IgG4-RD were approved by ACR and EULAR in 2019. Whether this new criteria mimics IgG4-RD even lacking the tissue confirmation. Patients with moderately elevated serum IgG4 were diagnosed as IgG4-RD, including the 4 definite patients using the 2011 CDC. Among the 20 IgG4-RD patients according to the 2019 ACR/EULAR criteria, 19(95.0%) were male and median age of symptom onset was 62(46~69) years. There were 6(30.0%) patients diagnosed at hematopathology, 5(25.0%) at gastroenterology, 3(15.0%) at general surgery, 2(10.0%) at radiology medicine and 1(5.0%) at cardiology, endocrinology, orthopedics and urinary surgery, respectively. There were 8(40.0%) patients with bilateral lacrimal or salivary glands involved, 9(45.0%) with pancreas and biliary tree involved, and 6(30.0%) with chest involved.

(4) The median serum IgG4 of the 20 IgG4-RD patients was 15.4(0.14~55.10) g/L, median serum IgG3 was 279(172~50.29)g/L. There were 20.0%(4/20) patients had elevated serum eosinophil and 93.3%(14/15) had elevated serum IgG4 of the 20 IgG4-RD patients was 15.40(4.14~55.10) mg/L vs. 73.7% (27.5% vs. 73.7%, P < 0.001), and percentage of serum IgG4 ≥5× upper limit of normal were also lower than those of IgG4-RD patients (27.5% vs. 73.7%, P = 0.0002).

Conclusion: The 2019 ACR/EULAR criteria can help diagnosing patients with IgG4-RD even lacking the tissue confirmation. Patients with moderately elevated serum IgG4 need more clinical evidence to diagnose IgG4-RD and exclude mimics.

Methods: Patients suspected of having IgG4-RD due to elevated serum IgG4 and swelling or masses in single or multiple organs were recruited in Sun Yat-Sen Memorial Hospital, Sun Yat-Sen University from May 2013 and November 2019. Demographic and clinical data were collected. The diagnosis was reevaluated with the 2011 comprehensive diagnostic criteria (CDC) for IgG4-RD and the 2019 ACR/EULAR classification criteria for IgG4-RD, respectively.

Results:

(1) There were 68 patients recruited and 59(86.8%) of them had elevated serum IgG4 (≥135mg/dl) and 53(77.9%) patients showed swelling or masses in single or multiple organs. Most patients first visit general surgery (17(26%), gastroenterology (16.2%), respiratory medicine (16.2%) and rheumatology (14.7%).

(2) According to the 2011 CDC for IgG4-RD, 4(5.9%) patients were definite IgG4-RD, (1.15%) was probable and 42(61.8%) were possible. According to the 2019 ACR/EULAR criteria, 20(29.4%) patients were diagnosed as IgG4-RD, including the 4 definite patients using the 2011 CDC.

(3) Among the 20 IgG4-RD patients according to the 2019 ACR/EULAR criteria, 19(95.0%) were male and median age of symptom onset was 62(46~69) years. There were 6(30.0%) patients diagnosed at hematopathology, 5(25.0%) at gastroenterology, 3(15.0%) at general surgery, 2(10.0%) at radiology medicine and 1(5.0%) at cardiology, endocrinology, orthopedics and urinary surgery, respectively. There were 8(40.0%) patients with bilateral lacrimal or salivary glands involved, 9(45.0%) with pancreas and biliary tree involved, and 6(30.0%) with chest involved.

(4) The median serum IgG4 of the 20 IgG4-RD patients was 15.4(0.14~55.10) g/L, median serum IgG3 was 279(172~50.29)g/L. There were 20.0%(4/20) patients had elevated serum eosinophil and 93.3%(14/15) had elevated serum IgG4 of the 20 IgG4-RD patients was 15.40(4.14~55.10) mg/L vs. 73.7% (27.5% vs. 73.7%, P < 0.001), and percentage of serum IgG4 ≥5× upper limit of normal were also lower than those of IgG4-RD patients (27.5% vs. 73.7%, P = 0.0002).

Conclusion: The 2019 ACR/EULAR criteria can help diagnosing patients with IgG4-RD even lacking the tissue confirmation. Patients with moderately elevated serum IgG4 need more clinical evidence to diagnose IgG4-RD and exclude mimics.

References: This work was supported by Guangdong Medical Scientific Research Foundation (grant no. A2017093).

Disclosure of Interests: None declared.

DOI: 10.1136/annrheumdis-2020-eular.4426

AB1021
CHARACTERISTICS AND MANAGEMENT OF RHEUMATIC MANIFESTATION UNDER ESTROGEN RECEPTOR-TARGETING CANCER THERAPIES, DATA FROM A PROSPECTIVE REGISTRY

A. Dr. Patro1, L. Diekmann2, H. M. Lorenz3, B. Kraemer1, K. Benesova1, J. Leipe1.

1. University Hospital Mannheim, Division of Rheumatology, Department of Medicine V, Mannheim, Germany; 2. University Hospital Heidelberg, Heidelberg, Germany, Department of Medicine V, Hematology, Oncology and Rheumatology, Heidelberg, Germany; 3. University Hospital Heidelberg, Heidelberg, Germany, Department of Medicine V, Hematology, Oncology and Rheumatology, Heidelberg, Germany

Background: The knowledge about interdependences between rheumatic manifestations and malignancies is limited. Further, reliable data on the occurrence of rheumatic symptoms as side effects of specific cancer therapies beyond checkpointinhibitor-induced immune-related adverse events are sparse. In this regard, although arthritis under estrogen receptor-targeting therapies (aromatase inhibitors and the estrogen receptor modulator tamoxifen) have been frequently reported in oncological clinical trials and case reports, prospective data including an assessment of rheumatic manifestations by rheumatologists are lacking.

Methods: To contribute to a better understanding of interdependencies between rheumatic manifestations and cancer/estrogen blockade and potentially improve the management of both entities, pilot data were analysed.

Results: We identified 11 patients with rheumatic manifestations under estrogen receptor-targeting therapies (3 anastrozol, 4 letrozol, 8 tamoxifen) as part of breast cancer treatment. In addition to breast cancer one patient had a lymphoma 3 years after and another patient had a non-small cell lung cancer 2 years before breast cancer diagnosis. The patients had different cancer stages (5 IA, 3 IIA, 1 IIB, and 1 IVA). Their mean age at cancer diagnosis was 60.4 ± 11.6 years and all patients are females. The time interval between diagnosis of cancer and onset of symptoms/ rheumatic symptoms was 49.5 ± 34.0 months. Of interest, the time interval between onset of rheumatic symptoms and first assessment by a rheumatologist was 16.9 ± 22.3 months. The following systemic and rheumatic symptoms were reported: arthritis in 10, arthritis in 8 (small joints in 5, large joints in 3 affected), morning stiffness (>30min) in 7, IBP in 1, myalgia in 7, sicca symptoms in 2, fever in 1 (new-onset FMF with heterozygous M694U mutation), class IV glomerulonephritis and polyserositis in 1 (with new-onset SLE patient) (5). Disease burden at baseline was rather high with a mean VAS pain of 65 ± (±12.9)(100). Laboratory analyses revealed an increased CRP in 6/11 (55%) with a mean of 10.3 ± 8.2 mg/ml (>5). Autoantibody positivity was observed for ANAs in 5/10 (50%, titers ranging from 1:80 to 1:160), anti-cdsDNA in 1, rheumatoid factor in only 1/10 (10%) patients, none was anti-CCP positive. Before cancer onset, one rheumatologist (16.0%) had been treated with NSAR 3/11 (27%), 1/12 systemic glucocorticoids (91%) with an initial dose of 17.5 ± 19.5 mg and intra-articular glucocorticoids 1/11 (9%). Rheumatological assessment lead to initiation of csDMARDs (3/11 MTX, 1/11 SSZ, 1/11 HCQ, 1/11 AZA (later MMF/ rituximab in the SLE patient) 1/11 colchicine) as corticosteroid-sparing agents with good response in the majority of patients.

Conclusion: Our data demonstrate heterogeneous rheumatic manifestations, partially with severe manifestations beyond arthritis, so far not reported by oncological studies including follow-up, which might suggest an underreporting. Furthermore, despite close monitoring in tumor aftercare, our data show a considerable delay in referral to a rheumatologist and initiation of suitable treatment. The prospective design of the MalheurR registry enables future validation of our pilot data.

Disclosure of Interests: Alina Dr, Patroi Consultant of: Advisory board Novartis, Leonore Diekmann: None declared, Hanns-Martin Lorenz Grant/research support from: Consultancy and/or speaker fees and/or travel reimbursements: Abbvie, MSD, BMS, Pfizer, Celgene, Medac, GSK, Roche, Chugai, Novartis, UCBI, Janssen-Cilag, Astra-Zeneca, Lilly, Scientific support and/or educational seminars and/or clinical studies; Abbvie, MSD, BMS, Pfizer, Celgene, Medac, GSK, Roche, Chugai, Novartis, UCBI, Janssen-Cilag, Astra-Zeneca, Lilly, Baxter, SOBI, Biogen, Actelion, Bayer Vital, Shire, Octapharm, Sanofi, Hexal, Mundipharma, Thermoc... Fisher., Consultant of: see above, Bernhard Kraemer: None declared, Karolina Benesova Grant/research support from: Study grants for SCREENED study by Abbvie, Novartis and Rheumatold Baden-Württemberg, Consultant of: One-time participation in Novartis advisory board, Jan Leipe Grant/research support from: Consultancy and speaker fees: Abbvie, AstraZeneca, BMS, Celgene, Hospira, Janssen-Cilag, LEO Pharma, Lilly, MSD, Novartis, Pfizer, Roche, Sanofi, UCBI. Scientific support: Novartis, Pfizer, Consultant of: Consultancy and speaker fees: Abbvie, AstraZeneca, BMS, Celgene, Hospira, Janssen-Cilag, LEO Pharma, Lilly, MSD, Novartis, Pfizer, Roche, Sanofi, UCB. Scientific support: Novartis, Pfizer, Consultant of: Consultancy and speaker fees: Abbvie, AstraZeneca, BMS, Celgene, Hospira, Janssen-Cilag, LEO Pharma, Lilly, MSD, Novartis, Pfizer, Roche, Sanofi, UCB.

DOI: 10.1136/annrheumdis-2020-eular.6431