AB1031

ETANERCEPT-INDUCED LUPUS-LIKE SYNDROME IN THE BOY WITH IDIOPATHIC DERMATOMYOSITIS OR DELAYED MANIFESTATION OF SYSTEMIC LUPUS ERYTHEMATOSUS?

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Background: Treatment of juvenile onset rheumatic diseases (RD) is an ongoing problem because of the more aggressive disease course and more various clinical manifestations than adult-onset RD. Since the main clinical symptoms may overlap between well-known various RD, in some cases the path to diagnosis and the search for optimal therapy takes longer, especially in patients of pre-adolescence and adolescence.

Objectives: We described a rare case of one boy with RD who presented initially with severe dermatomyositis with sequential evolution to systemic lupus erythematosus.

Methods: Case report.

Results: A 8 yo male patient with normal physical, psychosocial and cognitive development experienced muscle weakness, periorbital erythema, Gottron symptom, dysphagia in September 2015 y, which developed after insolation. When he was admitted to hospital, increased levels of muscle-derived enzymes and positivity of ANA 1:640 were found. He was diagnosed as having juvenile dermatomyositis and was treated with prednisolone (Pr) for 3 years (IV courses repeated and per os constantly, max dose 1.5 mg/kg/day, min dose 0.5 mg/g/day), IVIG courses repeated and sequentially methotrexate, hydroxychloroquine, cyclosporine with short time of remission. Muscle weakness disappeared after the treatment, but in October 2015 y he experienced new eruptions on his face, heavy aphthosis stomatitis, lymphadenopathy, distal polyneuropathy, weight loss without strong effect after increase the dose of Pr, IVIG courses repeated, cyclosporine and added of abatacept during 6 months. Because the response to this treatment was low, etanercept was started in June 2016 y, but after two injections he had flare of disease: bright rash on the face, scalp and legs, hard aphthosis stomatitis, cheilitis, cognitive disorders, diffuse alopecia. He received IV course of Pr without significant stable effect. We reviewed diagnosis as systemic lupus erythematosus and changed therapy in November of 2016. Because he had steroid's osteoporosis with spondylopathy, this combination that made us initiate treatment with mycophenolate mofetil and repeating courses of rituximab (3 courses in total) without increased dose of Pr. Clinical features gradually improved and now he feel good and receive mycophenolate mofetil and Pr 5 mg/day.

Conclusion: This clinical observation can be considered as a course of overlap-syndrome with a successive phase change that could be triggered by anti-TNF therapy, or as an unusual course of SLE in a male patient with a predominance of myositis at onset. Treatment strategy, including of Pr, mycophenolate mofetil and rituximab in a young patient proved successful for treatment and seems to be justified in similar cases.

Disclosure of Interests: None declared

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AB1032

FACTORS TO AID IN EARLY DIAGNOSIS OF KIKUCHI'S DISEASE IN ASIAN CHILDREN

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Background: The diagnosis of Kikuchi's Disease (KD), also known as histiocytic necrotizing lymphadenitis, is often delayed as patients often undergo multiple investigations to exclude other causes of cervical lymphadenopathy, such as infection and malignancy¹.

Objectives: To describe the clinical features and treatment response of pediatric patients diagnosed with KD in our institution to aid early recognition.

Methods: A retrospective study was conducted on patients < 18 years old diagnosed with KD in National University Hospital, Singapore, from Jan 2006 – Dec 2018. Demographics, clinical characteristics and treatment information were collected.

Results: 14 patients were diagnosed with KD. All were Asian and majority were Chinese (64.2%). 57.1% were male and the age of presentation ranged between 2.8 to 17.9 years old. Fever duration at diagnosis ranged between 8 - 50 days. Median height of temperature was 39.8 (IQR: 39.4 – 40.0) deg C. This was frequently associated with neck pain and swelling (57.1%) and constitutional symptoms (71.4%) such as anorexia and weight loss. Other symptoms include abdominal symptoms of pain, vomiting or diarrhea (57.1%), rash (35.7%) and oral ulcers (28.6%).

All patients had cervical lymphadenopathy either in the anterior cervical triangle or both anterior and posterior triangles, but not in the posterior triangle alone. This was usually multiple (92.9%), measuring ≥ 2 cm in diameter (100%), tender (85.7%), and tended to be unilateral (64.3%); none had overlying warmth or skin changes. 14.3% had isolated hepatomegaly while 14.3% had hepatosplenomegaly.

Common lab abnormalities included cytopaenias (50% bicytopenia, 28.6% leucopenia, 7.1% anaemia), elevated inflammatory markers C-reactive protein (CRP) (range: 12 – 117 mg/L), erythrocyte sedimentation rate (ESR) (range 15 – 116 mm/h) and lactate dehydrogenase (range: 480-1514 U/L). Only 3/14 had mild transaminitis. Interestingly, almost half had low alkaline phosphatase (ALP). All patients had normal to high complements and were negative for anti-dsDNA antibodies. Ultrasound of the neck typically revealed non-specific findings of reactive lymphadenopathy while 1 showed suspicious features of thickened cortex and loss of fatty hilum. 10/14 patients underwent excision biopsy which confirmed the diagnosis of Kikuchi Disease on histology. 3/14 did not have excision biopsy as they responded promptly to non-steroidal anti-inflammatory drugs (NSAIDs), one declined excision biopsy.

All patients had received empiric antibiotics with no response. 64.3% required treatment with NSAIDs for an average of 2-4 weeks. Of the 4 patients who required steroid therapy, one was started at the diagnosis of KD, while the other 3 received steroids upon failure of NSAIDs. Response to either NSAIDs or prednisolone, if effective, was usually prompt, within 24 hours.

Conclusion: Prolonged high grade fever with multiple tender cervical lymphadenopathy ≥ 2 cm without warmth or redness and non-response to antibiotics, should raise concerns for KD. The presence of cytopaenia with moderately elevated inflammatory markers and LDH, coupled with low ALP, may be considered supporting biochemical features of KD. Prompt response to a trial of NSAIDs can help with the diagnosis of KD and potentially avoid excision biopsy of the lymph node.

More research needs to be done to determine a clinical and biochemical diagnostic criteria for Kikuchi's Disease so as to aid early diagnosis and potentially avoid invasive excision biopsy of the lymph nodes.

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AB1033

MEASUREMENT OF DISEASE ACTIVITY LEVEL OVER TIME IN CHILDREN WITH JUVENILE IDIOPATHIC ARTHRITIS UNDER BIOLOGIC TREATMENT, USING THE JUVENILE DISEASE ACTIVITY SCORE HUNGARIAN DATA FROM THE JUVENILE IDIOPATHIC ARTHRITIS REGISTRY OF THE NATIONAL INSTITUTE OF RHEUMATOLOGY AND PHYSIOTHERAPY

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Background: A wide variety of measures are currently available to monitor the disease course over time in children with JIA. The Juvenile Disease Activity Score JADAS71 version was validated for 71 joints, global score for the JADAS71 is: 0-101.

Objectives: To report the changes of disease activity in JIA patients under biological treatment at age 18, using the JADAS 71 score for the extended oligoarticular and seropositive/seronegative polyarticular form of JIA.

Methods: The Hungarian Biologics JIA Registry of National Institute of Rheumatology and Physiotherapy, Budapest is a longitudinal, unicenter, observational study, that has been maintained since 2002. JIA-ILAR criteria were used for the diagnosis. Patients with polyarticular course, who failed to respond or did not tolerate methotrexate and were treated with biological agents at the Paediatric Rheumatologic Centre of the National Institute of Rheumatology and Physiotherapy, Budapest, Hungary from 2002 to 25th January, 2019, were enrolled in an open label observational study and have been monitored prospectively in the registry. At baseline patients and disease characteristics were registered such as, patients'

history, demographic characteristics, and previous DMARDs treatments. Adverse events(AEs), reason for the discontinuation of the biologic treatment, or switch to another biological treatment were documented. The study was approved by the Institutional Review Board. Written informed consent was obtained from parents of the patients and the study was conducted in accordance with the Declaration of Helsinki.

Results: In this study 64, 18-year-old Caucasian JIA patients were evaluated from the day of entrance into the registry, who had JADAS 71 results at age 18, from the 347 patients treated with biologics at the submission.

Demographic data and disease related variables by descriptive statistics:
Gender: Female: N(%): 49(77%).

Subtypes of JIA: Extended oligoarticular form: 11 patients, Polyarticular form: 53 patients (5 RF positive,48 negative)

Median age: 8 (range:4-17) years.

Mean disease duration: 3352 days.

Mean time on biological treatment:1030 days.

The first JADAS 71 summation at the entry into the registry: Median: 18,65 (range:14,4-55,4).

The age 18 JADAS71 summation: Median:3,75 (range: 2,1-38).

All patients were under biologic treatment (etanercept: 56, adalimumab: 29, tocilizumab: 12, abatacept: 4 patients).

Disease activity states and responses to treatments:

JIA extended oligoarticular subtype	Cutoffs for disease activity states JADAS10/71	Number of patients at the entry into the study	Number of patients at age 18
Inactive disease	≤1		1
Low disease activity	1.1 –2	1	
Moderate disease activity	2.1 –4.2		2
High disease activity	>4.2	10	8
JIA Polyarticular RF (negative+positive) subtype			
Inactive disease	≤1		10
Low disease activity	1.1 –3.8	3	20
Moderate disease activity	3.9 –10.5	3	9
High disease activity	>10.5	47	14

Conclusion: It appears from our study that at age 18, 72,3% JIA patients with extended oligoarticular subtype and 26,4% with polyarticular subtypes report a JADAS71 high disease activity sum, so, these young adults will need transitional care.

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AB1034 NO DIFFERENCES IN HEPATITIS B ANTIBODY POSITIVITY IN CHILDREN WITH JUVENILE IDIOPATHIC ARTHRITIS ON BIOLOGICAL THERAPY

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Background: Biologic disease-modifying antirheumatic drugs (DMARDs) are frequently used in the treatment of juvenile idiopathic arthritis (JIA) resistant to nonbiologic DMARDs recently. There is a concern for their reducing the effectiveness of vaccines due to immunosuppressive effects.

Objectives: To evaluate hepatitis B antibody (anti-HBs) positivity and antibody titers between JIA patients on biologic DMARDs therapy and healthy controls.

Methods: Anti-HBs antibody positivity and titers were compared between 88 patients with JIA on biologic DMARDs therapy followed in our clinic and 91 healthy controls aged 2-19 years. All participants were vaccinated according to the routine vaccination schedule in our country.

Results: Eighty-eight JIA patients included and fifty five (62,5%) patients were female. The median age of patients was 13,31 (4,07-18,08) years. The JIA subtypes were systemic JIA (10,2%), oligoarticular JIA (33%), polyarticular JIA (25%), entesitis related arthritis (26,1%), psoriatic JIA

(3,4%), and undifferentiated JIA (2,3%). The median duration of biologic DMARDs was 31,995 (1,33-115) months. 54 patients were treated only with one anti-TNF (34 etanercept, 17 adalimumab), 16 patients were treated more than one TNF-alfa inhibitor. 18 patients were treated with more than one different group biologic DMARDs, and 7 of them had more than two biologic DMARD due to disease activity. Anti-HBs positivity was 67% in the JIA group and 54,9% in the control group, and no significant difference detected between the two groups (p=0,097). The anti-HBs titer was 12,95 (min-max: 2-1000) in the controls and 22,44 (2-1000) in the biological DMARDs.

Conclusion: Patients with JIA on biologic DMARDs had not lower anti-HBs positivity than healthy controls in our study. Further studies are required to understand the effects of biologic DMARDs on protective anti-HBs antibody levels.

Disclosure of Interests: None declared

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AB1035 MAFB-VARIANTS IN MULTICENTRIC CARPOTARSAL OSTEOLYSIS WITH NEPHROPATHY DO NOT SEEM TO AFFECT SERUM C1Q CONCENTRATION

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Background: Multicentric carpotarsal osteolysis syndrome (MCTO)-associated progressive nephropathy is an orphan disease associated with well-known structural kidney damage that usually requires kidney transplantation in adulthood. Although ACE-inhibitors could potentially slow down the progression of kidney dysfunction, other treatments would be welcome. Furthermore, a slight proteinuria reduction in a patient with MCTO treated with cyclosporine has been reported. MafB-deficient mice develop systemic lupus erythematosus-like autoimmune disease by inhibited C1q production and efferocytosis. In the human, monoallelic mutations in MafB result in MCTO.

Objectives: To investigate whether C1q levels in patients with MCTO are reduced and might contribute to the development of renal disease.

Methods: In three individuals with clinical and radiographic signs of MCTO, the MAFB gene was sequenced. The serum C1q concentration was determined with a commercial laboratory kit.

Results: Three patients with MCTO were enrolled (table 1). Patient A presented recurrent urinary tract infections since 18 months of life for a third-degree vesicoureteral reflux. Despite correction surgery, he displayed proteinuria after one year, and kidney biopsy did not reveal significant lesions. After two years, he displayed bilateral tarsal malformations requiring orthopedic surgery. At the age of 6, a pathological fracture of the right wrist occurred, and local X-ray revealed osteolysis of carpal bones. - Because of a suspected juvenile idiopathic arthritis of the right wrist, patient B was treated with intra-articular steroid injection and subcutaneous methotrexate for one year without benefit. After one year, he developed proteinuria and kidney biopsy revealed focal segmental glomerular lesions with tubular fibrosis and renal vasa sub-intimal delamination. C3 and C4 positive mesangial deposits were also present. - Patient C was referred to us for suspected juvenile idiopathic arthritis of the right wrist. No inflammatory signs were present at the ultrasound examination. Sequencing of the MAFB gene revealed in all the patients a novel heterozygous mutation. Although specific variants have not been reported before, the nature and position of the variants associated with the clinical features of the patients strongly argued in favor of their pathogenic role. The serum C1q concentration was normal in all the patients.

Abstract AB1035 Table 1. Main features of three patients with MCTO.

Patient	A	B	C
Age (years); gender; country	14; M; Croatia	16; M; Italy	11; F; Russia
MAFB mutation	c.184A>G, p.Thr62Ala	c.172A>G, p. Thr58Ala	c.208T>C, p. Ser70Pro
Bone lesions onset (age; years)	5	8	8
Presentation Site	Bilateral Tarsus	Monolateral Carpus	Monolateral Carpus
Current state (except carpotarsal joints)	Elbows and knees contractures	Knees contractures	None