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AB0979

MEVALONATE KINASE DEFICIENCY AT TREATMENT WITH CANAKINUMAB: RARE BLEATED CUTANEOUS FEATURE

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Background: Mevalonate Kinase Deficiency (MKD) is an autosomal recessive autoinflammatory syndrome. Diagnostic criteria are based in clinical and genetic features (mutations in MVK gene) and its main treatment consists of blocking IL-1¹⁻⁵.

Objectives: To describe a case-series of a rare bleated cutaneous feature.

Methods: From January 2004 to September 2019, all cases diagnosed of MKD have been reviewed.

Results: 15 patients had MKD diagnosis (11 pathogenic mutations, homozygosis or double heterozygosis). Most common symptoms were oral and genital aphthous, abdominal pain, adenopathies and recurrent fever. Mean age of diagnosis was 10,67 years old (8,67 years later from the beginning of symptoms). Mean time of follow-up was 10 years. 3 patients developed this rare bleated cutaneous feature: suppurative hydrosadenitis. When the first injury appeared, all were at treatment with Canakinumab (mean time of treatment 4 years) and had 7,67 years of MKD course. This comorbidity began as repeating abscesses in folds with apocrine glands and hair follicles (armpits, inguinal, anal and genital folds). 2 patients continued Canakinumab and 1 switched to Adalimumab because of severity of cutaneous involvement.

Conclusion: This is the first case-series showing suppurative hydrosadenitis associated to MKD that has not been described in literature. Both theories have been found: as immune-mediated disease partnering this autoinflammatory syndrome (MKD) or as adverse event of treatment (anti-IL-1, less presumable).

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AB0980

CASE-REVIEW ON FAMILIAL MEDITERRANEAN FEVER IN A SPECIALIZED CENTRE

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Background: Familial Mediterranean Fever (FMF) is a genetic autoinflammatory disorder caused mostly by mutations in MEFV gene. Its inheritance is autosomal recessive and is the most frequent periodic fever syndrome. First-line treatment is based in colchicine use, so biologics (anti-IL-1) are reserved for refractory cases^{1,2}.

Objectives: To account for clinic and treatment features of patients with FMF in a specialized center as opposed to non-referent centers.

Methods: This study was developed in the Pediatric Rheumatology Service in Hospital Universitario y Politécnico La Fe de Valencia. Demographic, clinic and treatment data were collected from patients diagnosed of FMF since January 2004 to September 2019.

Results: 106 patients met last FMF criteria³. 55% had a pathogenic mutation in genetic analysis. 52% were female. Before 10 years old, 71% of patients had the diagnosis (51% before 4 years old). Arthralgia/myalgia (73%), periodic fever (62%) and abdominal pain (54%) were the most common symptoms. Systemic

Juvenile Idiopathic Arthritis (JIA, 6), other forms of JIA (9) and vasculitis (10) were the most prevalent comorbidities. When talking about treatment, 76,4% received Colchicine (60,5% with good response), 22,6% needed a classical disease modifying antirheumatic drug (mostly Methotrexate) and 22 patients got biologic treatment (73% anti-IL-1).

Conclusion: When analyzing this case-review, JIA has a strong association with our patients, so it could explain severe disease activity and more articular involvement. This could be an illustration to the higher use of Methotrexate. Also, the most relevant symptom was arthralgia while fever is the most frequent in literature. Likewise, age of diagnosis has been earlier than other case-series (this would be more frequent in other autoinflammatory syndromes, as literature relates)^{1,2,4}.

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AB0981

VALIDATION OF THE CAREGIVERS QUESTIONNAIRE (IMPACT OF PEDIATRIC RHEUMATIC DISEASES ON CAREGIVERS MULTIASSESSMENT QUESTIONNAIRE) IN PEDIATRIC RHEUMATIC DISEASES

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Background: The CAREGIVERS questionnaire is used as a multidimension screening instrument for burden of caregivers, it allows to examine the impact of juvenile idiopathic arthritis (JIA) on several areas of the caregiver's life, looking for specific concerns and needs in which treatment could be required. This instrument has shown a good understanding among participants, reliability and consistency to measure the psychosocial and economic impact on primary caregivers of patients with JIA.

Objectives: The aim of this study is to validate CAREGIVERS questionnaire with the most common pediatric rheumatic diseases, such as JIA, systemic lupus erythematosus (SLE) and juvenile dermatomyositis (JDM).

Methods: The external validation phase of the constructed questionnaire will be carried out following Feinstein's clinimetric and psychometrics methodology, to measure the impact of the disease on the caregiver and create a risk profile for treatment detachment.

Results: A total of 200 questionnaires are applied to caregivers of pediatric patients with rheumatic diseases: 109 caregivers of patients with JIA, 28 caregivers of patients with DMJ and 63 caregivers of patients with SLE. When analyzing all the questions together, a general Cronbach's alpha of 0.6751 was obtained (Table 1).

Table 1. External validation of the Questionnaire

Dimension	Numero de ítems	Alfa de Cronbach
I. Impacto Emocional	6	0.5669
II. Impacto Social	3	0.47
IIIA. Impacto Financiero	4	0.6736
IIIB. Impacto Laboral	3	0.3151
IV. Impacto Familiar	5	0.4948
V. Impacto en la Relación entre Cuidador-paciente	1	NA
VI. Impacto en la relación de pareja	1	NA
VII. Impacto en la Religión/Creencias/Espiritualidad	1	NA
VIII. Impacto de las Redes Sociales	4	0.6642
Total	28	0.6751

Conclusion: The CAREGIVERS questionnaire showed to be validated to assess the impact of pediatric rheumatic diseases.

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AB0982 SERUM ALBUMIN LEVELS AND DEPRESSION IN JSLE

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Background: Albumin is a negative acute phase response protein synthesized in the liver, being an important marker of inflammation. Under inflammatory conditions, the transcapillary escape rate of albumin may increase, leading to hypoalbuminaemia. Systemic lupus erythematosus (SLE) is a chronic condition involving multiple organ systems, inducing functional disability and psychological burden responsible for noteworthy depressive symptoms¹. Depression may be related with psychosocial, environmental and biological factors, disease activity and its severity, age and sex². Several studies show that immune activation and increased concentrations of positive and decreased concentrations of negative acute phase proteins are involved in the pathogenesis of depression³. As albumin has the capacity to bind homocysteine, lowered serum albumin levels leads to hyperhomocysteinemia, a well-known risk factor for depression. Moreover, hypoalbuminaemia decrease the availability of tryptophan, an essential amino acid from which the neurotransmitter serotonin is derived, and induce oxidative stress, which further decreases antioxidant levels in people with depression.

Objectives: To assess the association between serum albumin levels and depressive symptoms in juvenile-onset SLE (JSLE) patients.

Methods: A cross-sectional sample of JSLE patients, currently aged ≥ 16 years, completed a psychosocial assessment including quality of life (SF-36) anxiety and depressive symptoms (HADS) and cognitive assessment (MMSE), between October 2018- May 2019. Local Ethics Committee approved the study. All patients fulfilled both 2012 and 2019 EULAR/ACR classification criteria for SLE. Juvenile-onset was defined as age at diagnosis <18 years. Demographics and clinical characteristics were collected. Statistical analysis was performed with SPSS®. Variables were compared with spearman correlations tests.

Results: 30 JSLE patients were included (90%female) in the study, with median (min-max) age of 21 (16-35) years, with mean (SD) age of diagnosis of 15.8 ± 2.1 . Median albumin serum level was 41.7 (16.7-46.3) g/dL. Psychosocial assessment revealed a mean (SD) score in HADS - Depression of 3.9 (3.3), HADS - Anxiety of 9 (4.3), MMSE of 27.7 (1.8), Physical health SF-36 of 66.8 (9.9) and Mental health SF-36 of 68.9 (17.5). 23.3% JSLE showed mild cognitive impairment, 63.3% anxiety and 13.3% depression. We observed significant inverse linear relationships between serum albumin levels and depressive symptoms score ($p=0.042$, $\rho=-0.380$) and with anxiety symptoms score ($p=0.029$, $\rho=-0.406$). No significant correlations were detected between albumin serum concentrations and cognitive assessment.

Conclusion: Our findings are consistent with studies previously reporting the potentially protective effect of high serum albumin levels on mental health in different populations. A possible inflammation related aetiology for depression in JSLE patients is highlighted, further explained through the protective roles played by albumin in inflammation, infection, and oxidative damage.

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AB0983 LIPID METABOLISM AND DISEASE ACTIVITY IN JSLE PATIENTS

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Background: Systemic lupus erythematosus (SLE) is an autoimmune systemic disease associated with premature atherosclerosis. Risk factors include dyslipoproteinemia, inflammation, oxidized low-density lipoprotein (LDL), hyperhomocysteinemia and antiphospholipid antibodies. Hyperlipidemic condition is being reported to promote the production of proinflammatory cytokines such as IL-1 β , IL-6, and IL-27 and lowering blood lipid levels improves the disease. Oxidative stress is elevated, mainly due to mitochondrial dysfunction, further disrupting lipid metabolism. Some drugs also have an impact on lipid profile, such as chronic steroid use, which worsens LDL, HDL, and TG levels.

Objectives: To assess the relationship between lipid profile and disease activity in juvenile SLE (JSLE) patients.

Methods: Retrospective study of JSLE patients, fulfilling both 2012 and 2019 EULAR/ACR classification criteria for SLE. Juvenile-onset was defined as age at diagnosis <18 years. Demographics and clinical characteristics were collected. To evaluate the activity of JSLE, the Systemic Lupus Erythematosus Disease Activity Index (SLEDAI) was used. Statistical analysis was performed with SPSS®. Spearman's rank non-parametric test or Pearson's parametric test were used to assess the bivariate correlation for inflammatory and metabolic variables. P value <0.05 was considered significant for all the statistical tests.

Results: 35 patients were included, with current median (min-max) age of 22 (16-35) years, mean (SD) age of diagnosis of 15.8 (2.4) years; 91.4%female. Median ESR was 19 (2-75) mm/h, CRP 1.65 (0.1-9.6) mg/L, albumin 41.6 (16.7-46.3) g/L, proteinuria 0.2 (0-3) g/dL, leukocyturia 0 (0-1362.7)/uL, erythrocyturia 0 (0-501.9)/uL and anti-double stranded DNA 89.3 (10-800) U/mL. Mean C3 was 102.1 (21.6), C4 17.1 (7.4) mg/dL and creatinine 0.63 (0.1) mg/dL. Median SLEDAI was 2 (0-12). All were ANA positive, 40 % positive for antinucleosome antibodies, 25.7% anti-ribosomal P protein antibody, 11.4% anti-Sm, 8.6% autoantibodies against $\beta 2$ -glycoprotein I, 8.6% anti-cardiolipin, 14.3% lupus anticoagulant, 37.1% anti-SSA and 8.6% anti-SSB. Articular manifestations were present in 48.6%, mucocutaneous in 77.1%, haematological in 45.7%, lupus nephritis in 42.9%, serositis in 8.6% and pulmonary interstitial disease in 2.9%. Mean (SD) total cholesterol values (TC) was 165.5 (44.7) mg/dL and LDL 94.5 (29.9) mg/dL. Median high-density lipoprotein was 52 (28-92) and triglycerides (TG) 81.5 (41-253) mg/dL. Median daily prednisolone dose was 5 (0-40) mg. 88.6% were treated with hydroxychloroquine, 31.4% with mycophenolate mophetil and 14.3% with azathioprine. TC was negatively correlated with serum albumin ($p=0.043$, $\rho=-0.378$) and positively with SLEDAI ($p=0.032$; $\rho=0.392$), proteinuria ($p=0.009$; $\rho=0.469$) and leukocyturia ($p=0.031$; $\rho=0.394$). A positive correlation was found between LDL and proteinuria ($p=0.043$; $\rho=0.385$) and between TG and CRP ($p=0.001$; $\rho=0.575$). TG were also positively correlated with prednisolone daily dose ($p=0.035$; $\rho=0.394$). Mean LDL was higher in anti-Sm positive patients ($p=0.022$). No differences were found regarding anti-phospholipids antibodies. Nephritic lupus patients had worse lipid metabolism, but this did not reach statistical significance.

Conclusion: In out cohort, increased expression of TC, LDL and TG is associated with disease activity in SLE. As expected, higher doses of prednisolone also correlated with lipid metabolism.

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AB0984 BIOLOGICAL THERAPIES IN JUVENILE IDIOPATHIC ARTHRITIS: ARE THERE ANY DIFFERENCES BETWEEN CATEGORIES?

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