Methods: CMR findings in 40 patients (mean age 45.43 ± 14.7 years; male:female ratio 1:1) diagnosed with endomyocardial biopsy-proven VNLM at our Centre from January 2015 to December 2017, were retrospectively evaluated. CMR was performed at time of clinical presentation and before EMB. At contrast-enhanced CMR we analysed: ventricular function, T2-weighted sequences, short inversion time inversion recovery sequences (STIR), early (EGE) and delayed gadolinium enhancement (LGE). Demographic, clinical and histological data were correlated with CRM findings.

Results: Mean left ventricular (LV) ejection fraction (EF) was $52.5\% \pm 10.8\%$. 50% of patients had reduced LVEF ($<55\%$). Mean right ventricle EF was $55.25\% \pm 7.2\%$. Akinetic or hypokinetic areas were detected in 3 (7.5%) and 17 (42.5%) patients, respectively. LLC were identified in 39 patients (92.5%). LGE was the most common CMR finding. 20 patients (50%) met one LLC, 16 patients (40%) 2 LLCs. 1 patient met all 3 LLCs. STIR abnormalities were identified in the LV lateral wall in 8 (19%), septal wall in 9 (21.4%), LV inferior wall in 9 (21.4%), in the apex in 3 (7.1%), LV posterior wall in 3 (7.1%) and LV anterior wall in 2 (4.8%) patients. Myocardial oedema on T2 images was detectable in 21 (52.5%), EGE in 1 (2.5%) and LGE in 36 (90%) patients. LGE was intramural in 22 (55%), subepicardial in 23 (57.5%) and subendocardial in 4 (10%) patients. Multiple LGE patterns were disclosed in 10 (25%) patients. Pericardial effusion was detected in 9 patients (22.5%). 2 patients did not meet any LLC at CMR. When comparing CMR findings with Holter-ECG tape, clinical presentation, histology, biochemistry and autoimmunity results we found CRM oedema positively correlated with the detection of active myocarditis on EMB (11 vs 4, $p=0.027$). Serum anti-heart antibodies positively correlated with the detection of pericarditis at CMR (3 vs 1, $p=0.050$). Patchy/intramural LGE positively correlated with rhythm abnormalities at Holter-ECG tape (12 vs 6, $p=0.037$).

Conclusions: VNLM is an overlooked disease defined by EMB established histological, immunological and immune-histochemical criteria. Although the diagnostic power of CMR needs to be further investigated, it seems to correlate with EMB results and arrhythmic burden.

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AB1136 CLINICAL FEATURES OF ADULT CHINESE PATIENTS WITH PERIODIC FEVER, APHTHOUS STOMATITIS, PHARYNGITIS, AND ADENITIS: REPORT FROM AN SINGLE CENTRE

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Background: Periodic fever, aphthous stomatitis, pharyngitis, and cervical adenopathy (PFAPA) syndrome is a polygenic or multifactorial autoinflammatory disease (AUIDs) with unknown etiology. It mainly affects children, though recently it has been also recognised in adulthood. PFAPA syndrome has not been reported in the Chinese population up till now.

Objectives: We aimed to investigate the clinical features, treatment and prognosis of the first cohort of adult Chinese patients with PFAPA syndrome.

Methods: We prospectively evaluated clinical data of adult patients (≥ 14 years) with recurrent fever of unknown origin who were suspected monogenic AUIDs in the period April 2015 to December 2017, at the adult AUIDs centre, Department of Rheumatology, Peking Union Medical College Hospital. Gene tests of AUIDs associated pathogenic genes were examined to exclude monogenic AUIDs. The clinical features were evaluated for patients with PFAPA syndrome, and were compared with a newly reported large Italian cohort of adult patients with PFAPA syndrome.¹

Results: During the study period, a total of 7 adult patients with PFAPA syndrome were diagnosed (table 1), and the male to female ratio was 6:1. All these patients had a disease onset in adulthood, and the mean age of onset was 27 ± 10 years, and the mean time of delay of diagnosis was 4.9 ± 4.0 years. Clockwork periodicity fever was the main feature of PFAPA syndrome, and the median duration of attacks was 3.5 days, and the median interval between attacks was 4 weeks. Pharyngitis, adenitis and aphthous stomatitis were present in 85.7%, 57.1% and 42.9%, respectively. Other common symptoms included fatigue (100%), headache (71.4%), and myalgia (71.4%). The presence of arthralgia/arthritis was less frequent compared to the Italian cohort ($p=0.012$) (table 2). Corticosteroids given during attacks were effective in 3 patients. NSAIDs were partially effective but tonsillectomy was of no effect. Colchicine was rarely used compared to the Italian cohort ($p=0.046$).

Abstract AB1136 – Table 1. Summary of the clinical characteristics of our patients.

No. of patients	7
Male	6 (85.7%)
Female	1 (14.3%)
Age at onset (years)	27 ± 10 (16-46)
Age at diagnosis (years)	31 ± 11 (17-53)
Delay in diagnosis (years)	4.9 ± 4.0 (1.0-11.0)
Adult onset	7
Positive family history	0
Duration of febrile attacks (days)	3.5 (0.5-7.0)
Duration of attack intervals (days)	4 (1-12)
Frequency of attacks per year	10 (4-20)
Peak temperature (°C)	39.2 ± 0.6 (38.5-40)
Periodic Fever with 3 major clinical features	2 (28.6%)
Periodic Fever with only 2 major clinical features	2 (28.6%)
Periodic Fever with only 1 major clinical features	3 (42.9%)

Clinical features	Ours (n, %) (n=7)	Italian (n, %) (n=30)	p value
Pharyngitis	6 (85.7)	23 (76.7)	0.601
Cervical adenopathy	4 (57.1)	22 (73.3)	0.399
Aphthous stomatitis	3 (42.9)	15 (50)	0.734
Fatigue	7 (100)	19 (63.3)	0.056
Headache	5 (71.4)	13 (43.3)	0.181
Myalgia	5 (71.4)	16 (53.3)	0.384
Conjunctivitis	2 (28.6)	4 (13.3)	0.325
Arthralgia/arthritis	1 (14.3)	20 (66.7)	0.012
Nausea and vomiting	1 (14.3)	5 (16.7)	0.878
Chest pain	0 (0)	6 (20)	0.196
abdominal pain/diarrhea	0 (0)	8 (26.7)	0.123
Periorbital edema	0 (0)	5 (16.7)	0.245
Rash	0 (0)	5 (16.7)	0.245
Treatment			
Glucocorticoid	3/3 (100)	15/17 (88.2)	0.531
NSAIDs	1/3 (33.3)	10/12 (83.3)	0.080
Colchicine	0/1 (0)	3/3 (100)	0.046
Tonsillectomy	0/2 (0)	1/3 (33.3)	0.361

Abstract AB1136 – Table 2. Comparison of with an Italian cohort of adult patients with PFAPA syndrome

Conclusions: PFAPA syndrome is one of the main causes of adults Chinese patients with recurrent fever of unknown origin and might be misdiagnosed. Clinicians, not only paediatricians, should take into account this clinical entity, especially for patients with pharyngitis, cervical adenopathy and aphthous stomatitis.

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AB1137 INFLAMMATORY AND EROSIVE ARTHRITIS, A COMMON UNKNOWN MANIFESTATION OF VOGT-KOYANAGI-HARADA DISEASE (VKHD)

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Background: VKHD is a rare cause of bilateral granulomatous panuveitis leading to significant visual loss in patients who present it. VKHD is a multisystemic, autoimmune, inflammatory disease that usually displays extraocular manifestations that include meningismus, vitiligo, poliosis, and hearing loss. Arthritis is an unknown manifestation of the disease. In Mexico, VKHD affect mainly young women and is responsible for 13% to 27% of all uveitis.

Objectives: To describe the presence of inflammatory and erosive arthritis in patients with VKHD.

Methods: Observational study of patients with established VKHD that fulfilled the 2001 revised diagnostic criteria for VKHD was rolled out. Articular ultrasonography was performed to all patients by a trained rheumatologist in carpal, metacarpophalangeal (MCP) and proximal interphalangeal (PIP) joints evaluating

presence of sinovitis, power Doppler (PD) signal, bone erosions, and cartilage changes; serology and hand x-ray were requested and evaluated.

Results: We included 5 women and 1 man, mean age was 36.2 ± 10.3 years, and they all had characteristics of acute (4 patients) or chronic (2 patients) VKHD. They received treatment with intravenous methylprednisolone 1 g/day for 3 days combined with cyclophosphamide and thereafter methotrexate (12.5–17.5 mg/week) for maintenance therapy, scheme with which no ocular recurrences have been presented. Two patients had synovitis on physical examination. All patients had PD signal on the ultrasound and they had involvement of both wrists, MCPs and PIPs joints consistent with polyarthritis pattern; additionally bone erosions were detected in 2 patients. Rheumatoid factor and anti citrullinated peptide antibody were negative in all patients

Abstract AB1137 – Figure 1. a) Ocular ultrasonography of patient 1, showing bilateral exudative retinal detachment b) Articular ultrasound of patient 1, showing synovitis with PD signal in both wrists. c) Fundus photographs of the right and left eyes of patient 2 showing exudative retinal detachments d) Articular ultrasound of patient 2, showing synovitis with PD signal in both wrists.

Conclusions: The presence of inflammatory and erosive arthritis in patients with VKHD has only been described in one case recently. We propose that polyarthritis is common manifestation of this rare disease, including erosive arthritis, representing part of the spectrum of the disease, processes that share some features of the genetic susceptibility with rheumatoid arthritis as HLA-DR4, CTLA-4 and STAT4.

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AB1138

HIGH LEVELS OF ANTI-U1RNP AND ANTI-SM IN MIXED CONNECTIVE TISSUE DISEASE PATIENTS

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Background: The mixed connective tissue disease (MCTD) is an autoimmune systemic disease characterised by clinical manifestations that are included in systemic lupus erythematosus (SLE), scleroderma (SSc) or rheumatoid arthritis (RA). Moreover the MCTD exhibits anti-U1RNP high-titer antibodies. However, anti-U1RNP antibodies are not specific or exclusive from MCTD, because of can be detected in other systemic autoimmune diseases as SLE, SSc and RA.

Objectives: To verify the differentiation of MCTD patients from other systemic autoimmune diseases using anti-U1RNP titers. The secondary objective is to characterise anti-U1RNP titer in other systemic autoimmune diseases with clinical manifestation.

Methods: An observational retrospective study of patients with inflammatory autoimmune disease evaluated in the Rheumatology Department from 2012 since 2016 was performed. In all cases a blood-test with anti-U1RNP, anti-Sm, anti-Ro, and anti-La analysis was conducted. Clinical data was registered according to the patients' medical history, with special emphasis being placed on renal affection, vascular affection, pulmonary hypertension, arthritis-synovitis, tendonitis-tenosynovitis, dry eye syndrome and Raynaud's phenomenon. Biostatistical analysis was performed using R.

Results: We collected data from 355 patients with a mean age of 50.84 (15.49) years, 98.55% of them were female. 13.8% of patients showed anti-U1RNP high titers (up to 20 pg/mL), and a significant increase of anti-U1-RNP in MCTD patients in contrast to other connective pathologies ($p < 0.0001$) was observed. Anti-Sm antibody also exhibit significantly higher values in MCTD patients than in RA ($p = 0.025$) or scleroderma ($p = 0.003$). No differences in anti-Ro and anti-La levels among all diagnosis were observed. Patients with the high anti-U1-RNP levels, regardless of the diagnosis, showed more Raynaud's phenomenon and vascular affection, ($p < 0.001$ y $p = 0.008$). Related to Anti-Ro and anti-La, high titer of these antibodies in patients with Dry eye syndrome was observed ($p < 0.001$). Specifically in SLE patients, those with the highest levels of anti-U1-RNP exhibit Raynaud's phenomenon ($p < 0.001$), highest levels of anti-La was shown in those patients with renal affection ($p = 0.02$) and the highest levels of anti-Ro and anti-La was shown in those patients with Dry eye syndrome ($p = 0.002$ and $p = 0.006$).

Conclusions: In our patient series anti-U1RNP were significantly elevated in MCTD diagnosis, and in lesser extent anti-Sm antibodies. Anti-Ro and anti-La antibodies are increased in dry eye syndrome patients. In SLE patients, anti-La increased levels were associated to renal affection.

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AB1139

RITUXIMAB FOR SCLERITIS AND PERIPHERAL ULCERATIVE KERATITIS ASSOCIATED WITH RHEUMATOLOGIC DISEASE

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Background: Some types of ocular surface inflammatory diseases are often related to rheumatic conditions: 37% of scleritis (especially diffuse and necrotizing forms) and 60% of peripheral ulcerative keratitis (PUK). Rheumatoid arthritis (RA) and ANCA-associated vasculitis are the most frequently related conditions. Significant loss of visual acuity can be observed if these ocular diseases are not properly treated. To date, no approved therapies are available. Consequently, the management of these ocular diseases are based on published evidence coming from clinical trials (scarce and often with a low sample size), observational studies and case reports. There are positive efficacy data of rituximab (RTX) for ocular surface inflammatory disease.^{1–4}

Objectives: To describe our experience with RTX as a therapy for severe ocular surface inflammatory diseases associated to rheumatic conditions.

Methods: This is a retrospective observational study. It includes patients with severe scleritis or PUK associated to rheumatic diseases diagnosed and managed at our Multidisciplinary Uveitis Clinic between January 2006 and November 2017. We recorded demographic and clinical variables. As outcome variables we used the