During follow up appointments, the patient described new recurrent violaceous patches with episodes of inflammation of the pinna of the ear, suggesting a diagnosis of relapsing polychondritis and so the patient was started on high dose prednisolone (80mg per day [1mg per kg]) and referred for rheumatological assessment. He had an excellent response to prednisolone (fever, ear swelling and rash subsided). The overlapping features of relapsing polychondritis and Sweet's syndrome in an elderly man suggested a diagnosis of VEXAS (vacuoles, E1 enzyme, X-linked, autoinflammatory and somatic) syndrome. The prednisolone dose was rapidly reduced to 10mg per day and the patient was commenced on methotrexate, as a steroid-sparing agent. Further blood tests have been sent for genetic analysis for VEXAS syndrome but results are pending.

Objectives: N/A

Methods: N/A

Results: N/A

Conclusion: VEXAS syndrome is a newly identified genetically defined syndrome, described by Beck et al in October 2020 consisting of somatic mutations in the *UBA1* gene, affecting bone marrow stem cells. In a study of 25 patients with this mutation, diagnostic/ classification criteria for relapsing polychondritis (n=15), Sweet's syndrome (n=8), polyarteritis nodosa (n=3) or giant cell arteritis (n=1) were met and patients often had severe refractory disease with overlapping systemic inflammatory and haematologic features. Features of VEXAS include the presence of vacuoles in myeloid cells, somatic mutations in the *UBA1* (ubiquitin-activating enzyme) gene, X-linkage (therefore only occurring in males), in older people with autoinflammatory syndromes. Although VEXAS syndrome is a relatively rare condition, it was a relevant consideration in this case.

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HPR Measuring health (development and measurement properties of PROs, tests, devices)

POS1454-HPR AESTHETIC IMPAIRMENT IN PATIENTS WITH SYSTEMIC SCLEROSIS: A CASE CONTROL STUDY USING A SEMI-QUANTITATIVE SCALE FOR BODY IMAGE ASSESSMENT

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Background: Systemic sclerosis (SSc) is a fibrotic autoimmune disease characterized by dermatological involvement. Skin involvement can alter appearance, which can have a psychological impact(1). Assessment of body image could be central in optimizing care. Yet, data are scarce(2).

Objectives: The main objective of our study was to assess aesthetic impairment measured on a visual aesthetic scale (AES) (3) in patients with SSc compared to a group of healthy subjects. Secondary objectives were to assess associations between aesthetic impairment and standardized questionnaires for aesthetic impairment as well as clinical, biological, psychological/quality of life, and functional parameters of SSc.

Methods: The "Sclero-esthet" study is a descriptive single-center study of two populations: patients with SSc, included in a referral center for systemic and autoimmune diseases at Lille Hospital, France, and healthy controls.

Results: This study, from January 2017 to October 2017, included 88 patients (69 [78.4%] women) with a median age of 52 years (range: 43 to 59) and 88 controls (49 [55.7%] women) with a median age of 45 years (range: 32 to 55). Perception of physical changes assessed using the AES was greater in cases than in controls (3.7 ± 0.3 vs 2.8 ± 0.3 , $p=0.028$), with statistical correlation with ASWAP. SSc patients with anxiety or depressive symptoms had significantly higher AES scores. Cases presented poorer quality of life than controls.

Conclusion: The AES appears to be a good tool to evaluate aesthetic impairment. Correlations found with psychological and quality of life parameters support the need to improve patient management in this field.

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POS1455-HPR RELATIONSHIP BETWEEN PSYCHOLOGICAL FACTORS AND MEASURES OF RHEUMATOID ARTHRITIS ACTIVITY

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Background: The patient-reported outcomes are important components of quantitative methods of rheumatoid arthritis (RA) activity assessment which are used to choose the appropriate drug therapy. The value of these parameters can be significantly affected not only by the inflammatory process, but also by the psychological characteristics of the patient and, in particular, by hardness [1].

Objectives: To study the relationship between psychological factors and signs of RA activity.

Methods: Patients with RA who met the EULAR/ACR 2010 criteria, and observed at the V.A. Nasonova Research Institute of Rheumatology were included. Clinical examination was performed including patient global assessment (PGA), physician global assessment (PhGA), pain measurement on a visual analog scale, tender joint count (TJC), swollen joint count (SJC). The functional status was determined by HAQ, the quality of life – by SF-36 EQ-5D, the nature of pain – by painDETECT, the presence of anxiety and depression – by HADS. Patients also completed Hardiness Survey questionnaire to assess hardiness (HDS) and 3 components of the HDS – commitment (CMT), control (CT) and challenge (CLN). Disease activity was evaluated with DAS28, CDAI, and RAPID3. All patients signed informed consent to participate in the study. Analysis of the data was performed using Spearman's rank test, Fisher exact test, chi-square and t-tests.

Results: 85 patients with RA were included. There were 69 women and 16 men. Mean age was 56.7 ± 13.1 years, disease duration – 7.6 ± 2.7 years. 72 patients were positive for rheumatoid factor, 75 – for anti-cyclic citrullinated peptide antibody. CDAI showed high activity in 15, moderate – in 37, low – in 30, and remission in 3 patients, DAS 28 – in 10, 55, 12, and 8, and RAPID3 – in 24, 25, 15, and 21, respectively. 24 patients had subclinically or clinically expressed anxiety and 15 – subclinically or clinically expressed depression (≥ 8 according to HADS). In 31 patients, the painDETECT questionnaire revealed possible or probable neuropathic pain. Mean HDS was 84.8 ± 21.7 , CMT – 38.9 ± 9.2 , CT – 29.4 ± 8.6 , CLN – 17.3 ± 7.1 . These values were comparable with the corresponding population data for this age group. There was a significant inverse correlation between HDS and RA activity measures, including SJC, TJC, DAS28 ($p<0.05$), pain, PGA, PhGA, CDAI, RAPID3, and HAQ ($p<0.01$). In addition, HDS and all its components positively correlated with quality of life, assessed by SF-36 and EQ-5D ($p<0.01$). In patients with subclinically and clinically expressed anxiety and depression, HDS, CMT, and CT were significantly lower than in patients without anxiety and depression ($p<0.01$), while the values of CLN in these groups did not differ significantly.

Conclusion: The results of the present study suggest that low HDS may be one of the significant factors determining RA activity level because it does not allow patients to adapt adequately to a stressful situation produced by the disease.

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