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AB1069
HYPERZINCAEMIA AND HYPERCALPROTECTINEMIA SYNDROME: MORE THAN JUST AUTOINFLAMMATION?

Andrea Uva1,2, Claudia Bracaglia1, Silvia Federici1, Camilla Celani1, Manuela Pardeo1, Christoph Kessel1, Fabrizio De Benedetti1, Antonella Insalaco1,1
1IRCCS Ospedale Pediatrico Bambino Gesù, Division of Rheumatology, Rome, Italy; 2University Children’s Hospital, Department of Pediatric Rheumatology and Immunology, muenster, Germany

Background: Hyperzincaemia and hypercalprotectinemia (HandH) syndrome has been described as a new rare entity characterized by recurrent infections, dermatological involvement, increased inflammatory markers, hepato-splenomegaly and anemia. Little is known about its heterogeneous presentation, pathophysiology and treatment.

Objectives: To describe three cases with HandH syndrome

Methods: Serum calprotectin (MRP8/14) was measured according to the EULAR ELISA and plasmatic zinc by atomic absorption spectrometry.

Results: Three patients were referred to our centre because an history characterized by recurrent episodes of skin rash, severe oral aphthosis and increased level of serum amyloid A (SAA). Patient 1 presented, since the age of ten years, with recurrent episodes of fever and rash; skin biopsy showed a picture consistent with a lymphocytic lichenoid vasculitis resembling erythema multiforme. Patient 2 presented at birth with hemolytic anemia and thrombocytopenia. At the age of 5 she was admitted to another hospital due to EBV related hemophagocytic lymphohistio-cytosis (HLH). At the age of 8, she was first seen at our center because of a persistent desquamant erythematous rash with recurrent abdominal pain and recurrent arthritis. Intestinal biopsy showed small intestine inflammation (erosions in the duodenum). Patient 3 presented with recurrent episodes of fever, rash, two episodes of transient hip synovitis and musculoskeletal pain. A bone scintigraphy was performed resulting in the diagnosis of juvenile idiopathic arthritis. Patient 3 was admitted at the age of 11 to our hospital for a severe disease activity score (DAS28). The patient presented a complete response to treatment with methotrexate.

Conclusions: HandH syndrome includes recurrent infections and multisystemic involvement. The disease is associated with excessive inflammatory markers, neutrophil dysfunction, and immune dysregulation process in which the role of zinc metabolism needs to be assessed.

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AB1070
MICROVASCULAR NAILFOLD ALTERATIONS BY VIDEOCAPILLAROSCOPY IN PATIENTS WITH RHEUMATOLOGICAL SYMPTOMS WITH JUVENILE INFLAMMATORY BOWEL DISEASE

Ana Villareal-Trevino1, Fernando García-Rodríguez2, Nadina Rubio-Perez2, Sergio Fernández-Ontí2, 1Hospital Universitario “Dr. José Eleuterio González”, Pediatrics, Monterrey, Mexico; 2University Hospital “Dr. José Eleuterio González”, Pediatrics, Monterrey, Mexico; 4Hospital Universitario “Dr. José E. González”, Pediatrics, Monterrey, Mexico; 5Hospital Zambrano Hellion TEC salud, Pediatrics, Monterrey, Mexico

Background: Juvenile Inflammatory Bowel Disease (IBD) is a chronic relapsing inflammatory condition of the gastrointestinal system that includes Crohn’s disease (CD) and Ulcerative Collitis (UC) and develops during childhood or adolescence in up to 25% of patients and affects the patients and parent s quality of life. Endothelial dysfunction is considered one of the etiological factors of IBD. Nailfold videocapillaroscopy is one of the best and safest diagnostic non-invasive imaging techniques to analyze microvascular abnormalities. Previous studies describe the involvement of the microvasculature in the pathogenesis of this diseases, suggested that chronic mesenteric vasculitis is a pathogenetic mechanism in CD. Nailfold abnormalities founded are similar to those observed in some systemic vasculitides. Subcapillary venous plexus dropout and low vessel density were previously reported.

Objectives: In this abstract we describe nailfold videocapillaroscopy findings in patients with IBD and their correlation with disease activity.

Methods: This is a prospective and analytical study that recruited pediatric patients between 2 and 18 years with IBD. A single cross-sectional 8-finger nailfold videocapillaroscopy was performed using a 200x optical probe videocapillaroscope. The images were collected, encoded and stored using the Optipix® software. Qualitative, quantitative and semi-quantitative assessment for architecture were scored following the international definitions for the capillary abnormalities Blood chemistry, C-reactive Protein, erythrocyte sedimentation rate, antineutrophil cytoplasmic antibody, and calprotectin were performed. Sociodemographic data, clinical evaluations, confirmation of IBD criteria, disease history, and activity evaluation were collected from patients clinical records. Statistics were performed using Spearman test to evaluate the correlation coefficient between the variables under study.

Results: 10 patients with IBD were included 60% male, 50% Ulcerative Collitis and 50% Crohn’s disease. 20% had severe disease according the Pediatric Ulcerative Collitis Activity index, Pediatric Crohn’s Disease activity index showed mild activity in just one patient. Rheumatological manifestations were found in 40%, 4 had arthritis, systemic vasculitides in 2 and 10% hematological manifestations. Abnormal endoscopy in 60% showing pancolitis. Normal microvasculature pattern were only found in 2 and 10% hematological manifestations. Abnormal endoscopy in 60% showing pancolitis. Normal microvasculature pattern were only found in 3 patients (25%), microangiopathy was found in 75%, edema of the nailfold was present in 50% of patients, a low capillary density was found in 100% of the patients with disease activity. A statistical negative correlation between the number of capillarities per millimeter and disease activity was found (coefficient -0.936, p=0.001).

Conclusion: Previous data showed that low capillary density was found in patients with IBD, the data in this pilot study are consistent with those findings. The assessment of the microvasculature through the use of videocapillaroscopy could be useful in this diseases.

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