Conclusion: We report a rare case of synovial metastasis concomitant to a lung adenocarcinoma. It is important to make a rapid diagnosis as early recognition of malignant infiltration into joints ensure appropriate multidisciplinary management decisions.

Disclosure of Interests: None declared

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AB0771

HIGH DOSE INTRAVENOUS METHYLPREDNISOLONE INDUCES RAPID IMPROVEMENT OF VISUAL ACUITY IN NON-INFECTIOUS UVEITIS OF DIFFERENT IMMUNE MEDIATED INFLAMMATORY DISEASES

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Background: Rapid and effective remission-inducing therapy is mandatory in uveitis to avoid irreversible structural and functional damage. In some severe cases biological agents might be required (1-6).

High-dose intravenous methylprednisolone (IVMP) may achieve prompt control of inflammation in most immune mediated inflammatory diseases (IMID), including non-infectious uveitis (NIU).

Objectives: To evaluate the efficacy and safety of IVMP pulse therapy in NIU of different IMID.

Methods: Multicentre study of 71 patients with severe uveitis who received IVMP. The underlying diseases were: Vogt Koyanagi Harada disease (VKHD) (n=24), Behçet disease (BD) (19), Sarcoidosis (5) and idiopathic NIU (23). The main outcome variable was Best-Corrected Visual Acuity (BCVA) estimated using the Snellen chart. BCVA that was assessed at 0 (basal), 2-5, 7, 15 and 30 days after IVMP.

The results are expressed as mean $\pm$ SD in normally distributed variables, or as median [IQR] when are not. Comparison of continuous variables was performed using the Wilcoxon test.

Results: We studied 46♀/25♂ patients. The main features are shown in Table 1. IVMP dose ranged from 250 to 1000mg/day administered for 3-5 consecutive days, the dose was established according to the presence or not of other systemic manifestations apart from uveitis. All of them had active intraocular inflammation at the moment of the study. BCVA values improved considerably after 1 month (Figure 1). No major side effects were observed.

Figure 1. Improvement of best corrected visual acuity (BCVA).

Conclusion: High-dose IVMP pulse therapy is useful and safe for a prompt control of BCVA regardless of the underlying IMID.

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Table 1. Main features of 71 patients with NIU. Data are of affected eyes.

	VKHD (n=24)	Idiopathic (n=23)	Behçet's disease (n=19)	Sarcoidosis (n=5)	Overall (n=71)
Men/Women, n	5/19	9/14	9/10	2/3	71
Mean age (years) $\pm$ SD	42 $\pm$ 11	47 $\pm$ 15	33 $\pm$ 10	42 $\pm$ 22	-
Unilateral/Bilateral	2 (8.3)/22(91.7)	10 (43.5)/13(56.5)	4 (21)/15 (79)	3(60)/2(40)	19/52
NIU, n (%)					
NIU patterns, n (%)					
Posterior uveitis	6 (25)	9 (39.1)	3 (15.8)	1 (20)	19
Panuveitis	18 (75)	14 (60.9)	16 (84.2)	4 (80)	52
Laboratory data, n (%)					
ANA	2 (8.34)	2 (8.7)	0 (0)	1 (20)	5
HLA B27	0 (0)	4 (17.4)	0 (0)	0 (0)	4
HLA B29	0 (0)	1 (4.3)	0 (0)	0 (0)	1
HLA B51	0 (0)	5 (21.7)	8 (42)	3 (60)	16
Angiotensin	1 (4.17)	2 (8.7)	0 (0)	1 (20)	4
Converting Enzyme (ACE)					

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AB0772

JAK INHIBITORS IN REFRACTORY ADULT AND CHILDHOOD ONSET STILL'S DISEASE

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Background: Excessive and inappropriate production of pro-inflammatory cytokines such as interleukin (IL)-1, IL-6 or IL-18, is a pathogenic cornerstone in adult and childhood onset Still's disease. Beyond therapies targeting IL-1 or IL-6, Janus kinases (JAK) inhibitors have been proposed for adult-onset Still's disease (AOSD) patients refractory to or intolerant of treatment with biologicals. Recently, it has been suggested that JAK inhibitors might be efficient in refractory AOSD patients¹.

Objectives: To assess the efficacy and safety of JAK inhibitors in the treatment of refractory systemic juvenile idiopathic arthritis (sJIA) or AOSD.

Methods: This retrospective study was based on a national survey of the departments of rheumatology, paediatric rheumatology and internal medicine in all French hospitals from an online call of the "Club Rhumatismes et Inflammation" (www.cri-net.com). The data were collected using a standardized questionnaire, and analyzed at different time points (treatment initiation, M1, M3, M6 and end of the follow-up). The response to JAK inhibitors was categorized as: complete remission (resolution of all clinical and biologic signs), partial remission (clinical improvement with persistence of a few symptoms) or failure (lack of clinical or biological improvement).

Results: 6 patients (5 adults and 1 child) were recruited (Table 1). Mean age at treatment start was 39.6 years for the AOSD patient and 6 years for the sJIA patient, and mean disease duration was 5.3 years. The clinical expression was predominantly systemic in 5 five patients and chronic articular in one. Response to corticosteroids, conventional synthetic or biological Disease Modifying Anti-Rheumatic Drugs had been considered inadequate in all patients. Baricitinib was used in 3 patients, ruxolitinib in 2, and tofacitinib in 1. Steroids were concurrently used in all patients, anakinra in one, methotrexate and anakinra in one. At a mean (SD) follow-up of 9.5 months, partial response was observed in 4 (66.7%) cases (patients with ruxolitinib, tofacitinib

Table 1. Characteristics of the AOSD patient

No.	Sex	Age (year)	Main symptoms	Treatments before JAKi onset	JAK inhibitors	Steroids at onset (mg/day)	Concomitant treatment	Response at last F-U	Steroids at the end of F-U (mg/day)	F-U (months)
1	F	6	Fever, polyarthrititis, rash	AINS, ANAKI, TOCI, CANAKI, ADA, THALI, INFlix	RUXOLITINIB 5mgx2/day	3	0	P	1	23
2	M	28	Fever, polyarthrititis, rash	ANAKI	BARICITINIB 4mg/day	80	0	N	10	1
3	M	32	Fever, polyarthrititis, rash	TOCI+MTX, ANAKI+MTX, CANAKI+MTX, ADA, CICLO, IgIV	BARICITINIB 4mg/day	16	MTX 20mg/week ANAKINRA 100mg/day	P	12	19
4	F	40	Fever, polyarthrititis, rash	MTX, IMUREL, CICLO, ETANERCEPT, ANAKI+MTX, TOCI+MTX, IgIV	RUXOLITINIB 15mgx2/day	60	ANAKI 200mg/day	P	30	4
5	F	48	Fever, polyarthrititis, rash	TOCI, ANAKI, CICLO, CANAKI, IMUREL	TOFACITINIB 5mgx2/day	50	0	P	7.5	9
6	F	50	Fever, polyarthralgia, rash	ANAKI	BARICITINIB 4mg/day	60	0	N	40	1

F-U: Follow-up; N: No response; P: Partial response

or baricitinib) and failure in 2 (33.3%) (patients with baricitinib). No patient achieved complete remission. At the last visit, steroids could be decreased but not stopped in all patients. Patients with partial response had an average decrease of 72.8% (90% for tofacitinib, 70% for baricitinib, 58.5% for ruxolitinib between the start and the follow-up end date) and non-responder patients were yet able to reduce steroids by 60.5% (Table 1). Tolerance of JAK inhibitors was excellent, however patient 4 experienced an episode of infectious pulmonary disease.

Conclusion: JAK inhibitors therapy may be helpful for some patients with refractory Still's disease. However, no complete response was observed in this short series of cases. There might be a difference of response between the molecules, although the number of patients is too low to draw conclusions. Additional information is thus needed to evaluate more precisely the risk-benefit ratio of this treatment, and a possible difference in efficacy among the different groups of JAK inhibitors.

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AB0773 PATTERN OF MACROPHAGE ACTIVATION SYNDROME IN RHEUMATIC DISEASES

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Background: Macrophage activation syndrome (MAS) is a rare, potentially fatal condition complicating autoimmune diseases, as their initial manifestation or during its course.

Objectives: To describe the pattern of MAS and its salient features in patients with rheumatic diseases.

Methods: A retrospective observational study was conducted in 16 patients diagnosed with MAS, due to rheumatic diseases in Department of Rheumatology, SRIHER, Chennai from January 2018–December 2020.

Results: The primary diseases were Systemic Lupus Erythematosus (SLE) in 75% (n=12), Adult Onset Still Disease (AOSD) in 18.7% (n=3) and Kikuchi Disease in 6.25% (n=1). 50% (n=8) presented with MAS as the initial manifestation. Others had a known rheumatic disease with an average duration of 6.5 years. Occult MAS was seen in 25% (n=4). The most frequent clinical features were unremitting fever (n=15) and splenomegaly (n=15). All had hyperferritinemia—mean of 2971.10 ng/mL (531–12536). Sub group analysis showed significant hypofibrinogenemia (129.8 vs 200.62 mg/dl) in patients presenting with MAS as their initial manifestation. Transaminitis (666/252.5 vs 243/217.62 U/L), hypertriglyceridemia (316 vs 275.8 mg/dl), high LDH (2490 vs 938.75) were more severe in this group, but not significant. All with SLE had increased disease activity, with 50% (n=8) having SLEDAI 2K of ≥15. Bone marrow aspiration showed haemophagocytes in 81.25% (n=13). H score of >169 was seen in 93.75% (n=15). Infection was the commonest trigger for MAS (n=9)—bacterial (n=5), viral (n=4), fungal (n=1) and mycobacterial (n=2)—followed by parturition (n=1). All patients were managed with pulse steroids followed by intravenous cyclophosphamide (n=8), rituximab (n=2), mycophenolate mofetil (n=2) in SLE, tocilizumab (n=2) and etoposide (n=1) in AOSD. Mortality rate was 18.75% (n=3).

Conclusion: Spectrum of MAS in rheumatic diseases could be occult, associated with infection, high disease activity, or lead to mortality. High index of clinical suspicion is required, due to overlap of features like fever, cytopenias and organomegaly in MAS & SLE. Hyperferritinemia, high LDH, & transaminitis could aid in differentiating & early diagnosis. New criteria for rheumatic diseases, including these parameters are needed, especially in resource-limited settings where sIL-2Rα & NK cell activity tests are not feasible.

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