AB0942

RADIOLOGICAL SACROILIITIS AFTER 18 YEARS OF FOLLOW-UP IN THE POPULATION-BASED NORDIC JUVENILE IDIOPATHIC ARTHRITIS (JIA) COHORT

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Background: A challenge with the present classification of JIA is the evolution of the disease over time. One category that is especially difficult to classify is enthesitis-related JIA (ERA).

Objectives: To longitudinally study radiologically diagnosed sacroiliitis (SI) developed during the first 18 years in an aim to gain knowledge about classification challenges posed by the proposed, new classification (Martini A. et al. J Rheumatol. 2018 Oct; Epub ahead of print).

Methods: 510 consecutive cases of JIA with disease onset 1997 to 2000 were prospectively included in a Nordic, longitudinal, close to population-based 18-year follow-up study, and 434 (85%) had at least two follow-up visits during disease course. At the 18-year follow-up visit; 329 (76%) attended a clinical visit, and 105 (24%) a telephone interview. The follow-up period was 17.5 ± 1.7 years (mean ± SD) after onset. Mean age of the study participants was 24.0 ± 4.4 years. Clinical data, collected at one, eight and 18 years after disease onset, were evaluated regarding variables for enthesitis/spondylitis-related arthritis compared to the other JIA categories.

Results: In 376 participants evaluated for SI, radiology was performed on clinical suspicion, 26 (16 males, 10 females) developed radiologically verified sacroiliitis (rad-SI) during the first 18 years of disease. Age at onset was significantly higher in this group compared to the other participants, median 9.9 (IQR 6.4-12.0) vs. 5.6 (IQR 2.6-9.5) years, (p=0.001). Only 3/26 had rad-SI at eight-year follow-up.

Using the ILAR criteria 12/26 with rad-SI were classified as ERA after median 7 (IQR 6-10.2) months, 1/26 as juvenile psoriatic arthritis, 5/26 as undifferentiated JIA because of psoriasis-related variables, the remaining had oligo- or polyarticular arthritis. At 18-year follow-up, 18/26 fulfilled criteria for ERA, 2/26 juvenile psoriatic arthritis, and 3/26 the undifferentiated category because of psoriasis-related variables, 3/26 had polyarticular RF negative or oligoarticular extended disease. Enthesitis, inflammatory back pain and SI joint tenderness were more common with rad-SI (p<0.001; p<0.001; p<0.001), also HLA-B27, 18/26 vs. 62/351 (p<0.001), but not 1st degree heredity for ankylosing spondylitis. Uveitis developed in 10/26 (38.5%). Two participants developed IBD.

Conclusion: The majority of variables involved in the new proposed classification of enthesitis/spondylitis-related JIA were significantly more common during the first 18 years of disease in those that developed rad-SI. Psoriasis-related variables, no longer exclusion criteria for ERA, should be further evaluated as possible inclusion criteria, as well as higher age at onset.

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AB0943

CLINICAL AND LABORATORY PRESENTATION OF JUVENILE SJÖGREN'S SYNDROME IN A COHORT OF 30 PATIENTS

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Background: Sjögren's Syndrome (SS) is a chronic autoimmune disease, rare in childhood and adolescence, characterized mainly by xerostomia

and xerophthalmia, which compromises exocrine glands causing inflammatory response that leads to glandular hyposecretion. SS is probably underdiagnosed in pediatric range due to differences in presentation regarding adults with this condition, causing low recognition of the disease in children.

Objectives: The authors describe demographic, clinical, laboratory and therapeutic profiles of a cohort of children with primary SS attended at a university center.

Methods: Retrospective analysis of 30 selected patients' medical records between 2005 and 2017 allowed collection of various data. Laboratory and additional investigations included documentation of ocular dryness (Schirmer test, Rose-Bengal stain); evidence of parotid involvement (scintiscan, sialometry); and histological evidence of lymphocytic infiltration of the minor salivary glands or other organs.

Results: 30 patients diagnosed with juvenile SS were selected: 22 girls (73%) and 8 boys (27%) with an average age of 11 years. The clinical characteristics were: parotid enlargement as the initial manifestation of the disease in 11 patients (37%), and recurrent episode in 4 patients (13.3%). 12 patients (40%) had xerostomia and 17 (57%) xerophthalmia. One patient (3.3%) presented with leukocytoclastic vasculitis in lower limbs as first manifestation of the disease in association to recurrent parotid swelling. Two (6.6%) patients presented neurological symptoms (1 peripheral sensory neuropathy and 1 with dysautonomic manifestations). Positive Schirmer test was observed in 11 patients (37%), Rose Bengal stain in 10 (33%). 76% of patients had abnormal salivary glands scintigraphy. In 8 patients (30%) the salivary gland biopsy revealed compatible with Sjögren syndrome. 9 patients (30%) presented positive RF, 14 patients (46%) anti-Ro/SSA, 10 patients (33%) anti-La/SSB, 28 patients (93%) had +ve ANA. 50% received glucocorticoid. Hydroxychloroquine was the drug most often used in 25 patients (83%), followed by methotrexate in 12 patients (40%), azathioprine in 4 patients (13%) and cyclophosphamide in 3 patients (10%). Only one patient required the use of human immunoglobulin and one leflunomide (3.3%). Two patients (6.6%) received rituximab.

Conclusion: The present study demonstrated the demographic, clinical aspects and laboratory and treatment in a series of patients with primary juvenile Sjögren's syndrome, a relatively rare condition, presenting an overview of this population in our hospital.

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AB0944

SJÖGREN'S SYNDROME IN CHILDREN: A CASE SERIES

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Background: The symptoms of pediatric Sjögren's syndrome (SS) are different than in adults. There are currently no validated pediatric diagnostic criteria or treatment guidelines for SS. In most cases adult criteria are used, but they apply poorly to children.

Objectives: To present pediatric patients with primary SS who were treated at University Children's Hospital (UCH) Ljubljana in the past 10 years.

Methods: Eight children with primary SS were identified. Demographic data, clinical and laboratory findings and therapy were analysed by retrospective review of medical records at UCH Ljubljana.

Results: Six girls and 2 boys were evaluated. The mean age at disease onset was 12.3 years (range 6.5 – 17) and mean age at diagnosis was 13.8 years (range 7.5 – 17.5). The mean follow-up duration was 2.8 years (range 0.5 – 8.5). Four patients presented with recurrent bilateral parotitis, two with rash, one with arthralgia and fatigue and one with acute central nervous system vasculitis. The latter patient presented with rheumatic fever at the same time. During disease course arthritis and/or