was suspected and markers revealed serum ferritin 30,000 ng/ml, fibrinogen 250 mg/dl, Triglycerides 350 mg/dl and soluble CD25 12.000 U/ml. Bone marrow aspiration confirmed the diagnosis. Cytomegalovirus (CMV) IgM and PCR were significantly high. With regard to all the previous clinical and laboratory features, a CMV-induced secondary MAS in JSLE was confirmed. Patient was managed according to HLH-2004 treatment protocol (pulsed methylprednisolone 30mg/kg/day, cyclosporine A 6mg/kg/day, IV etoposide 150mg/m²/d) and IVIG. Ganciclovir was also added (5mg/kg IV q/12h x 7 days). Patient showed marked clinical improvement together with significant diminution of the MAS laboratory markers to normal. Two weeks post discharge on oral dexamethasone and cyclosporine A, the patient showed a severe clinical and laboratory MAS relapse (serum ferritin 50,000 ng/ml). The previous treatment protocol was restarted together with adding Rituximab 375mg/m² x 3 doses two weeks apart. Patient showed marked clinical and laboratory improvement and was maintained on full dose oral prednisone and oral Cyclosporine A. Tight gradual withdrawal of oral steroids was done with close clinical and laboratory follow up.

Conclusion: JSLE is a major diagnostic conundrum in pediatrics owing to its extremely variable clinical manifestations which can mimic many common pediatric conditions (e.g. malignant disease, other auto-inflammatory conditions, malignancy and any specific organ associated disease). Compared to adults JSLE has a more severe disease presentation. MAS is a life threatening condition that requires high index of suspicion and prompt management. In our case, controlling JSLE activity using Rituximab was significantly beneficial in arresting HLH evolution.

REFERENCES


AB0922

UPTDAYS ON THE EVIDENCE BASED INTERDISCIPLINARY GUIDELINES FOR HENOCH-SCHÖNLEIN PURPURA


Background: Henoch-Schönlein purpura (HSP) is the most common childhood vasculitis, it is characterized by inflammation of small vessels leading to non-thrombocytopenic purpura, arthritis, arthralgia, GI hemorrhage, evidence of nephritis/nephritic syndrome or renal impairment, evidence of neurological symptoms. Criteria for early referral to pediatric rheumatology and rehabilitation, School of Medicine Ain Shams University, Cairo, Egypt; 2Rheumatology and Rehabilitation, School of Medicine, Darent Valley Hospital, Dartford, United Kingdom; 3Rheumatology and Rehabilitation, School of Medicine Banha University, Banha, Egypt; 4Pediatrics, School of Medicine Cairo University, Cairo, Egypt; 5Rheumatology and Rehabilitation, School of Medicine Cairo University, Cairo, Egypt; 6Rheumatology and Rehabilitation, School of Medicine Cairo University, Cairo, Egypt; 7Rheumatology and Rehabilitation, School of Medicine Suez Canal University, Ismailia, Egypt; 8Pediatrics, School of Medicine Alexandria University, Alexandria, Egypt; 9Community and Public Health, School of Medicine Ain Shams University, Cairo, Egypt; 10Rheumatology and Rehabilitation, School of Medicine Assiut University, Assiut, Egypt

Methods: Interdisciplinary guidelines were developed with representative from PRINTO Egypt for pediatric rheumatology as well as patients’ group. A systemic literature analysis in MEDLINE was performed, evidence and recommendations were graded, an algorithm for treatment and final statements were discussed in a consensus meeting (Nominal Group Technique). A preliminary draft was fine tuned and discussed thereafter by all participants (Delphi Procedure).

Results: Consensus was reached on recommendations, including a standardized treatment strategy according to the HSP severity state in the individual patient. In this updated interdisciplinary guideline for HSP, treatment was tailored to the patient’s pathology and its severity (figure 1).

AB0923

EFFECTS OF EXON 10 MUTATIONS VS NON-EXON 10 MUTATIONS ON FMF PHENOTYPE AND RESPONSE TO TREATMENT

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Background: Familial Mediterranean fever (FMF) is the most common monogenic periodic fever syndrome. The MEFV gene mutation encoding the pyrin protein results in an uncontrolled increase in interleukin-1. Today, more than 333 MEFV mutations have been identified; however, exon 10 mutations are still seen to be best correlated with clinical findings.

Objectives: In this study, we aim to investigate the role of exon 10 mutations vs non-exon 10 mutations on clinical features and response to treatment in patients with FMF.

Methods: Data charts of children (n=935) with FMF from Dokuz Eylul University children’s hospital and Dr.B.Uz children’s hospital were reviewed. Patients were divided into two groups with regard to having