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 diagnosis[2]. One recent research reported 64% of RPC patients had a diagnostic delay with more than five years[3].

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 RPC in the department of rheumatology at Peking Union Medical College Hospital between October 2012 and October 2018, presenting as arthritis at onset, were retrospectively analyzed. 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. The most common joint affected was the knee. 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 ankyllosing 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.

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 above mentioned 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.

REFERENCES:
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THU0570

EFFICACY AND SAFETY OF ANAKINRA IN THE TREATMENT OF AUTOIMMUNE MYOCARDITIS

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Background: Virus-negative or “autoimmune” myocarditis/VNM) is a severe, inflammatory heart disease with a poor prognosis, and is a leading cause of inflammatory dilated cardiomyopathy (IDCM). Therapies are limited. Preliminary data indicate that interleukin-1(II-1) plays a key role in the initiation and maintenance of the inflammatory heart response, sustaining an auto-inflammatory cycle[1][2].

Objectives: to evaluate the efficacy and safety of anakinra(ANK) in in 9 patients with myocarditis/VNM, based on their present age (Group 1: 40-50 years, Group 2: >50 years).

Methods: Biopsy-proven VNM patients were enrolled and treated with ANK 100 mg daily subcutaneously. All patients received treatment with the maximum tolerated dose of any beta blockers and ACE-inhibitors, according to current guidelines. At baseline and 8±4 weeks after ANK therapy, all patients underwent a full evaluation with assessment of functional status (New York Heart Association[NYHA]), measurement of high-sensitive troponin T(hs-TnT) and NT-proBNP serum levels, electrocardiography (TTE) and TTE in patients with VNM.

Results: Eleven patients(F/M=5/6, mean age 46.2±12.2 years) diagnosed by hypothyroidism (16%), diabetes mellitus (10%) and cardiac disease (25%), class II-IV at baseline. Ten patients(90.9%) were in NYHA class I-II at the end of follow-up. Consistently, hs-TnT declined after 8 weeks(64.6±100.7 ng/L, p=0.028), and a similar trend was observed for NT-proBNP, even though not statistical significant(2582.6±5048.1 pg/ml,p=0.06). We did not observe any myocar-ditis-related death or complications, nor any ANK-related AEs.

Conclusion: Our pilot study supports the efficacy and safety of ANK in the treatment of inflammatory heart failure in VNM and provides the first clinical evidence to support the therapeutic blockade of IL-1 in myocarditis.

THU0571

THE IMPACT OF AGING ON FAMILIAL MEDITERRANEAN FEVER PATIENTS

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Background: Familial Mediterranean Fever (FMF) is a monogenic autoimmune-inflammatory disorder with innate immune activation with an onset before age 20 in approximately 90% of the patients. There is scarce data on the effect of aging on FMF patients over 40 years of age.

Objectives: This study aims to collect data on FMF patients who have survived over 40 years of age. Here we report our preliminary data on disease course and treatment status and comorbidities of our patients with FMF.

Methods: Among the FMF patients who have been followed in our FMF outpatient clinic with a pool of approximately 5000 patients, those who have aged 40 and over are being included to the study. As by today 180 patients are considered for evaluation. The files of patients were reviewed and a standard questionnaire was used to interview the patients. Here we report the results of 100 of these patients (56%) who were contacted for this purpose. These patients were questioned on their demographic characteristics, comorbid conditions, colchicine treatment details, and attack information. In order to see the trend of the change in the parameters assessed, the patients were divided into two groups based on their present age (Group 1: 40-50 years, Group 2: >50 years).

Results: A total of 100 (78 F, 22M) patients were evaluated. There were 61(46%, 15M) patients aged between 40-50 years and 39 (32M, 7M) over 50. The demographic characteristics and clinical features of these patients are given in Table 1. Besides, 3 patients were still on colchicine therapy. Ninety-six percent of the patients declared overall benefit from colchicine therapy; however 38% experienced a side effect related to this treatment. Over 88% of the patients reported decrease in severity and frequency of FMF attacks. The mean daily colchicine dose was lower in the age 50 and over group (1.7±0.77 mg versus 1.35 ±0.38 mg). There were no patients with AA amyloidosis in neither age group. The mean duration from the last attack increased from 15.3 ±17.9 months to 35.6 ±52 months in the older patients. One or more additional disease was present in 75% of this patient group. Among the comorbidities hypertension was the most frequent, diagnosed in 25% of the patients, followed by hypothyroidism (16%), diabetes mellitus (10%) and cardiac disease (5%). Sixty-five of the patients were receiving other medications in addition to colchicine.

Table 1. Clinical course and co-morbidities in two age groups over 40 years

<table>
<thead>
<tr>
<th>n</th>
<th>Group 1*</th>
<th>Group 2**</th>
<th>p</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sex (F:M); current age (mean±SD) (yr)</td>
<td>46(15):45.5 ±32.7 = 57.05 ±5.81</td>
<td>&lt;0.001</td>
<td></td>
</tr>
<tr>
<td>Mean duration since the last episode, (mean±SD, mo)</td>
<td>15.3 ±17.9 (1) = 35.67 ±52.05 (1)</td>
<td>0.012</td>
<td></td>
</tr>
<tr>
<td>Number of patients on colchicine therapy, n (%)</td>
<td>59(96.7)</td>
<td>38(97.4)</td>
<td>NS</td>
</tr>
<tr>
<td>Mean colchicine dose, mg/day (current)</td>
<td>1.70±0.76</td>
<td>1.41±0.45</td>
<td>0.03</td>
</tr>
<tr>
<td>Number of patients with decrease in attack severity, n (%)</td>
<td>54(88.5)</td>
<td>35 (89.7)</td>
<td>NS</td>
</tr>
<tr>
<td>Number of patients with decrease in attack frequency, n (%)</td>
<td>57(93.4)</td>
<td>37 (94.8)</td>
<td>NS</td>
</tr>
</tbody>
</table>