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THU0292 DIAGNOSTIC ACCURACY OF ULTRASOUND AND ULTRASONOGRAPHIC FEATURES OF SALIVARY GLANDS IN PATIENTS WITH PRIMARY SJOGREN'S SYNDROME

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Objectives: To analysis ultrasonography (US) changes of salivary glands (SG) in patients with primary Sjogren's syndrome (pSS) and assessment of their accuracy for diagnosis pSS.
Methods: This study included 205 pSS patients (mean age 53.9±11.5, disease duration 5.6 years) and 87 healthy controls (mean age 52.3±14.7). All pSS patients fulfilled the AECG diagnostic criteria. The disease activity was measured by EULAR SS disease activity index (ESSDAI). Parotid and submandibular glands on both sides were assessed for size, parenchymal echogenicity and inhomogeneity, posterior glandular border and presence of intraglandular lymph nodes. Inhomogeneity of the salivary glands were graded according to the De Vita scoring system [0) homogenous glands; 1) mild inhomogeneous - isolated hypoechogenic areas; 2) evident inhomogeneous - scattered hypoechogenic areas, and/or multiple punctate or linear densities; 3) grossly inhomogeneous – large or confluents hypoechogenic areas, and/or to linear densities, and/or multiple cysts. The global SGUS score (0–6) was the sum of the scores of each pair of salivary glands. Statistical analysis was performed by SPSS v16. Data were compared using *t*-test, χ^2 test and Mann-Whitney U test. The optimal cut-off value for SGUS score was calculated as the area under the receiver operating characteristic curve (AUC-ROC).
Results: Xerophtalmia and xerostomia were presented in 185/205 (90.2%) and 186/205 (91.2%), respectively. According to ESSDAI, the majority of pSS patients 88/205 (43%) had moderate disease activity. Seventy-eight per cent of pSS patients were anti-SSA antibody positive, 44% anti-SSB/La antibody positive. Biopsy of LSG was positive in 140/172 (81.4%) pSS patients. US abnormalities were established in 197 (96%) pSS patients and in 16 (18%) controls ($p<0.0001$). Pathological sizes of salivary glands were more frequently in pSS patients than controls, 111 (54.2%) vs. 3 (3.4%) patients, respectively ($p<0.0001$). The echogenicity of the salivary glands was pathological changes in 142 (69.3%) pSS patients and in only 5 (5.7%) control group ($p<0.0001$). The pathological glandular border was frequently in pSS patients than control group, 48 (23.4%) vs. 2 (2.3%), $p<0.0001$. No differences were detected between the two groups of patients for enlarged intraglandular lymph nodes. Most of pSS patients had pathological inhomogeneity, 197/205 (96.1%) vs. 16/85 (18.4%) in control group ($p<0.0001$). The median SGUS was significantly higher in pSS patients in comparison with control group [median (range) 4 (0–6) vs. 0 (0–2), $p<0.0001$]. Forty-five percent of pSS patients had SGUS score 4. The SGUS cut-off ≥ 2 showed specificity of 89.5% and sensitivity 89.3%. Diagnostic accuracy of the parenchymal inhomogeneity was very good (AUC-ROC 0.89), followed by the glandular echogenicity (AUC-ROC 0.81), the glandular size (AUC ROC 0.75), the posterior border (AUC ROC 0.60), and the presence of intraglandular lymph nodules (AUC ROC 0.49), respectively.
Conclusions: Our findings confirm that most of established pSS patients had pathological SGUS features. Among US parameters, parenchymal inhomogeneity was the most discriminant feature for diagnosis of SS. There is the growing evidence that ultrasound should be considered as the useful method for evaluation of salivary glands in pSS patients.
Disclosure of Interest: None declared
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THU0293 B-CELL RESPONSES TO TYPE I INTERFERON DEFINE DISEASE ACTIVITY IN SLE AND CAN BE MEASURED BY CELL SURFACE TETHERIN (CD317)

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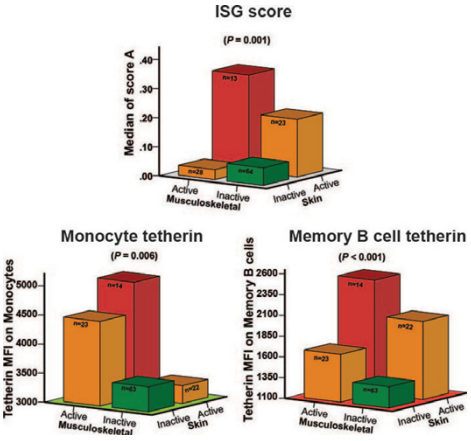
Background: SLE is a Type I interferon (IFN-I) mediated disease with autoreactive B cells. Whole blood interferon-stimulated gene (ISG) expression is used to measure IFN-I status but does not consistently correlate with clinical features and therapy response. ISG expression may be influenced by differing response in individual cell subsets as well as IFN-g. Tetherin is a cell surface protein encoded by the interferon-stimulated gene *BST2*.
Objectives: To evaluate tetherin expression as a novel cell-specific flow cytometric biomarker for IFN-I response.
Methods: *In vitro,* we tested response of expression of *BST2* and 30 other ISGs to stimulation with IFN-a and IFN-g, as well as dose response of *BST2* and tetherin protein in B cells. Sorted cells (Monocyte, T, NK, naïve and memory B, plasmablast) from 8 controls and 10 SLE patients were used to test variation in expression of 31 ISGs between cell subsets and whether tetherin measured by flow cytometry is a cell specific marker for IFN-I response. Samples from 156 SLE patients, 30 ACPA+ANA- RA (DAS28>3.2) patients and 22 healthy controls (HC) were used in 3 clinical validation studies of tetherin vs. ISG score against diagnosis, disease activity (BILAG-2004) and plasmablast repopulation after rituximab.
Results: Some ISGs' expression in B cells increased in response to both IFN-a and IFN-g. Others, which included *BST2*, were selective for IFN-a. An 18-gene ISG score derived from this latter group was calculated for comparison with tetherin.
Results from cell sorting showed that ISG Score was highest in monocytes; other subsets were 75–85% lower ($p<0.001$). SLE-associated increase in expression (SLE:HC ratio) varied between 2.56 (T cells) to 4.93 (plasmablasts).
Diagnosis: ISG score differentiated HC from SLE with ratio 3.58 (1.94 – 6.61) and large effect size 0.14 (partial eta squared). Using tetherin for cell specific IFN response revealed marked differences between subsets. Monocytes did not differentiate HC and SLE at all with ratio 1.19 (0.87 – 1.61) and effect size 0.03. Memory B cells had medium-large effect size of 0.11 with ratio 1.59 (1.21 – 2.09). Comparing SLE and RA the largest effect size was for plasmablast Tetherin at plasmablasts at 0.23, with ratio 2.20 (1.66 – 2.93).
Disease activity: ISG score was associated with cutaneous disease activity (BILAG A/B) but not musculoskeletal (Fig 1). Monocyte tetherin was associated with musculoskeletal disease activity but not cutaneous. Memory B cell tetherin was associated with disease activity in both these organs. Memory B cell tetherin was increased with renal (p=0.005) or haematological (p=0.005) activity with no differences in ISG score for these domains (p=0.152, p=0.989 respectively).
Plasmablast numbers after rituximab were associated with Memory B cell tetherin (R=0.38, p=0.047) but not ISG score (R=0.24, p=0.219).



Conclusions: ISG expression in unsorted blood is influenced by IFN-g and cellular composition of the sample as well as IFN-I. Flow cytometric measurement of surface tetherin is a cell-specific assay for IFN-I that avoids these problems. Memory B cell tetherin was better associated with plasmablast numbers and clinical features of disease than monocyte tetherin or ISG score.
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THURSDAY, 15 JUNE 2017

Vasculitis

THU0294 DIFFERENTIAL PATTERNS OF ATROPHY IN HIPPOCAMPUS AND BRAINSTEM BETWEEN CHRONIC PROGRESSIVE NEURO-BEHCET'S DISEASE AND ALZHEIMER'S DISEASE

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Background: Central nervous system involvement is one of the most serious complications in Behçet's disease (BD). This condition is referred to as neuro-Behçet's disease (NB) and can be classified into acute type (ANB) and chronic progressive type (CPNB) based upon differences in the clinical course and responses to corticosteroid treatment. Cerebellar ataxia, such as gait disturbances and dysarthria, is one of the representative manifestations in CPNB. Accordingly, brainstem atrophy is frequently observed in CPNB, but not in ANB. Notably, progressive neurobehavioral changes mimicking those in Alzheimer's disease (AD) are also frequently observed in CPNB, but these changes cannot be accounted for by brainstem atrophy.