1154

INCREASE GENERATION BUT DEFECTS OF SECRETING IFN- α PLAY A ROLE IN THE PATHOGENESIS OF IGG4-RD

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Background: IgG4 related disease (IgG4-RD) is a multi-organ involvement, fibro-inflammatory disease of unknown etiology. Both innate and adaptive immunity played vital roles in the pathogenesis of IgG4-RD. Plasmacytoid dendritic cells (pDC) had major roles in antigen presentation and secreting IFN- α upon infection. However, the characteristics and relevant function of this cell population in IgG4-RD was poorly understood. So we aim to study the expression and function of pDC in IgG4-RD.

Objectives: To study the expression and function of Plasmacytoid dendritic cells (pDCs) in IgG4-RD

Methods: Flow cytometry was performed to analyse the expression of pDC cells in untreated IgG4-RD patients (n=12) and healthy controls (n=12). The immunohistochemistry technique was used to assess the location of pDC in the involved tissues of IgG4-RD patients. Furthermore, by cells culture *in vitro*, the abilities of pDC secreting INF- α and the activation of NF- κ B signal in IgG4-RD were explored.

Results: The frequencies of pDC in the IgG4-RD patients were significantly higher in the peripheral blood and involved tissues compared with healthy controls. The cell surface marker of CCR7 in pDC was lower in untreated IgG4-RD patients, but after treatment, the expression of CCR7 increased. There was on significance of the expression of pDC maturation marker (CD83), activation marker (CD80, CD86), CCR1, CCR2, CCR3, CCR4, CCR5, CCR6, CCR8, CCR9, CCR10, CXCR2, CXCR3 and CX3CR1 compared with healthy controls. Interestingly, the frequencies of CD123hiCD303+cells were also higher in untreated patients, and reduced after treatment, and the cell surface marker CD83 was also elevated. By cell culture *in vitro*, pDC had defects in secreting INF- α of IgG4-RD patients than healthy controls.

Conclusions: The excessive infiltration of pDC in peripheral blood and tissue but less CCR7 Defects of secreting INF- α of pDC in IgG4-RD may indicate less function of eliminating infection which may induce constant infection. pDC may played vital roles in the pathogenesis of IgG4-RD,

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AB1155

IMMUNOGLOBULIN G4 – RELATED DISEASE, A DIAGNOSTIC CHALLENGE

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Background: The IgG4-related disease (IgG4-RD) is a chronic, inflammatory, multi-organic, systemic disease. The pathological assessment is the gold standard for the diagnosis, hallmarked by the lymphoplasmacytic tissue infiltration of mainly IgG4-plasma cells, the storiform fibrosis and the obliterative phlebitis.

Objectives: The aim of this study was to analyse the features of IgG4- RD cases followed in a specific autoimmune diseases unit, since 2013.

Methods: Descriptive, retrospective study, through the review of clinical charts. Medical records were reviewed for demographics information, clinical

presentation, underlying conditions, laboratory and radiological data, medical and surgical treatments and clinical outcomes. All patients diagnosed by biopsy were included (international consensus pathological criteria of 2012). We excluded the possible cases of IgG4-RD when biopsy was not available or did not meet the criteria previously mentioned.

Results: Eight patients with characteristic clinical, histological and laboratory features of IgG4-RD were included. The patients were predominantly male (87.5%), the mean age at diagnosis was 66±11 years.^{48–80} 87.5% of the cases were consulted from other hospital Services, mainly surgical: Urology 25% (hydrocele and urinary retention), General Surgery 25% (obstructive jaundice and forearm tumour), Vascular Surgery 25% (abdominal aortic aneurysm), Cardiac Surgery 12.5% (thoracic aortic aneurysm) and Neurology 12.5% (hypertrophic pachymeningitis). The most commonly involved organs were vascular 87.5% (6 aorta, 1 pulmonary veins) and retroperitoneum 75%, followed by renal 37.5%, pancreatic 12.5%, and central nervous system 12.5%. It was found isolated organic involvement in only one patient (autoimmune pancreatitis). Two patients had previous malignancy: renal cells and prostate. Magnetic resonance imaging, computed tomography and FDG-PET were made in 62.5% each of them. Serum IgG4 was determined in all cases, 62.5% of patients had normal IgG4 serum levels (the upper limit of normal for serum IgG4 is 135 mg/dL). Three patients (37.5%) had elevated serum IgG4, the mean level was 188±38 mg/dL ((152–266). The pathological findings were dense lymphoplasmacytic infiltrate in 6 cases (75%) with obliterative phlebitis in 5 of them, storiform-type fibrosis in 4 cases (50%) and IgG4/IgG ratio >40% in 75% of the samples. Glucocorticoids treatment was initiated in 75% of patients, 25% were untreated for predominantly fibrotic involvement. There were treatment response in 60% of cases. 25% had a relapse after corticoid withdrawal. Only one patient received a second drug due to lack of response to Prednisone (rituximab).

Conclusions: According to the literature, IgG4-RD patients were predominantly male in their sixth and seventh decade of life. The predominant involvement in our serie was vascular and retroperitoneal, with no cases of glandular manifestations. Highlight the high number of cases with normal serum IgG4. Most of our patients responded to corticosteroid therapy.

Disclosure of Interest: None declared

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AB1156

DRESS (DRUG REACTION WITH EOSINOPHILIA AND SYSTEMIC SYMPTOMS): LOOKS MAY BE DECEPTIVE!

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Background: Drug reaction with eosinophilia and systemic symptoms(DRESS) is a rare, potentially life threatening multisystem disorder with fever, skin eruptions, lymphadenopathy, eosinophilia and systemic involvement most commonly after a drug exposure. These cases may mimic various rheumatological conditions. We report a series of 14 cases of DRESS who presented to Rheumatology Clinic as suspected connective tissue disease or sepsis in Northern India.

Objectives: To highlight DRESS as a common mimic of common rheumatologic conditions and sepsis and report its etiology, characteristics, treatment and prognosis

Methods: We manually searched the inpatient records of Immunology Department in SGPGI 2007–18 for the cases discharged with a diagnosis of possible/probable/definite DRESS. The records of the patients with probable and definite DRESS according to regiSCAR criteria¹ were reviewed.

Results: All the 14 patients fulfilled clinical criteria for diagnosis (4 probable, 10 definite). The age of patients ranged from 9–53 years with majority in their 3rd decade. Majority of the patients were referred to us when their counts were rising in the setting of fever and skin rashes with a suspicion of rheumatic disease/sepsis. The clinical and laboratory features of these patients are as follows (table 1): TLC: Total Leucocyte Count, AEC: Absolute Eosinophil Count, ED: Exfoliative dermatitis, F: facial oedema, M: maculopapular rash, L: Liver, K: Kidney, P: Lung, G: GI, SSZ: Sulphasalazine, HRZE: Isoniazid, Rifampicin, Pyrazinamide, Ethambutol, FU: Follow up

All the patients were treated with oral steroids showing signs of clinical improvement within 4–5 days. Rashes and leucocyte count were first to respond. Transaminitis responded within a week. Mean Followup is 9.3 months. They were gradually tapered off steroids over next 3–4 months except for two patient who were lost to follow up.