AB0739

MULTISYSTEM INFLAMMATORY SYNDROME ASSOCIATED WITH SARS-COV-2 INFECTION AND E.COLI SEPSIS: THE POTENTIAL ROLE OF PROCALCITONIN AS A RAPID DIAGNOSTIC BIOMARKER TO DISTINGUISH TWO DIFFERENT PHASES OF SYSTEMIC INFLAMMATORY RESPONSE SYNDROME

M. Gilio¹, S. B. Morella¹, F. Picaro¹, C. Acerno¹, D. Palazzo¹, A. Erezanu¹, M. Frontuto¹, G. Mastroberti¹, G. De Stefano¹. ¹San Carlo Hospital, Infectious Diseases, Potenza, Italy

Background: Severe acute respiratory syndrome coronavirus 2 (SARS-CoV-2) infection is typically very mild and often asymptomatic in children. A complication is the rare multisystem inflammatory syndrome in children (MIS-C) associated with COVID-19, presenting 4-6 weeks after infection as high fever, organ dysfunction, and strongly elevated markers of inflammation. The pathogenesis is unclear but has overlapping features with Kawasaki disease suggestive of vasculitis and a likely autoimmune etiology.

Objectives: We report a case of multisystem inflammatory syndrome in children (MIS-C) in patient with SARS-CoV-2 infection and Enteropathogenic Escherichia coli (EPEC) sepsis due to acute enteritis, observed at end of December 2020 to a tertiary-care center (San Carlo Hospital), in Basilicata region (Italy).

Methods: This healthy 12-year-old male patient was tested positive for severe acute respiratory syndrome coronavirus 2 (SARS-CoV-2) infection. Clinical presentations was characterized by fever, abdominal pain, gastrointestinal complaints and evanescent rash. Laboratory values were remarkable for high levels of procalcitonin, C-reactive protein (CRP), D-dimers, B-type natriuretic peptide (BNP), and troponin. He also had low albumin levels. Autoantibodies tests were negative. Chest tomography showed ground-glass opacities in less than 25% of the lungs, small bilateral pleural effusion and increased cardiac area; abdominal tomography showed enlargement of the lymphnodes and ascites. Evaluation for other infectious etiologies showed molecular test positivity on fecal samples for EPEC E. coli. He received broad spectrum intravenous antibiotics (macrolids and quinolones and then carbapenems). On the seventh day the enteritis resolved and procalcitonin normalized, however he continued to have lymphopenia, thrombocytopenia, hypoalbuminemia, elevated levels of CRP, D-dimers, ferritin, troponin, and increased BNP. On the ninth day he was feverish again and developed severe cardiac and respiratory failure requiring advanced respiratory support and admission to the intensive care unit. He received IVIG (intravenous immunoglobulin at 2g/Kg, glucocorticoids (Methylprednisolone 1mg/kg) and enoxaparin.

Results: The patient was discharged asymptomatic at home after 28 days of hospital stay.

Conclusion: We observed multisystem inflammatory syndrome in children (MIS-C) in a previously healthy patient with SARS-CoV-2 infection and E.coli sepsis, who became critically ill with multisystem involvement. In this case viral and bacterial infections could be considered as a double hit for the etiopathogenesis of MIS-C. The trend of procalcitonin was better than C-reactive protein for differentiating bacterial from non-bacterial phase of systemic inflammatory response syndrome (SIRS) in this critically ill child. Although the accuracy of both tests is moderate. Diagnostic accuracy could be enhanced by combining these tests with bedside clinical judgment.

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AB0740

IMPACT OF DIAGNOSIS DELAY ON DISEASE PARAMETERS DURING JUVENILE IDIOPATHIC ARTHRITIS

H. Bettaieb¹, H. Ferjani¹, K. Maatallah¹, D. Kaffel¹, W. Hamdi¹. ¹Mohamed Kassab Institute of Orthopedics, Rheumatology, Tunis, Tunisia

Background: Juvenile idiopathic arthritis (JIA) is the most common type of arthritis in childhood (1). Prompt diagnosis is mandatory to avoid joint destruction and growth abnormalities. However, it's often misdiagnosed by pediatricians and general practitioners leading to longer diagnosis delay (2).

Objectives: The aim of this study was to evaluate the lag time between JIA symptoms onset and diagnosis and its impact on disease activity and bone loss.

Methods: A retrospective monocentric study was carried out on JIA patients (ILAR criteria). Diagnosis delay was collected from the patient's medical files. Disease activity at JIA diagnosis was evaluated by JADAS10 (Juvenile Arthritis Disease Activity Score) in poly and oligoarticular subtypes and by BASDAI (Bath Ankylosing Spondylitis Disease Activity Index) in arthritis related enthesitis form. The data were analyzed using the SPSS statistical package. A p value < 0.05 was considered significant.

Results: We enrolled 48 JIA (31 male and 17 female) with a mean age at disease onset of 11.2 ± 3.8 years. The median disease duration was 84 months [2-408]. The median JIA diagnosis delay was 8 months [1-108]. The JIA subgroups were in decreasing order of frequency: Enthesitis-related Arthritis (n=32), Polyarticular RF- (n=4), Polyarticular RF+ (n=2), Oligoarticular (n=6), Systemic (n=2), Psoriatic Arthritis (n=1) and Undifferentiated (n=1).

At diagnosis, median ESR and CRP were 44 mm/hour [2-100] and 24 mg/l [2-86] respectively. Median JADAS10 score was 4 [0-21]. Median BASDAI score was 6.2 [2-9.4].

At follow-up, five patients (10.4%) had atlantoaxial subluxation and 17 had coxitis (43.8%).

At bone densitometry, 45% of patients had osteoporosis and 27.5% had osteopenia.

An agreement was assessed between a long diagnosis delay and the following parameters: male gender (p=0.04) and osteoporosis (p=0.018). A Significant positive correlation was found between delay in JIA diagnosis and BASDAI score (p=0.047, r=0.63). No association was found between JIA diagnosis delay and JADAS score (p=0.56). Neither ESR (p=0.19) nor CRP (p=0.42) was associated with JIA diagnosis delay.

Finally, no link was observed with the occurrence of hip arthritis (p=0.281) or atlantoaxial subluxation (p=0.137).

Conclusion: In this study, delay in diagnosis was associated with higher disease activity scores and bone loss. Our results suggest that early identification and treatment of JIA leads to improved outcomes as well as bone mass.

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AB0741

PREVALENCE AND ASSOCIATED FACTORS OF ATLANTO-AXIAL INSTABILITY IN PATIENTS WITH JUVENILE IDIOPATHIC ARTHRITIS

H. Bettaieb¹, H. Ferjani¹, K. Maatallah¹, H. Boussaa¹, D. Kaffel¹, W. Hamdi¹. ¹Mohamed Kassab Institute of Orthopedics, Rheumatology, Tunis, Tunisia

Background: Atlanto-axial instability (AAI) is a serious complication during Juvenile Idiopathic Arthritis (JIA). It can lead to severe neurological morbidity or mortality if left untreated (1).

Objectives: To determine the prevalence of AAI in patients with JIA and to identify factors associated with an increased risk of its occurrence.

Methods: A retrospective monocentric study was carried out on JIA patients (ILAR criteria). Data, including age at disease onset, JIA type, disease activity at AAI diagnosis and treatment were collected. Disease activity at JIA diagnosis was evaluated by JADAS10 (Juvenile Arthritis Disease Activity Score) in poly and oligoarticular subtypes and by BASDAI (Bath Ankylosing Spondylitis Disease Activity Index) in arthritis related enthesitis form. Standard radiographs of the cervical spine were analyzed. The data were analyzed using the SPSS statistical package. A p value < 0.05 was considered significant.

Results: We enrolled 48 JIA (31 male and 17 female) with a mean age at disease onset of 11.2 ± 3.8 years. The median disease duration was 84 months [2-408]. The median JIA diagnosis delay was 8 months [1-108]. The JIA subgroups were in decreasing order of frequency: Enthesitis-related Arthritis (n=32), Polyarticular RF- (n=4), Polyarticular RF+ (n=2), Oligoarticular (n=6), Systemic (n=2), Psoriatic Arthritis (n=1) and Undifferentiated (n=1).

At diagnosis, median ESR and CRP were 44 mm/hour [2-100] and 24 mg/l [2-86] respectively. Median JADAS10 score was 4 [0-21]. Median BASDAI score was 6.2 [2-9.4].

At follow-up, five patients (10.4%) had atlantoaxial subluxation and 17 had coxitis (43.8%).

At bone densitometry, 45% of patients had osteoporosis and 27.5% had osteopenia.

An agreement was assessed between a long diagnosis delay and the following parameters: male gender ($p=0.04$) and osteoporosis ($p=0.018$). A Significant positive correlation was found between delay in JIA diagnosis and BASDAI score ($p=0.047$, $r=0.63$). No association was found between JIA diagnosis delay and JADAS score ($p=0.56$). Neither ESR ($p=0.19$) nor CRP ($p=0.42$) was associated with JIA diagnosis delay.

Finally, no link was observed with the occurrence of hip arthritis ($p=0.281$) or atlantoaxial subluxation ($p=0.137$).

Conclusion: In our study, the prevalence of AAI was 10.4%. Prolonged corticosteroid use and elevated inflammatory markers were the major factors associated with an increased risk of upper cervical spine involvement. Hence, targeted treatments are required to prevent cervical spine instability.

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AB0742

PREVALENCE AND RISK FACTORS OF OSTEOPOROSIS IN PATIENTS WITH JUVENILE IDIOPATHIC ARTHRITIS

H. Bettaieb¹, H. Ferjani¹, K. Maatallah¹, H. Boussaa¹, D. Kaffel¹, W. Hamdi¹.

¹Mohamed Kassab Institute of Orthopedics, Rheumatology, Tunis, Tunisia

Background: Childhood rheumatic diseases are associated with reduced Bone mass and increased risk of fractures (1). Several factors may interact to determine osteoporosis other than direct bone detrimental effects of the disease or its treatment.

Objectives: In this work, we aimed to investigate the prevalence of bone loss in patients with JIA and to determine the relative factors associated with osteoporosis during this chronic disease.

Methods: A retrospective monocentric study was carried out on JIA patients (ILAR criteria). Dual-energy x-ray absorptiometry (DEXA) was used to determine bone status. Disease activity was evaluated by JADAS10 (Juvenile Arthritis Disease Activity Score) in poly and oligoarticular subtypes and by BASDAI (Bath Ankylosing Spondylitis Disease Activity Index) in arthritis related enthesitis form. Erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) were noted. The data were analyzed using the SPSS statistical package. A p value < 0.05 was considered significant.

Results: The sample included 40 JIA (25 male and 15 female) with a mean age at disease onset of 11.3 ± 3.6 years. The median disease duration was 90 months [7-408]. The median JIA diagnosis delay was 8 months [1-108]. The JIA subgroups were in decreasing order of frequency: Enthesitis-related Arthritis ($n=27$), Polyarticular RF- ($n=4$), Polyarticular RF+ ($n=1$), Oligoarticular ($n=4$), Systemic ($n=2$), Psoriatic Arthritis ($n=1$) and Undifferentiated ($n=1$). Median ESR and CRP were 29 mm/hour [2-98] and 14.5 mg/l [0-70] respectively. Median BASDAI score was 4.3 [1-9.7]. Median JADAS10 score was 1[1-21].

Overall, 45% of patients had osteoporosis, 27.5% had osteopenia, and 27.5 % had normal bone densitometry. None of the patients had a history of vertebral or peripheral fractures.

Thirty per cent of patients ($n=12$) were on long term corticosteroid therapy with a mean dose of 6.6 ± 2.8 mg/day. Only 12.5% ($n=5$) of them had a regular physical activity.

Osteoporosis was associated with age at JIA onset ($p=0.005$), disease duration ($p=0.001$), ESR ($p=0.08$), CRP ($p=0.04$), BASDAI score ($p=0.017$) and sedentarily ($p=0.026$). Osteopenia was only associated with corticosteroid therapy ($p=0.01$). Neither osteoporosis ($p=0.37$) nor Osteopenia ($p=0.25$) was associated with disease activity score.

Conclusion: In our study, osteoporosis was a common feature during JIA. A long term corticosteroid therapy and sedentarily seem to be correlated with more impaired bone abnormalities. Hence, targeted interventions are urgently required to preserve bone health during JIA.

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AB0743

SINGLE CENTER EXPERIENCE OF BIOLOGICAL THERAPY IN PATIENTS WITH JUVENILE SYSTEMIC SCLERODERMA

D. Alexseev¹, I. Nikishina¹, L. Ananieva¹, V.A. Nasonova Research Institute of Rheumatology, Pediatric, Moscow, Russian Federation

Background: Juvenile systemic sclerosis (jSSC) is a rare multisystem disease with a poor prognosis. The current therapy is imperfect. Biologics, as a modern efficient option for treatment of rheumatic diseases don't have official indication for jSSC, but it seems to be useful in severe cases, especially lung and heart damage.

Objectives: To analyze the existing experience of Biologics therapy in children with jSSC in single center in order to improve the understanding of the indications for use and the results of using these drugs.

Methods: The retrospective study include 9 patients with jSSC with severe course of disease, 7 of them were girls.

Results: Among this group of patients the age at disease onset was 9.1 years (min-max 1.1-15.1). Clinical manifestations: Raynaud's syndrome ($n=9$), ulcers ($n=3$), skin changes ($n=9$), joints ($n=8$), muscles ($n=4$), interstitial lungs disease ($n=6$), heart ($n=3$), esophagus ($n=5$), calcification ($n=3$). The main therapy prior to the use of biological agents: glucocorticoids (GC) ($n=9$), cyclophosphamide (CF) ($n=2$), hydroxychloroquine (HC) ($n=5$), therapy for the microcirculation improvement. Biologics were prescribed in the case of insufficient effectiveness of previously conducted therapy, primarily in relation to lung damage (interstitial disease) and heart (with reduced contractility, arrhythmia), as well as changes in the skin and musculoskeletal system. In addition, the possibility to reduce the dose of GC ($n=6$), withdrawal of CF ($n=2$) was taken into account. Our experience consist of follows: rituximab (RTM) ($n=7$) at a dose of 375 mg/m² interval 3-6 months, abatacept i.v.(ABT) ($n=2$) 10 mg/kg every 4 weeks, tocilizumab i.v.(TCZ) ($n=2$) 8 mg/kg. In all cases Biologics was prescribed not earlier than 1.5 year after disease onset (4.6 years in average). By present most patients continued to use biological therapy; of them - RTM was used for 2.2 (0.5-6.0) years, in 1 patient – was canceled after 6 years due to the loss of efficacy; ABT is used in 2 children for 0.5 and 3.6 years, TCZ - in 2 children for 0.3 (withdrawal) and 2.0 years (continues to use), TCZ was prescribed in 1 patient after 6 years of RTM use. In 1 patient TCZ was withdrawal due to an infusion reaction, and ABT was prescribed. As a result of the therapy, it has been found to improve habitus, quality of life and well-being, reduce skin tightening, improve joint function; no progression of changes in the lungs according to CT and functional tests (FVC, DLCO); improvement in cardiac lesions. It seems that disease improvement became notable at least 3-6 months after the Biologics start. In most patients there were no serious adverse events (AE). No significant infections have been reported. But we met an unexpected case of substantial increasing of neutrophil's count just after each TCZ infusions up to 21,000 and reversed by following infusions for a one year of treatment from 2 years of total duration of using.

Conclusion: Our data shown that Biologics could be efficient and well-tolerated option for severe and resistant to «traditional» therapy course of jSSC. Because this kind of condition is extremely rare further multicenter study is needed.

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AB0744

HOW JUVENILE IDIOPATHIC ARTHRITIS PATIENTS PERCEIVE THEIR ILLNESS?

K. Maatallah¹, M. Yasmine¹, H. Ferjani¹, W. Triki¹, D. Ben Ncib¹, D. Kaffel¹, W. Hamdi¹. ¹Kassab Institute of Orthopedics, Rheumatology, Tunis, Tunisia

Background: Juvenile idiopathic arthritis (JIA) represents the most common inflammatory rheumatism of childhood. Because of a prolonged course of active disease, many patients experience physical and psychosocial limitations. Illness perceptions involve the personal beliefs that patients have about their illness and may influence health behaviors considerably.

Objectives: The present study aimed to investigate the illness perception of patients with JIA.

Methods: We conducted a cross-sectional study including children with JIA (according to the International League of Associations for Rheumatology (ILAR)). We recorded sociodemographic data and the subtype of the JIA. To assess a child's perception of how arthritis affects their life, the Brief Illness Perception Questionnaire (IPQ) was completed by the parent (child age ≤ 10 years) or by