HYPERZINCÆMIA AND HYPERCALPROTECTINÆMIA SYNDROME: MORE THAN JUST AUTOINFLAMMATION?

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Background: Hyperzincæmia and hypercalprotectinæmia (HandH) syndrome has been described as a new rare entity characterized by recurrent infections, dermatological involvement, increased inflammatory markers, hepatitis 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 Euhleman assay (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 aphtosis 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 hemolitic anemia and thrombocytopenia. At the age of 8 months, she was admitted to another hospital due to EBV related hemophagocytic lymphohistiocytosis (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 digiunum). 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 2 and 10% hematological manifestations. Abnormal endoscopy in 60% of patients and arthritis, systemic vasculitidies in 2 and 10% hematological manifestations. Abnormal endoscopy in 60% of patients and arthritis, systemic vasculitidies in 2 and 10% hematological manifestations. Abnormal endoscopy in 60% of patients and arthritis, systemic vasculitidies in 2 and 10% hematological manifestations. Abnormal endoscopy in 60% of patients and arthritis, systemic vasculitidies in 2 and 10% hematological manifestations. Abnormal endoscopy in 60% of patients and arthritis, systemic vasculitidies in 2 and 10% hematological manifestations. Abnormal endoscopy in 60% of patients and arthritis, systemic vasculitidies in. Endothelial dysfunction is considered one of the etiological factors of jIBD. 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 vasculitidies. Subapillary venous plexus dropout and low vessel density were previously reported.

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

Methods: This is a prospective and analytical study that recruited paediatric patients between 2 and 18 years with jIBD. 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 evaluation, confirmation of IBD criteria, disease history, and activity evaluation were collected from patients clinical records. Statistics were performed using Spearman to evaluate the correlation coefficient between the variables under study.

Results: 10 patients with jIBD were included 60% male, 50% Ulcerative Colitis and 50% Crohn’s Disease. 20% had severe disease according the Pediatric Ulcerative Colitis 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 vasculitidies 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 bed was present in 4 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 jIBD, 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 these diseases.

REFERENCES

Disclosure of Interests: None declared