articular and extra-articular damages and the need for prosthetics and the psychological status.

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FRI0493

THE INTERLEUKIN-1B INHIBITOR CANAKINUMAB FOR REFRACTORY STILL'S DISEASE: LONG-TERM EXPERIENCE IN 50 CONSECUTIVE PATIENTS

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Background: Interleukin-1 (IL-1) is a major mediator of the inflammatory cascade in Still's disease and an established therapeutic target.

Objectives: To assess the efficacy and safety of the IL-1b inhibitor canakinumab in adolescent and adult patients with refractory Still's disease.

Methods: We conducted a retrospective longitudinal outcome study of 50 consecutive patients aged 39 years (median, range 14-72), fulfilling the Yamaguchi disease classification criteria, with active disease despite treatment with corticosteroids (CS) (n=11) and/or methotrexate (n=9) and/or biologics (n=30) [tumor necrosis factor inhibitors (n=13), IL-6 blockade (n=7), abatacept (n=2), anakinra (n=24); ≥1 biologics (n=13)]. Canakinumab 150-300 mg was administered sc, starting every 4 (n=48) or 8 weeks (n=2), for a median of 24 months (range 3-84). Concomitant treatment included CS (n=41), methotrexate (n=12) and leflunomide (n=3).

Results: Complete remission was initially achieved in 78% of patients within a median time of 3 months, irrespective of age at disease onset. Partial clinical and laboratory response was evident in 20%. Canakinumab was discontinued in one patient with resistant disease (primary failure) and in 6 out of 10 initial responders, who relapsed during treatment (secondary failure). Of 39 patients in complete remission, increase in drug administration interval and/or drug dose reduction was attempted in 7, of which only 1 relapsed, whereas drug discontinuation was attempted in 19 patients for a median time of 8 months (range 3-68), of which 8 relapsed. Overall, in half of all disease flares, canakinumab re-introduction or intensification was successful. Canakinumab had a significant CS sparing effect permitting weaning in 21 of 41 cases. Infections (20%, severe 4%) and leucopenia (6%) led to treatment cessation in one patient.

Conclusion: In this largest so far real-life patient cohort with refractory Still's disease, high rates of sustained remission were induced by canakinumab both in adolescent and adult patients.

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RHEUMATIC IMMUNE RELATED ADVERSE EVENTS OF CHECKPOINT INHIBITORS: A RETROSPECTIVE REVIEW OF 70 PATIENTS

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Background: Immune Checkpoint inhibitors (ICI) have revolutionized cancer therapy by achieving remarkable survival benefits however, at the cost of a myriad of immune-related adverse events (irAEs)[1]. Rheumatic irAE can develop in 5-10% of patients although the true incidence is unknown given the lack of prospective studies [2]. Symptoms are heterogenous and probably underreported with few data available about their management and outcome [3].

Objectives: To describe the clinical, biological, and radiological features of the largest cohort of rheumatic irAEs from ICI along with their therapeutic management, outcome and follow-up in real-world practice.

Methods: A referral process for emergent rheumatic irAEs was initiated in February 2016 between the oncology and rheumatology departments at the Cleveland Clinic Foundation. All patients were evaluated by authors CC and/or LHC. Patients' characteristics were retrospectively collected from medical charts after IRB approval.

Results: 70 patients referred for one or more rheumatic irAEs between February 2016 and January 2020 were included. 66% were male, median age was 60.8 years. Among them, 24 (34%) had pre-existing rheumatic complaints. Melanoma was the most frequent malignancy (56%). ICI therapy included anti-CTLA4 (40%), anti-PD1/L1 (79%), and dual therapy ipilimumab/nivolumab (41%). Rheumatic irAE occurred in a median 4 months after ICI initiation, with phenotypes including inflammatory arthritis (32 patients), sicca-like symptoms (12), polymyalgia rheumatica-like (7), and myositis (2). Oral, intravenous or intraarticular glucocorticoids (GC) were administered to 54 patients (77%). Of these 54 patients, 22 (41%) required long term GC, 19 had bone density scan and 15 received pneumocystis (PJP) prophylaxis. One PJP case, 1 osteoporotic fracture and 2 avascular necrosis cases were reported. 16 patients received conventional DMARDS (23%) and 9 received biologics (13%). ICI therapy was held for rheumatic irAE in 31% of cases and for another systemic irAE in 29%. Median follow-up was 13.6 months, at end of follow-up 51 patients were still on treatment for rheumatic irAE and 41% of them were still symptomatic despite ongoing treatment.

Conclusion: Rheumatic irAEs are heterogeneous and often chronic requiring prolonged immunomodulatory therapy. Prospective studies are required to define optimal management of rheumatic irAEs that maintain long-term oncologic outcomes.

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FRI0495

FOLLOW UP OF INTERSTITIAL PNEUMONIA WITH AUTOIMMUNE FEATURES – THE EXPERIENCE OF ONE CENTRE

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Background: Interstitial Lung Diseases (ILD) may present features suggesting an underlying autoimmune process, which seem to differentiate them from idiopathic interstitial pneumonias, although without fully meeting the classification criteria (CC) for a specific connective tissue disease. Different terms had been used to describe these conditions and, to reach a consensus, the European Respiratory Society/American Thoracic Society proposed the CC for an entity named Interstitial Pneumonia with Autoimmune Features (IPAF). Clinical evolution and prognosis of this entity are still poorly understood.

Objectives: To evaluate clinical evolution and prognosis of a population of patients with IPAF.

Methods: Retrospective analysis of clinical files of patients followed by the Pulmonology Department since 02/2012 until 06/2019, who met the CC for IPAF, regarding clinical, functional and radiological evolution. Patients were considered to have a progressive phenotype in 24±3 months from their 1st evaluation if they fulfil 1 of the 4 criteria: relative decline in FVC ≥10% predicted; relative decline in FVC ≥5–<10% predicted and worsened respiratory symptoms; relative decline in FVC ≥5–<10% predicted and increased extent of fibrosis on High-resolution Computed Tomography (HRCT); worsened respiratory symptoms and increased extent of fibrosis on HRCT.

Results: 22 (7.4%) of 296 ILD patients met IPAF CC. 59.0% were female with an age at the 1st evaluation of 66.7±12.4 years. They were all non-smokers (63.6%) or ex-smokers (36.4%). Serologic and morphologic criteria were both present in 21 (95.4%) and clinical criteria in 5 patients (22.7%). Antinuclear antibodies (ANA) were identified in 19, rheumatoid factor in 4, SSA in 3 and anti-Jo-1 in 1 patient. HRCT patterns were identified in 21 patients: 15 nonspecific interstitial pneumonia (NSIP), 5 organizing pneumonia (OP) and 2 lymphocytic interstitial pneumonia (LIP). One NSIP and 1 LIP identified on HRCT were confirmed by histopathology. Three patients had inflammatory arthritis and 2 had Raynaud's phenomenon. Immunosuppressive therapy was introduced in most cases (18 patients, including systemic corticotherapy in 17, azathioprine in 4, mycophenolate mofetil in 1), azithromycin was prescribed in 2 patients and 3 remained without therapy. Regarding the follow up at 24±3 months from the 1st evaluation (3 patients were excluded due to too recent follow-up), 4 patients (18.2%) had progressive phenotype, 7 (31.8%) had a favourable evolution and 3 (13.6%) patients had died. During a follow-up of 31.1±19.8 months, this number rose to 6 patients (27.3%), all of them died by respiratory cause and had NSIP pattern. No differences were found in age, last FVC, therapy and time of disease evolution between those who died and the others.

Conclusion: Our study showed that a small proportion of IPAF patients had a progressive phenotype and the NSIP pattern seemed to be a poor prognosis factor for survival.

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FRI0496

FREQUENCY OF POLYAUTOIMMUNITY IN RHEUMATOID ARTHRITIS AND SYSTEMIC LUPUS ERITHEMATOSUS

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Background:

Objectives: To study the frequency of polyautoimmunity and multiple autoimmune syndrome (MAS) in patients with rheumatoid arthritis (RA) and systemic lupus erythematosus (SLE).

Methods: Study design: We performed a cross-sectional study in patients with RA and SLE, and compared them with healthy subjects. Cases: RA

patients classified by ACR/EULAR 2010 criteria and SLE patients classified by ACR/EULAR 2019 criteria. SLE and RA patients were compiled consecutively from a rheumatology clinic of the Regional University hospital of Malaga. Controls: subjects without rheumatologic autoimmune disease (AD) from the same population area. Protocol: All subjects filled out a pre-designed questionnaire for the collection of polyautoimmunity data on the cut-off date. Main variables: polyautoimmunity was defined as co-occurrence of SLE or RA and other AD. Secondary variables: Rheumatologic, cutaneous, endocrine, digestive and neurological AD. MAS was defined as presence of three or more AD. Family history of SLE, RA and other autoimmune diseases were also collected. Statistic analysis: descriptive analysis, bivariate analysis and multivariable analysis were done. (Dependent variable: Polyautoimmunity).

Results: We recruited 109 patients with RA, 105 with SLE and 88 controls. Fifteen patients with RA (13.8%), 43 with SLE (41%) and 2 controls (2.2%) reported polyautoimmunity. Table 1 describes the epidemiological characteristics, comorbidities and polyautoimmunity in study population. The most frequent AD associated with RA was Sjögren's syndrome (SS) (53.3%) and SS (55.8%) followed by the antiphospholipid syndrome (30.2%) were associated with SLE. Hashimoto's thyroiditis and psoriasis were the next most frequent AD. According to family history, 5 patients with RA (33.3%) and 12 with SLE (27.9%) had a family history of first degree of other AD. Obesity was associated with polyautoimmunity in RA (OR = 3.362, p = 0.034). In SLE, joint damage (OR = 2.282, p = 0.038) and anti-RNP antibodies (OR = 5.095, p = 0.028) were factors associated with polyautoimmunity and taking hydroxychloroquine was a protective factor (OR = 0.190, p = 0.004).

Conclusion: Polyautoimmunity in RA and especially in SLE is frequent. It was associated with obesity in RA and in SLE with joint damage and anti-RNP antibodies. The hydroxychloroquine appeared as a protective factor.

Variables	RA (N=109)	SLE (N=105)	Controls (N=88)	P value
Epidemiological characteristics				
Sex: female, n (%)	85 (78.0)	99 (94.3)	68 (77.3)	0.001
Age, mean (SD), years	56.5 (10.8)	50.8 (13.2)	57.1 (10.6)	0.133
Caucasic race, n (%)	107 (98.2)	105 (100)	88 (100)	0.168
Comorbidities				
Smoking				0.001
No smoking, n (%)	55 (50.5)	78 (74.3)	56 (63.6)	
Smoking history, n (%)	54 (49.5)	27 (25.7)	32 (36.4)	
Obesity, n (%)	38 (34.9)	21 (20.0)	22 (25.0)	0.044
Dyslipidemia, n (%)	24 (22.0)	22 (21.2)	17 (19.3)	0.896
Hypertension, n (%)	27 (24.8)	28 (26.7)	23 (26.1)	0.746
Diabetes mellitus, n (%)	7 (6.4)	1 (1.0)	1 (1.1)	0.031
Polyautoimmunity, n (%)	15 (13.8)	43 (41)	2 (2.2)	<0.001
MAS, n (%)	1 (0.9)	9 (8.6)	0 (0.0)	<0.001
FH polyautoimmunity, n (%)	19 (17.6)	26 (24.8)	15 (17.0)	0.060

RA: rheumatoid arthritis; SLE: systemic lupus erythematosus; SD: standard deviation; MAS: multiple autoimmune syndrome; FH:Family history

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FRI0497

THERAPEUTIC STRATEGIES AND LONG-TERM OUTCOME IN PATIENTS WITH INTERSTITIAL PNEUMONIA WITH AUTOIMMUNE FEATURES: A SINGLE CENTER LARGE-SCALE OBSERVATIONAL COHORT STUDY

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Background: Patients with idiopathic interstitial pneumonia (IIP) may have features of connective tissue diseases (CTDs). The term interstitial pneumonia with autoimmune features (IPAF) has been recently proposed for such patients [1]. To date, only few studies have comprehensively described outcomes over a long-term period and choices of treatment [2-4].

Objectives: The aim of this study was to investigate the therapeutic strategies and long-term outcome among patients with IPAF, IIP, and CTD-ILD.

Methods: Six hundreds- and seventy-two patients who had visited our department between April 2009 and March 2019 and were evaluated by chest HRCT scan. They were clinically and radiologically diagnosed as having interstitial lung disease (ILD), including IIP, CTD-ILD, undifferentiated connective tissue diseases associated ILD or other ILD. Then, we applied IPAF criteria to these patients, 68 patients were diagnosed as IPAF. We extracted the