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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