Methods: A total of 155 PsA patients were enrolled between May 2018 and December 2021. Adult-onset PsA CASPAR criteria were used to classify patients affected by PsA, while ASAS criteria were used to classify patients affected by axial-PsA (axPsA). All patients completed the PsA-STS, a simple instrument with five “thermometers” that incorporate pain, fatigue, physical function, skin disorders, and depression into a single assessment of disease activity. Additional continuous measures of disease activity (e.g., DAPSA, PASDAS, CPDAI) and patient-reported outcomes (PROs) measures of disease health status (e.g., PSAID and SF-36) were analyzed as comparisons. Spearman’s correlations and cross-tabulations were used to examine concurrent validity. Receiver operating characteristic (ROC) curve method was used to assess discriminant validity. As an external criterion, the Minimal Disease Activity (MDA) levels were used.

Results: The 96 female and 59 male patients ranged in age from 20 to 79 years old and had been living with PsA for an average of 8.352 yrs (6 months to 22 years). The PsA-STS area under the ROC curve (AUC-ROC) was 0.944. (95 percent CI 0.895 to 0.974) (Figure 2). The PsA-STS had same discriminant validity as the DAPSA, CPDAI, PASDAS, and PSAID, but it was better than the SF-36 (p=0.001). PsA-STS subscales were highly significantly different between the MDA status (all at p<0.0001) when categorizing patients into those who reported their condition as reaching MDA (Kruskal-Wallis test). PsA-STS were also shown to have a remarkable (p<0.0001) correlation with known PsA activity measurement techniques.

Conclusion: For the evaluation of disease activity in PsA, PsA-STS is a useful alternative to continuous composite indices and other patient-centered assessments. The PsA-STS make data collection simpler, and they should be used in both clinical studies and everyday clinical treatment. In order to establish the tool’s clinical relevance, longitudinal construct validity, which concerns the measure’s capacity to identify a real change in health status as well as its accuracy in detecting changes of various magnitudes, must be addressed.

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Disclosure of Interests: None declared

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Challenging cases to make you think

**POS0012**

A NEW TNFRSF1A GENE MUTATION IN A TURKISH FAMILY WITH TNF RECEPTOR-ASSOCIATED PERIODIC SYNDROME (TRAPS).

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**Background:** Tumour necrosis factor (TNF) receptor-1 associated periodic syndrome (TRAPS) is a rare autosomal dominant autoinflammatory syndrome caused by pathogenic variants in the tumour necrosis factor receptor 1 (TNFR1) gene (TNF receptor superfamily member 1A, TNFRSF1A). For definitive diagnosis, genetic tests are needed to show variants in the TNFR1 gene (1). Information on genotype-phenotype relationships is limited due to the rarity of the disease.

**Objective:** This study presents novel R97S mutation in TNFRSF1A gene and relevant clinical findings in a patient with amyloidosis of unknown etiology and his sister.

**Methods:** Case; Proband 33-year-old male patient presented with swelling in the legs 9 years ago. In his history, he described muscle pain, unilateral eye redness, swelling and discharge, which occurred several times in 2-3 months and lasted for up to 2 weeks. Kidney biopsy was found to be consistent with AA amyloidosis.

**Results:** The proband was started first and then continued with hemodialysis due to end-stage renal failure. The renal biopsy revealed apple-green birefringence with congo-red stain, which was confirmed to be AA amylodiosis. There were moderate to good, with slightly better scores for both the WPS-RA and WPAl instruments both measuring the impact of OA and IA on productivity.

Our country is endemic for FMF disease, a study by Bilge et al. (2) demonstrated that mutation analysis of 9% of all subjects with FMF in our country provided negative results. In our opinion genetic analysis should be performed to detect other autoinflammatory diseases in cases that classical FMF attacks are not seen, as in this case series. These and similar cases followed as FMF are likely to be associated with other autoinflammatory diseases. Diagnosing these rare diseases will provide both appropriate and effective treatment options to patients and a better understanding of the clinical feature - mutation relationship.

**Disclosure of Interests:** None declared


**Figure 1.** Family’s pedigree

**PO0013**

SJÖGREN-LIKE SYNDROME, MULTIPLE AUTOIMMUNE DISEASES, THYMOID AND GENETIC VARIANTS IN AIRE

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**Background:** The autoimmune regulator gene AIRE is the main controlling factor for developing central immunotolerance. Variants in AIRE lead to autoimmunity and underly the autoimmune polyendocrine syndrome type 1 (APS-1 or APECED), defined by the clinical triad chronic mucocutaneous candidiasis, hypoparathyroidism and adrenal insufficiency. (1, 2) Manifold additional manifestations have been described, including Sjögren-like disease lacking the typical serology; (3) Furthermore, serological abnormalities, e.g. antibodies against IL-22, IL-17F and IFNα are a hallmark of the disease. Due to the diversity in clinical manifestations and the genetic variants in AIRE, diagnosis is often delayed.

**Objective:** Case report of a patient with 2 new variants in AIRE with diverse rheumatological and autoimmune disorders as well as thyroma.

**Methods:** Case report

**Disclosure of Interests:** None declared


Our country is endemic for FMF disease, a study by Bilge et al. (2) demonstrated that mutation analysis of 9% of all subjects with FMF in our country provided negative results. In our opinion genetic analysis should be performed to detect other autoinflammatory diseases in cases that classical FMF attacks are not seen, as in this case series. These and similar cases followed as FMF are likely to be associated with other autoinflammatory diseases. Diagnosing these rare diseases will provide both appropriate and effective treatment options to patients and a better understanding of the clinical feature - mutation relationship.

**REFERENCES:**
