Musculoskeletal, hematologic, enterohepatic, and ocular findings as initial findings were significantly high in group 1 ($p < 0.05$). Pediatric Rheumatology, general pediatrics and pediatric neurology are the three clinics which the assay was requested most common. The patients who were examined in pediatric rheumatology, pediatric hematology-oncology, pediatric gastroenterology departments had a higher incidence of autoimmune disease ($p < 0.05$). In patients with autoimmune disease, $\geq 1/1000$ ANA titer and homogenous staining were frequent ($p < 0.05$).

Conclusion: In our center, which is a tertiary health center, the autoimmune disease was not detected in most of the patients whose ANA assay was positive. The proportion of patients with autoimmune disease was low in most of the commonly requested departments. The ANA assay should be requested after the patient's clinical findings are evaluated in detail.

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AB1044

CYTOKINE PROFILE OF MACROPHAGE ACTIVATION SYNDROME ASSOCIATED WITH KAWASAKI DISEASE

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Background: Macrophage activation syndrome (MAS) is a severe, potentially life-threatening complication of childhood systemic inflammatory disorders. MAS occurs most often in children with systemic juvenile idiopathic arthritis and less commonly in children with Kawasaki disease (KD).

Objectives: Our study aimed to assess the kinetics of cytokine release and compare the accuracy of serum biomarkers for diagnosis of MAS, including neopterin, IL-18, IL-6 and soluble TNF receptor type I (sTNFR-I) and sTNFR-II levels, we analysed these levels in patients with KD, including those with MAS, and compared them to the clinical features of KD and MAS.

Methods: Serum neopterin, interleukin (IL)-18, IL-6 and soluble tumour necrosis factor receptor type I (sTNFR-I) and sTNFR-II levels were determined using enzyme-linked immunosorbent assay in 78 patients with KD, including five with MAS. Results were compared to the clinical features of MAS.

Results: Serum neopterin, IL-18, sTNFR-II levels and sTNFR-II/I ratio were significantly elevated in KD patients with MAS compared to those in the acute phase. Receiver operating characteristic curve analysis revealed areas under the curve and cutoff values of neopterin, IL-18, sTNFR-II levels and sTNFR-II/I ratio were 0.9750/30.0 nmol/L, 0.9813/1165 ng/mL, 0.9969/16,600 pg/mL and 0.9875/4.475, respectively. Serum sTNFR-II levels correlated positively with disease activity.

Conclusion: These findings indicate that interferon (IFN)- γ and tumour necrosis factor- α (TNF- α) are closely associated with the pathogenesis of MAS associated with KD. Serum sTNFR-II levels might be a useful marker to diagnose the transition to MAS.

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AB1045

MULTICENTRIC CARPOTARSAL OSTEOLYSIS (MCTO) IN PRACTICE OF PEDIATRIC RHEUMATOLOGIST: DIFFERENTIAL DIAGNOSIS WITH JUVENILE IDIOPATHIC ARTHRITIS

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Background: Rare genetic pathologies involving the musculoskeletal system can be erroneously misdiagnosed as juvenile idiopathic arthritis (JIA).

Idiopathic multicentric carpotarsal osteolysis (MCTO) is a congenital disease, linked to MAFB-gene mutation, described in 2012. MCTO is characterized by progressing osteolysis, mostly of carpal and tarsal bones, leading to articular deformities and functional impairment, with or without nephropathy. The incidence has not yet been established. And there is no available treatment as of today

Objectives: To share the experience of MCTO identification in pediatric rheumatologist practice at federal center.

Methods: Totally 2 MCTO cases were identified in boys 6 and 13 years old during the period 2009 – 2018. Standard rheumatological examination was performed. Genetic testing (Sanger sequencing) in 1 patient identified MAFB-gene mutation.

Results: Both patients had pain and swelling in wrist and ankle joints, flexion contractures in elbow joints, and gait abnormalities. Disease duration at the time of MCTO verification was 4 and 11 years. Both patients went through erroneous polyarticular JIA diagnoses. During the follow up patients' ESR and CRP were normal, HLA B27 - negative, ANA and RF - negative. No visceral pathology, including kidneys, was found. Therapy included NSAIDs, glucocorticosteroids – in one case, MTX and Tocilizumab gave no effect in the second patient. Radiographic findings were severe osteolysis of wrist and feet bones. JIA diagnosis was ruled out and MCTO suspected, and later confirmed in both patients by a geneticist. Heterozygous MAFB gene c.206C>T (p.Ser69Leu) mutation was detected in one patient.

Conclusion: Specific phenotypical features and patient's articular status, osteolysis of the carpal and tarsal bones, absence of laboratory activity signs and of response to antirheumatic therapy is a sound motive to continue diagnostic elaboration in order not to miss rare genetically-linked conditions of the musculoskeletal system.

Disclosure of Interests: None declared

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AB1046

PHYSICAL ACTIVITY ASSESSMENT IN CHILDREN WITH JUVENILE IDIOPATHIC ARTHRITIS COMPARED TO CONTROLS

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Background: Physical activity (PA), known to maintain optimal metabolic function and normal development could be impaired during Juvenile Idiopathic Arthritis (JIA).

Objectives: The aim of our study was to assess PA in children and adolescents with JIA compared to healthy peers using the physical activity questionnaire for children (cPAQ) and adolescents (aPAQ).

Methods: This is a cross-sectional study of measured level of PA in children and adolescents with JIA, compared to age and gender-matched healthy Tunisian schoolchildren. PA was estimated by cPAQ and aPAQ filled by the patient group and the reference group. If the child is unable or unsure to answer the questions we have helped with the parents' response. The PAQ scores 2 as "low activity," >2 and ≤ 3 as "moderate activity," and >3 as "high to vigorous activity".

Results: A total of 55 patients (38 boys and 17 girls) with JIA and 60 healthy control schoolchildren were included. No significant difference in demographic background was found between the two groups. The mean age was 8.5 ± 4.12 years in the JIA group and 9.2 ± 3.51 years in the control group. Thirty-one patients (53%) had persistent oligoarticular JIA, 15 (27%) had polyarticular JIA, 5 patients (9%) had systemic JIA, and 4 (7%) had enthesitis-related arthritis. The median disease duration was 3.2 ± 2.8 years. The mean cPAQ was 2.101 ± 0.722 in the JIA group and 4.112 ± 0.644 in the control group ($p=0.0001$). Children and adolescents with JIA had a significantly lower levels of PA compared with their healthy peers as assessed by cPAQ/aPAQ ($p=0.012$). The time spent in each cPAQ/aPAQ item was significantly lower in the JIA group compared to the healthy group: sports activities (3.4 ± 0.5 versus 4.7 ± 1.4 hours/week, $p=0.002$), leisure time activities (2.2 ± 0.3 versus 6.2 ± 1.3 hours/week, $p=0.001$), activities at school (1.1 ± 0.3 versus 2.1 ± 0.5 hours/week, $p=0.001$), and after school activities (0.5 ± 0.5 versus 2.5 ± 0.8 hours/week, $p=0.001$). Seventy six percent of the JIA group spent the day on the two lowest PA categories: sleeping and sitting, which was significantly higher compared with the reference group ($p=0.001$ and $p=0.055$, respectively).

Conclusion: In our study, children and adolescents with JIA were less physically active than the healthy peers as assessed by the PAQ. More objective methods are needed to better evaluate and quantify the PA.

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