Catastrophic antiphospholipid syndrome in pregnancy: Case series

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Background: Catastrophic antiphospholipid syndrome (CAPS) is a rare condition associated with high mortality. Prompt recognition and treatment is important for optimal outcomes. Clinical features of CAPS can overlap with obstetric complications, leading to diagnostic challenges.

Objectives: To describe two patients with CAPS during immediate post-partum period.

Methods: We report two cases in a single centre over two years. Results: A 38 year old with obstetric antiphospholipid syndrome (APS) presented with epigastric pain, transaminitis and thrombocytopenia in keeping with evolving HELLP syndrome. Spontaneous complete miscarriage ensued. Post-delivery, she developed worsening abdominal pain and transaminitis (Table 1). Imaging demonstrated hepatic infarcts and patchy ground glass lung opacities. She was treated for CAPS with intravenous steroids and anticoagulation, and recovered.

A 39 year old with primary APS (deep venous thrombosis and obstetric features) whose antenatal course was fraught with minor per vaginum bleeding (PVB) presented with heavy PVB at 22 weeks gestation requiring cessation of anticoagulation. She developed a deep venous thrombosis. Risk of clot propagation versus bleeding prompted cautious anticoagulation. Labour was induced for declining maternal health and poor foetal prognosis at 24 weeks, 3 days. Post-delivery, she developed abdominal pain, headache, transaminitis and worsening thrombocytopenia with heavy PVB requiring uterine artery embolization. Given features that were consistent with micro thrombi and ischaemia (Table 1), she was treated for CAPS and HELLP syndrome with intravenous steroids, plasma exchange (PLEX) and intravenous immunoglobulin (IVIG), and recovered.

Conclusion: A significant correlation was observed between serology, ocular surface damage (OSS), and ocular surface inflammation (MMP-9). The positivity of RF isotypes IgM and IgA could alert the clinician about the ocular damage to initiate a study directed to this organ. Nevertheless this present study shows preliminary outcomes, we need to enroll more patients to obtain better outcomes.

REFERENCES:

Disclosure of Interests: None declared

FR10221 CATASTROPHIC ANTIPHOSPHOLIPID SYNDROME IN PREGNANCY: CASE SERIES

FR10222 ANALYSIS OF CLINICAL AND SEROLOGICAL PROFILE OF PATIENTS WITH PRIMARY SJÖGREN’S SYNDROME AND AN EARLY DISEASE ONSET AT AGE BEFORE 35 YEARS

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Background: Primary Sjögren’s Syndrome (pSS) is a chronic systemic autoimmune disease with diverse clinical picture, extending from exocrinopathy to systemic disease and non-Hodgkin’s lymphoma (NHL). Usually, it affects middle aged women, but early disease onset (<35 years old) has been also observed. So far, there is a limited number of studies to explore whether the age of SS onset affects the clinical SS phenotype. To definitely answer this question a well-organized, large, harmonized cohort of patients with a continuous follow up is needed, through the ongoing European project (HarmonicSS, grant agreement no: 731944).

Objectives: To investigate whether the clinical and serological picture of pSS patients with early disease onset (<35 years old) differs to that of middle-age onset.

Methods: Medical records of 717 pSS patients, included in two Greek cohorts fulfilling the 2002 revised European/American International classification criteria for SS, were evaluated. The study group included 133 patients with disease onset <35 years (median disease duration, range: 10, 0-32 years; group A) matched at 1:1 ratio according to gender and disease duration with 133 pSS patients with middle-age onset (mean...
age at disease onset 52±5 years; median disease duration, range: 10, 0-30; group B). Clinical and laboratory data were collected through an extensive clinical chart review. All parameters were compared by chi-square or Fisher’s exact test, when appropriate.

Results: The two pSS groups had similar frequencies of non-specific clinical findings (chronic fatigue, arthralgias-myalgias, arthritis, Raynaud’s phenomenon), periarteriitis nodosa, interstitial nephritis, lung and liver disease), splenomegaly, leukocyte and neutrophil counts. However, patients with disease onset ≤35 years old, exhibited increased proportion of salivary gland enlargement (SGE; 43.8% vs 25.4%, p=0.003), lymphadenopathy (18.2% vs 7.6%, p=0.016), palpable purpura (20.3% vs 9.8%, p=0.025), anti-Ro/SSA antibodies (90.2% vs 74%, p=0.001), rheumatoid factor positivity (73.3% vs 53.6%, p=0.002), low C4 levels (64.5% vs 42.4%, p=0.001) and NHL (18% vs 8.3%, p=0.028).

Conclusion: Our findings suggest that young patients with pSS display increased prevalence of systemic features, B cell hyperreactivity, as well as heightened risk for lymphoma development. Further prospective studies with a larger number of patients are needed to address whether early disease onset may also serve as an additional risk factor for NHL development.

REFERENCES:

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FR10223 EXTRACELLULAR VESICLES AS A SOURCE OF BIOMARKERS IN SJÖGREN’S SYNDROME: A SWATH-MS PROTEOMIC APPROACH

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Background: Primary Sjögren’s syndrome (pSS) is a multifactorial autoim-mune disorder characterized by lymphocytic infiltration of the exocrine glands. In the recent past several proteomic studies attempted to look at valid biomarkers for pSS in whole saliva, however little is known about the composition of salivary extracellular vesicles (EVs), nor to what extent their content may reflect the phenotypic state of the disease.

Objectives: In this study, we search for pSS specific biomarkers by using a sequential window acquisition of all the theoretical fragment ion spectra (SWATH-MS) approach to monitor the dynamics of saliva EVs sub-pro- teome of pSS patients compared to healthy controls.

Methods: We included patients with a diagnosis of pSS made according to the AECG 2002 criteria and healthy volunteers as controls. Saliva was collected under standardized conditions. EVs were enriched by sequential ultracentrifugation steps from saliva samples. Peptide identification and quantitation was performed by matching SWATH data against an assay database. Protein expression and network analyses were performed using Cytoscape software.

Results: We included 20 pSS patients (AECG 2002 criteria) and 10 healthy subjects. Quantitative data evidenced a distinct separation between the patient group and control group, indicating that pSS may modulate the phenotype of protein cargoes of salivary EVs. The majority of the proteins up-regulated in pSS compared to controls were found to be involved in several inflammatory processes. Particular emphasis was given to proteins conveying to IL-12 signaling that included annexin A2, coiflin, macrophage migration inhibitory factor (MIF), S100A8-A9 and plastin-2 proteins.

Conclusion: Our results revealed that the inflammatory phenotype observed in pSS patients is also extended to salivary EVs, which protein content may represent a novel source for potentially useful biomarkers for pSS.

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FR10224 PREVALENCE OF SJÖGREN’S SYNDROME IN THE COMMUNITY OF MADRID

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Background: Several studies on the epidemiology of autoimmune diseases performed within the past several decades revealed a prevalence of Sjögren Syndrome (SS) between 0.3% and 4.83% (1). More recent studies revealed a lower prevalence between 31/100,000 to 49/100,000 (2-3). The exact prevalence of SS is unknown due to the heterogeneity of the populations and geographic areas studied, the utilization of different diagnostic tests or the lack of a unique classification criteria for the disease. Also, the disease may have an insidious onset and a variable course with a broad spectrum of clinical manifestations, so the diagnosis may be delayed or SS patients may be missed and misclassified as other rheumatic disease. Besides, SS can occur alone (primary SS) or in association with other specific systemic autoimmune rheumatic diseases (secondary SS).

Objectives: The aim of our study was to determine the prevalence of SS in the Community of Madrid (CM) and to describe the sociodemographic and clinical characteristics of these patients.

Methods: Population-based cross-sectional study in the CM, Spain. The information source for SS cases was the Registry of rare diseases in the CM (SIERMA).

A descriptive analysis of the main sociodemographic and clinical characteristics of SS cases was performed. Prevalence per 10,000 inhabitants in people with 18 years of age and over, stratified by sex and their 95% confidence intervals (CI) were calcu-lated for 2015. The denominator was the people registered in the Popu-lation Register in the middle of the period (July 2015).

Results: There were 4,778 cases of SS in SIERMA and 389 (8.1%) of them were already dead. 4434 (92.8%) were women. The median age was 64.7 years (15.4), and the maximum age was 103 years. The disease was most frequent in the sixth decade with 1079 cases (22.6%), in the seventh decade with 1120 cases (23.4%) and in the eighth decade with 935 cases (19.6). 3116 cases (65.2%) were classified as primary SS (pSS) and 1662 (34.8%) as secondary SS (sSS).

Among the sSS the main rheumatological conditions associated were rheumatoid arthritis (58%), lupus (25.4%), systemic Sclerosis (10.5%); mixed connective tissue (4%), inflammatory muscle disease (2%) and vasculitis (1%).

The prevalence of SS in adults (≥ 18 years of age) was 8.4/10,000 with a 95% CI: 8.2 – 8.7 (14.9% in women and 0.8% in men). The prevalence of pSS was 5.5/10,000 (CI 5.3 – 5.7) and for sSS was 2.8/10,000 (95%CI: 2.7-3.1).

Conclusion: SS mainly affects females during the sixth, seventh and eight decades of life, and shows a female/male ratio of 9:1. Two out of three cases of SS identified were classified as pSS. The main rheuma-tological conditions associated were rheumatoid arthritis and lupus. The prevalence of sSS in the Community of Madrid population is lower than the observed in previous studies whereas the prevalence of pSS is similar than the observed in current studies.

REFERENCES: