converting enzyme (ACE), liver enzyme test, chest radiography, chest computed tomography scan, treatment and biopsy if performed.

TABLE. Demographic and clinical features of the 50 patients with ocular sarcoidosis.

	Patients (n=50)
Age, mean (SD) years	45.5 (16.7)
Sex, men/women, n/n	23/27
Affected organs, n (%)	
Lung	30 (60)
Skin	14 (28)
Liver	4 (8)
Kidney	3 (6)
Musculoskeletal system	3 (6)
Central nervous system	3 (6)
Hearth	2 (4)
Ocular signs, n (%)	
Bilaterality	22 (44)
Snowballs/strings of pearls	19 (38)
Mutton-fat KPs	12 (24)
Multiple chorioretinal peripheral lesions	7 (14)
Periphelebitis	5 (10)
Anterior synechiae	1 (2)
Optic disc granulomas	1 (2)
IWOS criteria, n (%)	
Definite	22 (44)
Presumed	13 (26)
Possible	4 (8)
Treatment, %	
Prednisone (oral)	44 (88)
Methylprednisolone (bolus)	5 (10)
Conventional IS	
.....MTX	21 (42)
.....AZA	9 (18)
.....CsA	2 (4)
.....MMF	2 (4)
Biologic therapy	
.....IFX	5 (10)
.....ADA	4 (8)
.....GLM	2 (4)

Abbreviations: ADA, adalimumab; AZA, azathioprine; CsA, cyclosporine A; GLM, golimumab; IFX, infliximab; IS, immunosuppressor; KPs, keratic precipitates; MMF, mycophenolate mofetil; MTX, methotrexate; SD: standard deviation.

Results: We selected patients with ocular inflammation from a cohort of 381 patients with sarcoidosis (n=50, 13%). Most of the cases were women (54%) and median age was 45.5±16.7 years. In these 50 cases, the most affected organ was lung (60%), followed by skin (28%). Forty patients had uveitis, 32 of them with ocular symptoms. Thirty-nine out of 50 patients (78%) met one of the 4 IWOS diagnostic categories: 22 with definite (44%), 13 presumed (26%) and 4 with possible (8%) sarcoidosis. Eleven patients did not meet IWOS criteria. The most common ocular signs were bilaterality (44%), snowballs or strings of pearls (38%), mutton-fat KPs (24%), multiple chorioretinal peripheral lesions (14%) and periphelebitis (10%). The median value of ACE was 69 U/l. Forty-four patients (88%) received oral corticosteroids, 21 (42%) received methotrexate, 11 (21%) received another conventional immunosuppressor and 11 (21%) a biological treatment. TABLE shows demographic and clinical features.

Conclusion: In our population the IWOS Criteria had a sensitivity of 78%. Even though there is no gold standard for diagnosing ocular sarcoidosis yet, IWOS signs can help clinicians suspect it.

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THU0567

POLYMYALGIA RHEUMATICA-LIKE SYNDROME FROM CHECKPOINT INHIBITOR THERAPY: CASE SERIES AND SYSTEMATIC REVIEW OF THE LITERATURE

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Background: Checkpoint inhibitor therapy has caused a paradigm shift in the field of oncology, producing significant survival benefits in patients with an ever-growing list of malignancies, but their use is attended by a spectrum of immune related adverse events (irAEs). Many rheumatic irAEs have been described. A critical and presently unanswered question is what proportion of these rheumatic irAEs represent new appearances of classic rheumatic diseases or, alternatively, represent new clinical variants with potentially different pathogenesis, clinical course and treatment responsiveness. Little is known about the PMR-like entity that has been described in the setting of ICI therapy.

Objectives: To assess whether the polymyalgia rheumatica (PMR)-like syndrome reported as an irAE from checkpoint inhibitor therapy is consistent with the 2012 EULAR/ACR provisional criteria for PMR¹.

Methods: The cases were derived from two sources. Group 1 represents reported cases from three contributing centers. Group 2 was derived from a systematic review of the literature searching for all cases reported as PMR or PMR-like illness associated with checkpoint inhibitor therapy. Cases were assessed for the quality of reporting and then analyzed to determine whether they fulfilled the 2012 EULAR/ACR provisional criteria for PMR.

Results: A total of 49 patients were included for analysis, including 9 cases from Cleveland Clinic (Table 1), 4 cases from Johns Hopkins University, and 7 cases from University Hospital of Bordeaux, in addition to the 29 separate cases found by systematic review. Among the entire group, 37 (75%) were designated "complete" indicating that they had sufficient data to reliably apply the 2012 EULAR/ACR criteria of which 28 (75%) cases fulfilled complete criteria for PMR. A number of cases also demonstrated some clinical features unusual for idiopathic PMR.

Conclusion: This study suggests a high proportion of reported cases of checkpoint inhibitor-related PMR fulfill preliminary criteria for PMR yet in one quarter clinical details were incomplete making verification problematic. Furthermore, in the absence of a gold standard for the diagnosis of PMR, the relationship of checkpoint inhibitor-related PMR to the idiopathic form remains unclear.

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THU0568

CLINICAL CHARACTERISTICS OF SEVEN RELAPSING POLYCHONDROITIS CASES MISDIAGNOSED AS ARTHRITIS

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Background: Relapsing polychondritis (RPC) is a rare autoimmune disease. It mainly affects the tissue abundant of cartilage, such as ear, nose, and tracheoles. The multiple systems including eye, joint, mucocutaneous, and kidney could be involved¹. As the presentation of arthritis was nonspecific and could appear in several connective tissue diseases,

it was easily misdiagnosed. The delay in establishing the diagnosis of RPC was common. A survey from the United Kingdom found a median length of 1.9 years from the first disease attack to diagnosis². One recent research reported 64% of RPC patients had a diagnostic delay with more than five years³.

Objectives: We made a retrospective study to explore distinct characteristics of relapsing polychondritis with arthritis as the first attack. By comprehending the nature of disease fully, misdiagnosis at the early phase of illness could be avoided.

Methods: The clinical features and prognosis of 7 RPC patients in Peking Union Medical College Hospital between October 2012 and October 2018, presenting as arthritis at onset, were retrospectively analyzed.

Results: There were five female patients. The female to male ratio of the case series was 5:2. The age of 7 patients was 43.43 ± 9.86 years. The number of affected joints was 20.14 ± 10.92 . The joint involvement was most common in the bilateral proximal interphalangeal joints (PIPs) and interphalangeal joints of the thumb. Recurrent arthritis occurred in 5 patients. There was no accompanying enthesitis. The specific serum indexes of RA, such as rheumatoid factor (RF), anti-cyclic citrullinated peptide (CCP) antibody, were negative. There were three patients administered with conventional synthetic disease modifying anti-rheumatic drugs firstly, and they did not respond well. The median delay time from onset to diagnosis of RPC was ten month (Figure 1). The deformity and destruction of the joint were not observed in all patients, by clinical and radiological assessment. The average duration from arthritis to the occurrence of other involved system was 8.3 months. Four patients were misdiagnosed as rheumatoid arthritis at first. One patient was considered as ankylosing spondylitis at first. Two patients were diagnosed as arthritis at the beginning. As regards to treatment, glucocorticoids (GCs), cyclophosphamide and mycophenolate mofetil were used. Two patient was complicated with palmoplantar pustulosis and Kikuchi-Fujimoto disease. During follow up, myelodysplastic syndrome occurred in one patient. After following up for 35.43 ± 30.92 months, all patients survived and were not with the recurrence of arthritis.

Figure 1. The demonstration of multiple system involvement and the interval from onset to diagnosis in seven patients with relapsing polychondritis.

Conclusion: RPC patients with arthritis onset were characteristic of middle-aged women and symmetric multiple joint involvements. The RF and anti-CCP antibody were negative. The arthritis was recurrent and not erosive. The interphalangeal joints of the thumb and PIPs were the most frequently involved joints. In clinical practice, physicians should be alert to the possibility of RPC, when encountered the patients with abovementioned arthritis. The inquiry about manifestations of other system involvement in RPC and a regular follow-up was warranted. After the combined therapy of GCs and immunosuppressants, all patients had a favorable prognosis.

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THU0569

MANAGEMENT OF ADULT-ONSET STILL'S DISEASE (AOSD) WITH IL-1 INHIBITORS: EVIDENCE- AND CONSENSUS-BASED STATEMENTS BY A PANEL OF ITALIAN EXPERTS

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Background: Still's disease is a rare autoinflammatory disease, presenting in both pediatric [systemic juvenile idiopathic arthritis (SJIA)] and adult patients [adult-onset Still's disease (AOSD)]. Due to the rarity of the disease, clinical trials are limited and treatment guidelines are not available. In patients refractory to the classical therapy with NSAIDs, corticosteroids and DMARDs, the introduction of drugs targeting IL-1 has greatly expanded treatment options. Among these, canakinumab, a human monoclonal anti-IL-1 β antibody, and anakinra, a human recombinant IL-1RA, have been recently approved for the treatment of refractory patients.

Objectives: To produce recommendations, based on evidence and expert consensus, that can help clinicians in choosing the most appropriate treatment of AOSD, with particular attention to anti-IL-1 therapies, in order to achieve disease remission before the development of complications.

Methods: The recommendations development process took place from April to October 2018 and consisted of three steps. The first step was dedicated to a comprehensive literature review and development of statements. Two separate literature searches were performed: a) similarities and differences between SJIA and AOSD; b) efficacy and safety of IL-1 blockade in AOSD. The issue related to the treatment of AOSD with anti-IL1 therapies was specified into 4 questions: 1) efficacy and safety; 2) comparison among IL-1 inhibitors; 3) early versus late treatment; 4) systemic versus chronic articular pattern of the disease. In the second step, the statements were submitted in a Delphi process to a panel of 67 rheumatologists. Consensus threshold was set at 66%: positive, > 66% of voters selected scores 3 to 5; negative, > 66% of voters selected scores 1 or 2. At the third step of the consensus process, the voting results were analyzed, and the statements were finalized.

Results: In the two literature searches, 332 and 358 publications were identified; 30 and 25 publications, respectively, were selected according to the inclusion criteria. Based on the review of the literature and personal clinical experience, 11 statements were developed. 48/67 rheumatologists (72%) participated to the Delphi process. Positive consensus was reached after the first round of voting and was full (> 95%) on the majority of statements. A large consensus was achieved in considering AOSD and SJIA as the same disease. The use of anti-IL-1 therapies in refractory patients was considered quite safe and effective both as first and as subsequent line of biologic treatment, especially in systemic patients. Because of the lack of head-to-head comparisons, a different profile of efficacy among IL-1 inhibitors could not be established. There was a large consensus that failure of the first IL-1 inhibitor does not preclude a therapeutic response with another one. The lack of studies comparing early versus late treatment in AOSD patients did not allow to draw conclusions, however data from SJIA suggest a better response in early treated patients.

Conclusion: The Delphi method was used to develop recommendations that we hope will help clinicians in the management of patients with AOSD refractory to conventional therapies.

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