AB0751 JUVENILE UVEİTİS: THE 3RD LEVEL EXPERIENCE

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Background: Uveitis is the inflammation of the uveal components of the eye (iris, choroid and retina) and is the third leading cause of blindness in the world. It is less common in children than adults. Young children are usually diagnosed late, as they cannot express their complaints.

Objectives: With this work; We aimed to contribute to the literature by determining the underlying primary diagnosis and evaluating the treatment response in patients who were followed up in our clinic with a diagnosis of non-infectious uveitis, mostly juvenile idiopathic arthritis.

Methods: 93 patients under 18 years of age who were followed up with a diagnosis of non-infectious uveitis were included in the study. The data of the patients were scanned retrospectively. According to the last biomicroscopic examinations, the patients were classified as remission, inactive period and active disease. Clinical characteristics, laboratory findings, family history, drugs used during the disease and duration of treatment were examined.

Results: The male to female ratio was 1,06 (48/45). The mean age at diagnosis of childhood uveitis was found to be 10±4.02. 49 Patients (52.6%) were diagnosed with idiopathic uveitis, 33 (33.5%) with juvenile idiopathic arthritis, 11 (11.7%) with Behçet's, 2 (2.2%) with FMF. Juvenile idiopathic arthritis was the most common systemic disease in the patient group. The mean age of patients with uveitis to be diagnosed with rheumatological disease was 10.1±5.32. ANA test was positive in 30 (32.2%) patients. 23 (69.7%) of juvenile idiopathic arthritis patients were oligoarticular type. Anterior uveitis was present in 37 (39.8%) of the patients. Anterior uveitis was most common in the juvenile idiopathic arthritis cases. Of the patients with uveitis, 24 (25.8%) were intermediate, 11 (11.8%) were posterior, 21 (22.6%) were panuveitis. Anti-TNF α agents were administered to patients with unresponsiveness to immunosuppressive therapy and high risk of vision loss. Most used anti-TNF α agents were infliximab and adalimumab. Glaucoma, cataract, and posterior synechiae were common complications in the patients during follow-up.

Conclusion: The most common systemic disease that cause noninfectious uveitis in children is juvenile idiopathic arthritis. In juvenile idiopathic arthritis, the most common involvement of the eye was the anterior segment, and it was thought that the location of the eye involvement could be useful in determining the underlying systemic disease. It is important to ensure regular eye examination in patients with rheumatologic symptoms.

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AB0752 CHRONIC RECURRENT MULTIFOCAL OSTEOMYELITIS: A SINGLE-CENTER CASE SERIES

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Background: Chronic recurrent multifocal osteomyelitis (CRMO) is a chronic autoinflammatory disease that primarily affects the skeleton of children and adolescents in the absence of an infectious etiology[1]. CRMO lesions presentation varies widely ranging from episodic bone pain to growth disturbance; lytic and sclerotic bone lesions can be found on X-ray, however magnetic resonance imaging is very useful for evaluating the extent of disease and follow-up [2].

Objectives: To report demographic, clinical and laboratory characteristics and response to therapy in a single center Italian cohort of CRMO patients.

Methods: We reviewed retrospectively the clinical records of seven patients affected by CRMO diagnosed between 2008 and 2019 at rheumatologic service of South Tyrol region in Italy.

Results: We identified 7 patients with CRMO, 4 were female patients, median age of onset of symptoms was seven years. Median delay in diagnosis was seven months; all patients had bone pain as initial symptom and 6 patients presented joint swelling. Median number of lesions was two; all patients had a recurrent pattern, 4 patient had multifocal distribution and 5 had appendicular skeleton involvement. None of our patients had antinuclear antibodies or HLA-B27 positivity. Mean erythrocyte sedimentation rate was 52mm/h and C-reactive protein 1,96mg/dL. Regarding other organs involvement one patient presented palmo-plantar pustulosis, one psoriatic arthritis and one psoriasis guttata; three patient referred familiarity for psoriasis. Histopathological diagnosis was confirmed in 3 patients. NSAIDs were used in six patients, methotrexate in two patients and bisphosphonates in 6 patients. Among biological DMARDs Adalimumab was used in two patients, one patient was treated subsequently with Secukinumab and Ustekinumab. 3 patients achieved remission (Table 1).

Table 1. Demographic, clinical and laboratory characteristics of patients with chronic recurrent multifocal osteomyelitis.

	Total (n=7)
Demographics	
Female (%)	4 (57)
Age at disease onset, years, median (range)	7 (6-24)
Delay in diagnosis, months, median (range)	7 (2-14)
Follow-up, months, median (range)	69 (11-151)
Initial symptoms	
Bone pain, n (%)	7 (100)
Swelling, n (%)	5 (71)
Limp, n (%)	3 (43)
Fever, n (%)	1 (14)
Clinical features	
Number of lesions, median (range)	2 (1-10)
Axial skeleton involvement, n (%)	2 (29)
Appendicular skeleton involvement, n (%)	5 (71)
Distribution of involvement	
Femur, n (%)	3 (43)
Radius/ulna, n (%)	2 (29)
Humerus, n (%)	1 (14)
Tibia/fibula, n (%)	5 (5)
Tarsal bones, n (%)	1 (14)
Others, n (%)	3 (43)
Laboratory tests	
Hemoglobin, g/dL, mean $\pm$ SD	13.1 $\pm$ 0.6
Leukocytes $\times 10^3/\text{mm}^3$, mean $\pm$ SD	8.48 $\pm$ 1.7
Platelets $\times 10^3/\text{mm}^3$, mean $\pm$ SD	384 $\pm$ 130
Erythrocyte sedimentation rate mm/h, mean $\pm$ SD	52 $\pm$ 37
C-reactive protein, mg/dL, mean $\pm$ SD	1.96 $\pm$ 2.35
Positive antinuclear antibodies, n (%)	0 (100)
HLA-B27 positive, n (%)	0 (100)
Comorbidities	
Any type of autoimmunity, n (%)	2 (29)
Others, n (%)	3 (43)
Histological confirmation, n (%)	3 (43)
Treatment	
NSAIDs, n (%)	6 (86)
Glucocorticoids, n (%)	0 (0)
Bisphosphonates, n (%)	6 (86)
TNF blocking agents, n (%)	2 (29)
Biological DMARDs, n (%)	1 (4)
Methotrexate, n (%)	2 (29)
Antibiotics, n (%)	3 (43)
Reached remission, n (%)	3 (43)

Conclusion: CRMO remains a rare diagnosis. From our experience most of the patients had delay in diagnosis, laboratory tests showed mild elevation of inflammatory parameters, was found association with psoriatic arthritis and psoriasis, remission was achieved with combination of bisphosphonate and immunosuppressive therapy in less than half of the cohort

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AB0753 ASSESSMENT OF SLEEP PROBLEMS IN PATIENTS WITH KAWASAKI DISEASE: A SURVEY-BASED STUDY

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Background: Kawasaki disease (KD) is a type of vasculitis that affects mainly medium and small arteries, especially the coronary arteries (1). It mainly affects children under the age of five (2). Sleep disturbances are more prevalent among medically ill children and adults and may have a negative impact on their health related quality of life (3, 4). To the best of our knowledge, sleep problems have not previously studied in KD patients.

Objectives: This study aimed to investigate sleep problems, sleep disturbances and the relation between sleep problems and clinical manifestations among patients with KD.

Methods: A cross-sectional study was performed using a written survey. Participants were invited through social media groups including patients and parents of children with KD. The survey included questions about demographic data, KD manifestations, disease duration and treatment received for management of the disease. Children's Sleep Habits Questionnaire (33-item version) was also included in the survey (5).

Results: There was a total of 262 survey participants (130 KD and 132 age- and sex matched healthy controls). There was no significant difference in the frequency of clinically significant sleep disturbance between patients with KD and healthy controls. However, the total sleep score was significantly higher in KD patients than healthy controls ($p < .001$). Total sleep duration was significantly higher in patients with KD (4.94 ± 1.88 vs 3.74 ± 1.14 hr, $p < .001$). Additionally KD patients had significantly higher scores regarding sleep onset delay ($p = .001$), night wakings ($p < .001$), parasomnias ($p < .001$) and sleep disordered breathing ($p < .001$) when compared to healthy controls. KD Patients who manifested with chronic heart disease or lethargy had significantly higher sleep score than patients without ($p = .015$, $.039$ respectively).

Conclusion: This study showed that patients with KD had more sleep disturbance than their healthy peers. The presence of either chronic heart disease or lethargy is associated with poor sleep quality. This study highlights the need for assessment of sleep disorders in KD patients.

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AB0754

THE INFLUENCE OF PHYSICAL ACTIVITY ON BODY COMPOSITION AND SELF-ESTEEM IN ADULTS WITH JUVENILE IDIOPATHIC ARTHRITIS

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Background: With improvement of treatment, long-term factors influencing quality of life have to be addressed in the care of juvenile idiopathic arthritis (JIA) patients.

Objectives: To compare body composition, level of physical activity and self-esteem of adult patients with JIA with control group (CG).

Methods: The levels of physical activity were analyzed using the International Physical Activity Questionnaire (IPAQ) short version. The body composition data were collected using the Omrom HBF 514 C vertical electric bioimpedance device. The Rosenberg Self-Esteem Scale was used to assess positive and negative attitude and feelings about themselves, where the lower the score, the higher the individual's self-esteem. For the statistical analysis of the data, the IBM SPSS software was used with a significance index of $p < 0.05$.

Results: We included 81 individuals >18 years, 38 with JIA and 43 healthy people. The female sex was the largest participant in this research, being. There was no significant difference regarding sex [female =28 (73.7%) in JIA and 26 (60.5%) in CG] and age of the groups (29.39 ± 7.6 vs. 31.1 ± 3.1 , $p = 0.18$). The CG showed greater height and body weight (163.5 ± 9.8 vs. 169.6 ± 8.0 , $p < 0.001$; 63.07 ± 16.65 vs. 69.33 ± 6.88 , $p = 0.003$, respectively), but no difference in Body Mass Index (BMI) was observed (24.32 ± 4.9 vs. 24.12 ± 2.09 , $p = 0.8$). JIA had a higher percentage of total fat mass (31.65 ± 10.20 vs. 28.47 ± 4.9 , $p = 0.07$), while the CG had a higher percentage of total muscle mass (29.7 ± 6.5 vs. 35.86 ± 6.05 , $p < 0.001$). The self-esteem of JIA patients was lower than in the CG (21 ± 3.9 vs. 16.26 ± 4.3 , $p < 0.001$). In the IPAQ classifications, JIA patients were classified as less physically active than the CG (2.63% vs. 20.93%, $p = 0.022$), less irregularly active (5.26% vs. 23.25, $p = 0.023$) and a higher level of sedentary lifestyle (65.78% vs. 25.58%, $p < 0.001$). A higher percentage of muscle mass was associated with greater self-esteem ($r = -0.3$; $p = 0.01$).

Conclusion: The low practice of physical activities by patients with JIA seems to directly influence both, their body composition and their self-esteem. These patients should be encouraged to increase physical activity from an early age by a multiprofessional healthcare team to seek improvement their quality of life.

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AB0755

THE FIRST CASE OF AN AUTOLOGOUS STEM CELL TRANSPLANTATION FOR AN EXTREMELY REFRACTORY PROGRESSIVE SYSTEMIC JUVENILE IDIOPATHIC ARTHRITIS PATIENT IN BRASÍLIA, BRAZIL

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Background: Systemic juvenile idiopathic arthritis (sJIA) is one of the most perplexing diseases of childhood, with a wide range of presentation features and severity. Advances in treatment have improved the outcome, such as the use of modulators of proinflammatory cytokines and their receptors, but in a considerable number of children, especially in polyarticular or sJIA, refractoriness tends to be progressive. As of late, extreme cases have required Autologous Stem Cell Transplantation (ASCT) as a last resort.

Objectives: To describe an extremely refractory clinical evolution in a sJIA patient, the first one to be submitted to an ASCT for an autoinflammatory condition in Brasília, Brazil's capital.

Methods: Case Report

The patient was classified according to the Pediatric International League of Associations for Rheumatology for sJIA.

I.T.S.M, female, 15-years-old, was diagnosed with sJIA at 6 with the onset of polyarthritis, daily fever for 6 weeks, rash and enlargement of liver and spleen. In the initial investigation infectious and neoplastic causes were ruled out.

The use of prednisone and methotrexate lead to a preliminary effective clinical control, however after an year, there was clinical deterioration and etanercept was associated, leading to a short term improvement. At that time, in addition to anti-tumour necrosis factor medications, no other biological drugs were available in Brazil.

Despite some brief periods of clinical improvement, after different regimes, the patient manifested recurrent flares. Since the diagnosis she has already been treated with:

- [1] Methotrexate (2011 to 2014*, 2016 to 2020). *In 2015, it was replaced by Leflunomide (up to May 2016, discontinued due to intolerance);
- [2] Etanercept (2012);
- [3] Cyclosporine (August to December 2012);
- [4] Adalimumab (2013);
- [5] Tocilizumab (May 2014 to January 2015);
- [6] Canakinumab (March 2015 to April 2016);
- [7] Mycophenolate Mofetil (February 2016, discontinued in May 2017 due to start of Cyclophosphamide);
- [8] Rituximab (May 2016 to April 2017);
- [9] Cyclophosphamide at a dose of 2 grams /m² as well as Filgrastim for mobilization and collection of stem cells (May 2017, in a attempt of ASCT), but the parents were unable to reach a mutual agreement to consent to the continuation of this therapy;
- [10] Cyclophosphamide (500mg /m²) in a mensal scheme application for nine consecutive months since June 2017, once was observed a clinical improvement after the intensive immunosuppression previous to ASCT attempt;
- [11] Several courses of methylprednisolone and human immunoglobulin since the initial diagnosis;
- [12] Abatacept (November 2019 to October 2020);
- [13] ASCT was suggested again in 2020, this time with the consent of all family members.

Results: An intensive immunosuppression [Cyclophosphamide and anti-thymocyte globulin (ATG)] followed by ASCT (October 2020) resulted in apparent sustained remission. The patient's evolution since the transplant has been optimistic. She is currently on a low dose prednisone prescription. To ensure a less profound depletion of T cells, a better control of systemic disease and antimicrobial and antiviral prophylaxis after transplantation, slow tapering of corticosteroids were performed.

Conclusion: The goal of hematopoietic stem cell transplantation in patients with autoimmune disease is to reprogram the immune system with the eradication of autoreactive cells, renew the population of regulatory T cells and restore the diversity of T cell receptor function.

ASCT has been used in some refractory children with sJIA as well as hematologic malignancy and some progressive autoimmune diseases. However it is associated with significant morbidity and mortality, due to prolonged and severe depression of T cell immunity.

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