had high risk of bias, small sample size and low quality (Level 2 evidence) with high attrition rate but it revealed reduction of daily prednisolone with Azathioprine. A high quality RCT (Level 1) did not confirm a steroid sparing effect with Infliximab vs placebo, and there was no significant difference between relapse or remission rate. Methotrexate studies showed conflicting results: one high quality RCT (Level 1) and one low quality RCT (Level 2) on Methotrexate revealed statistically significant steroid sparing effect, however the remaining study did not demonstrate between Methotrexate and placebo. Two methotrexate studies assessed the risk of relapse, with conflicting results (relapses 73% placebo vs 47% methotrexate; or no difference).

Methotrexate was not associated with increased adverse effects in any of the studies. Azathioprine was associated with significant adverse events resulting in high attrition.

A meta analysis was not performed for methotrexate as the studies were heterogeneous.

Conclusion: There is a lack of evidence regarding DMARDs and biologics in PMR. Methotrexate is an effective steroid sparing agent, and is not associated with increased adverse events. Azathioprine may be effective but is associated with significant adverse events. Infliximab is not an effective steroid sparing agent in PMR. More high quality RCTs are needed to study the efficacy of steroid sparing agents.

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FRI0476

RHEUMATIC MANIFESTATIONS DURING INFLAMMATORY BOWEL DISEASE

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Background: Rheumatologic manifestations are frequent extraintestinal manifestations (MEI) of chronic inflammatory bowel disease (IBD). Some of these manifestations develop in parallel with the underlying disease, others evolve on their own account. They sometimes lead to reconsider the initial therapy for intestinal purposes.

Objectives: The aim of our study is to specify the epidemioclinical characteristics of osteoarticular manifestations of chronic inflammatory bowel disease and their possible impact on intestinal disease

Methods: This is a retrospective study conducted between January 2000 and December 2015 including patients hospitalized in our department for chronic inflammatory bowel disease (IBD).

Results: During the study period, 206 patients with IBD were hospitalized in our department, 78 of whom had rheumatic MEI (frequency equal to 37.8%). These 78 patients were divided into 48 men and 30 women with a sex ratio of 1.6 and an average age of 40.7 ± 13.6 years (18–79). They had Crohn's disease in 60% of the cases. The average length of service for IBDs was 83 ± 73 months (4–360). Osteoarticular MEIs were peripheral in 56.4% of cases, axial in 29.5% of cases, mixed in 6.41% of cases with the presence of osteopenia in 6.41% of cases and osteoporosis in 1.28%. The activity of IBD associated with these MEI was moderate with an average number of outbreaks / year of 1.6 ± 0.8. These patients were treated with salicylates in 30.7% of the cases using corticosteroid therapy at least once in 23% of the cases. Maintenance treatment based on immunosuppressants was found in 38.46% of cases and anti-TNF alpha in 10.25% of cases. During follow-up, 3 patients died (3.84%). There was no significant difference between patients with IBD with rheumatic MEI and without rheumatic MEI regarding epidemiological data and disease activity.

Conclusion: Rheumatic MEIs are found in almost 40% of IBDs. They affect men more frequently than women and Crohn's disease than UC. They are rather peripheral and do not associate with a more important activity of the disease.

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FRI0477

SYSTEMIC TREATMENT IN SARCOIDOSIS. STUDY OF 377 PATIENTS FROM A SINGLE UNIVERSITY HOSPITAL

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Background: Sarcoidosis is a multisystemic disease characterized by the formation of non-necrotizing granulomas. The most frequently affected organs are lungs, skin and eyes. Systemic corticosteroids are the most used drugs in the treatment of this disease. Conventional and biological immunosuppressants (IS) may also be used (1-3).

TABLE. Systemic treatment of sarcoidosis according to clinical domains.

ORGAN INVOLVEMENT	CONVENTIONAL IS							BIOLOGICAL IS									
	Cases	NT	OCS	MD of OCS (mg/d)*	IVMP	MTX	AZA	CFM	MMF	BT	IFX	ADA	ETN	GLM	TCZ	RTX	CR
Pulmonary, n(%)	319(84.6)	128(40.1)	185(58)	43.6±18.8	13(4.1)	46(14.4)	13(4.1)	2(0.6)	3(0.9)	21(6.6)	12(3.8)	16(5.0)	0	1(0.3)	2(0.6)	3(0.9)	64(33.5)
Cutaneous, n(%)	124(32.9)	50(40.3)	66(53.2)	38.0±20.2	4(3.2)	18(14.5)	4(3.2)	1(0.8)	0	9(7.3)	5(4)	9(7.3)	1(0.8)	2(4.2)	0	1(0.8)	29(39.2)
Ocular, n(%)	48(12.7)	5(10.4)	38(79.2)	41.5±17.6	10(20.8)	23(47.9)	8(16.7)	1(2.1)	1(2.1)	13(27.1)	5(10.4)	11(22.9)	1(2.1)	2(4.2)	2(4.2)	0	10(23.3)
Musculo-eskeletal, n(%)	108(28.6)	41(38)	65(60.2)	39.2±18.7	5(4.6)	26(24.1)	6(5.6)	0	0	12(11.1)	8(7.4)	8(7.4)	1(0.7)	2(1.9)	0	1(0.9)	24(35.8)
Hepatic, n(%)	41(10.9)	12(29.3)	27(65.9)	43.8±15.2	3(7.3)	6(14.6)	1(2.4)	1(2.4)	0	1(2.4)	1(2.4)	1(2.4)	0	0	0	0	6(20.7)
Neurologic, n(%)	27(7.2)	4(14.8)	23(85.2)	50.9±20.2	5(18.5)	14(51.9)	5(18.5)	0	0	9(33.3)	7(25.9)	5(18.5)	1(3.7)	2(7.4)	1(3.7)	1(3.7)	6(26.1)
Cardiac, n(%)	8(2.1)	4(50)	4(50)	40.0±28.3	1(12.5)	2(25)	1(12.5)	1(12.5)	0	2(25)	2(25)	2(25)	0	0	0	0	1(25)
Renal, n(%)	22(5.8)	2(9.1)	20(90.9)	47.1±13.8	1(4.5)	6(27.3)	2(9.1)	0	0	2(9.1)	1(4.5)	2(9.1)	0	0	0	0	3(15)
Löfgren's síndrome, n(%)	44(11.7)	18(40.9)	24(54.5)	29.8±15.3	0	3(6.8)	1(2.3)	0	0	2(4.5)	1(2.3)	2(4.5)	0	0	0	1(2.3)	19(69.2)
Heerfordt's síndrome, n(%)	2(0.5)	0	2(100)	57.5±17.7	2(100)	2(100)	1(50)	0	0	2(100)	2(100)	2(100)	0	0	0	0	0
TOTAL, n(%)	377(100)	161(42.7)	206(54.6)	48.2±19.0	15(4.0)	53(14.1)	16(4.2)	2(0.5)	3(0.8)	28(7.4)	15(4.0)	20(5.3)	1(0.3)	2(0.5)	3(0.8)	3(0.8)	72(33.3)

NT: no treatment; MD: maxim dose; OCS: oral corticosteroids; IVMP: intravenous methylprednisolone; BT: biological therapy, CR: complete response

* Mean (±SD)

Table 1. Characteristics and treatment of Rh-irAEs patients.

	Age, Sex	Cancer	ICI	Onset (months)	Rhem-irAE	Treatment	Response	ICI stop	AutoAb
1	69, M	Lung, adeno	Nivo	3	Myo	CS, MMF, ANK, TCZ	LDA	Y, D	pANCA
2	70, M	Bladder	Atezo	1	A, A,	CS, MTX	Rem	Y, T	Neg
3	79, M	Lung SCC	Pembro	1	A	CS	Rem	No	Ro52
4	52, M	Lung, LCC	Nivo	6	A, CV	CS, MTX, ANK, TCZ	LDA	No	ANA 1:160 s
5	82, M	Lung, LCC	Pembro	3	PMR	CS	Rem	No	Neg
6	56, M	Melanoma	Pembro	26	PMR, PM	CS, MTX	Death	Y, D	ANA 1:160 s, pANCA
7	72, M	Lung, adeno	Pembro	5	A, CV	CS, MTX, TCZ	HDA (TCZ)	Y, T	ANA 1:160 s
8	74, M	Lung, adeno	Pembro	1	PMR	CS, MTX	LDA	No	ANA 1:160 s
9	75, M	Lung, adeno	Pembro	1	PM, Myo	CS	Rem	Y, D	Neg
10	75, M	Lung, adeno	Nivo	9	A	CS	Rem	No	Neg
11	68, F	Lung, adeno	Atezo	18	A	CS, MTX	LDA	Y, T	Neg
12	35, M	Melanoma	Nivo	2	A	CS, MTX	LDA	No	ANA 1:640 s
13	79, M	Lung, SCC	Nivo	15	A, PMR	CS, MTX, ANK	Death (NR)	Y, D	Ro52
14	77, M	Lung, adeno	Pembro	8	DM	CS, HCQ, IVIG	LDA	Y, D	ANA 1:320 PCNA
15	67, M	Lung, adeno	Nivo	13	A	CS, MTX	LDA	No	ASMA
16	74, M	Lung, SCC	Atezo	1	A, CV	CS, HCQ, Colch	LDA	Y, D	Neg
17	70, M	Lung, adeno	Nivo	5	A, CV	CS	Rem	Y, D	Neg
18	70, M	Lung, adeno	Pembro	15	A	CS	LDA	No	ASMA
19	69, M	Lung, adeno	Nivo	5	DM	CS, HCQ, MMF, IVIG, Tacro	MDA	Y, T	ANA 1:160 s
20	84, M	H&N, SCC	Nivo	3	Ph	CS	Rem	Y, D	Neg
21	67, M	Lung, adeno	Durva	5	A, PM, Myo	CS	LDA	Y, D	ANA 1:160 h
22	50, F	Lung, LCC	Nivo	1	A, CV	CS, HCQ	Rem	Y, D	Neg

Objectives: To evaluate the systemic treatment of sarcoidosis according to clinical domains.

Methods: Study of all consecutive patients diagnosed with sarcoidosis between 1/1/1999 and 1/1/2019 in a tertiary university hospital. The diagnosis was established following the ATS/ERS/WASOG criteria (*Eur Respir J*. 1999;14(4):735-737): compatible clinical and radiological presentation, evidence of non-caseifying granulomas and exclusion of other granulomatous diseases.

Results: We studied 377 patients (188 men/189 women), mean age at diagnosis of 46.0±14.8 years. After a mean follow-up of 13.0±9.3 years, 161 (42.7%) patients did not require treatment. The remaining 216 (57.3%) received oral glucocorticoids (206, 54.6%) with a maximum mean dose of 43.2±19.0 mg/day, conventional IS (60, 16.2%), biological therapy (28, 7.4%) and/or endovenous methylprednisolone (15, 4.0%). Biological therapy was indicated by pulmonary (9, 32.1%), ocular (9, 32.1%), neurological (3, 10.7%), musculoskeletal (3, 10.7%), cutaneous (2, 7.1%), nephrological involvement (1, 3.6%); and Heerfordt's syndrome (1, 3.6%). Adalimumab and Infliximab were the biologics used more frequently (Table).

Conclusion: Compared to other studies, the high percentage of patients who required systemic treatment is remarkable. It also highlights the frequency of the use of biological drugs in more severe organ involvement (ocular and neurological), which is consistent with the trend in recent years.

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FRI0478

SEROLOGICAL AUTOIMMUNITY IN PATIENTS WITH RHEUMATIC IMMUNE-RELATED ADVERSE EVENTS: CORRELATION WITH SEVERITY AND TREATMENT

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Background: Immune checkpoint inhibitors (ICIs) can induce a variety of rheumatic immune-related adverse events (Rh-irAEs). Few data are available on which features can predict the occurrence of long-lasting and severe Rh-irAE

Objectives: To describe the serological features associated with long-lasting and severe Rh-irAE.

Methods: ICI-treated patients were identified. Patients' demographics, histotype of cancer, ICI, time interval from ICI start to Rh-irAE onset, characteristics of Rh-irAEs were recorded. Patients were tested for autoimmunity panel (a-IP): RF, ACPA, ANA, anti-SSA, anti-SSB, anti-Sm, anti-RNP, anti-Jo1, ANCA, ASMA, AMA, anti-dsDNA, anti-tireoglobulin, anti-tireoperoxidas, IgM and IgG anti-cardiolipin and anti-β2 glycoprotein I, crioglobulins. All patients were treated with steroids (CS). In case of flare of the Rh-irAE, csDMARDs or bDMARDs were started. Associations between a-IP status and need for DMARD start was evaluated. Non parametric tests were used.

Results: 22 Rh-irAE were included (see Table 1). Median age at Rh-irAE onset was 70 (50 – 84) years. 2 patients (9%) had a personal history of psoriasis. Median time from ICI start to Rh-irAE onset was 5 (1 – 26) months. 11 patients (50%) developed 1 Rh-irAE, 10 (45.4%) 2 and 1 (4.5%) 3. The most frequent were arthritis (A, 14, 63.6%), cutaneous vasculitis (CV, 5, 22.7%), PMR-like (4, 18.2%), polymyositis (PM, 4, 18.2%), myocarditis (Myo, 3, 13.6%) and dermatomyositis (DM, 2, 9.1%). Median initial prednisone dose was 25 (10 – 75) mg daily. In 14 patients (63.6%) a csDMARD was started upon steroids tapering. 9 patients (41%) were treated with methotrexate (MTX, 4, 18.2%) with hydroxychloroquine (HCQ, 2, 9.1%) with mycophenolate (MMF, 2, 9.1%) with colchicine (colch). 6 patients were treated with bDMARDs. 3 patients (50%) were treated with anakinra (ANK), 2 (33.3%) with IVIG and 3 (50%) with tocilizumab (TCZ). 13 patients (59.1%) were a-IP+. A significantly higher percentage of a-IP+ patients received DMARDs (11, 84.6%) compared to a-IP- patients (2, 22.2%, p = 0.0007). A significantly higher percentage of a-IP+ patients were treated with bDMARDs (5, 38.5%) compared to a-IP- patients (0, 0%, p = 0.05). We analysed whether in csDMARD-treated patients the need for bDMARDs was higher in a-IP+ but we found no statistical significance (45.4% vs 0%, p = 0.487).

Conclusion: The presence of serological autoimmunity might be helpful in detecting patients with Rh-irAEs refractory to steroid therapy.

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FRI0479

REWired CELL ENERGY METABOLISM IN A NOVEL KRAS-MUTATED HISTIOCYTOSIS CHARACTERIZED BY SEVERE SYNOVITIS

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