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AB1069

HYPERZINCAEMIA AND HYPERCALPROTECTINEMIA SYNDROME: MORE THAN JUST AUTOINFLAMMATION?

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Background: Hyperzincaemia and hypercalprotectinemia (HandH) syndrome has been described as a new rare entity characterized by recurrent infections, dermatological involvement, increased inflammatory markers, hepatosplenomegalia 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 Buehlmann 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 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 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 normal. Patients 1 and 2 suffered from recurrent infections (pneumonia, otitis, skin abscesses). Immunological studies revealed in patient 2 a reduction of memory B cells and a reduced response to Toll like receptor 9 agonist. Of note, all the patients presented in their medical history at least one episode of vasculitis: patients 1 and 3 suffered from Schönlein-Henoch's purpura at the age of 11 and 3 respectively and patient 2 had at the age of 2 years an undefined vasculitis (evaluated elsewhere). Laboratory tests showed in all patients elevated inflammatory markers, zinchemia and serum calprotectin (table)

	Patient 1	Patient 2	Patient 3
Age at 1 st evaluation (years)	14.3	7.8	4.9
Sex	F	F	M
Clinical features	Recurrent fever, recurrent infection, vasculitis, skin rash, musculoskeletal pain	Hemolytic anemia, thrombocytopenia, HLH, vasculitis, recurrent infections, recurrent fever, arthritis, skin rash, gastrointestinal involvement	Recurrent fever, skin rash, vasculitis, arthritis, musculoskeletal pain
SAA	13-23	27,5-76,4	20
Zinchemia (mcg/dl) (n.v. 80-125)	132-143	102-233	147
Serum MRP8/14 (ng/ml) (n.v. < 2900)	22431	10383	10363

Figure 1

Conclusion: We report three patients with high serum levels of calprotectinemia and zinchemia presenting with a clinical phenotype consistent with previously reported cases. The presence of vasculitis in all of the patients suggest that it may represent the first symptom of this condition. Vasculitis could lead to an increase of serum calprotectin already proposed as a marker of vascular impairment. Moreover, considering the immunological defect detected in one of our patient, we speculate that

recurrent infections described in this syndrome may underline an immune-dysregulation process in which the role of zinc metabolism needs to be assessed

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

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Background: Juvenile Inflammatory Bowel Disease (jIBD) is a chronic relapsing inflammatory condition of the gastrointestinal system that includes Crohn's disease (CD) and Ulcerative Colitis (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 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 these diseases, suggested that chronic mesenteric vasculitis is a pathogenetic mechanism in CD. Nailfold abnormalities founded are similar to those observed in some systemic vasculitides. Subpapillary 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 pediatric 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 test 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 to 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 vasculitides 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 n=6 patients, a low capillary density was found in 100% of the patients with disease activity. A statistical negative correlation between the number of capillaries 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.

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