Abstracts of the 15th International Congress of the Egyptian Society of Pediatric Allergy and Immunology

Oral abstract: 01

The fatal consequences of delayed administration of cyclophosphamide in a case of neuropsychiatric lupus

Ahmed Seri I Mohamed, Tomador Siddig MZ.
Clinical Immunology Section, Jaafer Ibn Ouf Pediatrics' Hospital, Sudan

Neuropsychiatric lupus is a clinical entity of systemic lupus erythematosus (SLE). It comprises a variety of neuropsychiatric manifestations arising from autoantibody attacks on neural cells and/or cerebral vessels' endothelium. A combination of corticosteroids and cyclophosphamide is the first line of treatment of neuropsychiatric lupus. This case represents a "ring the bell" on the time factor in administration of cyclophosphamide in those patients. A nine-year-old Sudanese girl presented to the Pediatric Neurology Unit at Jaafer Ibn-Ouf hospital - Khartoum, with acute onset of right upper and lower limbs' weakness and aphasia. There was no disturbance of consciousness or convulsions and she gave no history of febrile illness or head or back trauma. She had no history suggestive of autoimmunity including joint or skin manifestations. Neurological examination showed depressed mood and aphasia, normal sensation, right sided paresis with power of grade one over the upper and lower limbs and normal power on left side. SLE diagnosis was confirmed based on 2015 ACR/SLICC revised criteria with overall score of 4 points which includes psychosis, leukopenia in on two occasions, positive antinuclear antibody and positive anti-phospholipids antibodies. CT brain showed Parieto-temporal ischemia. Pulsed methyl-prednisolone therapy was started together with hydroxychloroquine. She was planned to receive cyclophosphamide but – due to restricted resources – it was not initiated. Five days later, her condition deteriorated, she developed convulsions and was transferred to the pediatric intensive care unit for hypertensive crisis; after which she developed coma and had to be intubated. On day seven, she died. This case illustrates that the prompt administration of IV Cyclophosphamide along with the corticosteroids in neuropsychiatric lupus may be as lifesaving.

Oral abstract: 02

Upregulation of T cytotoxic 1 in Gaucher disease patients

Asmaa M. Zahran 1, Naglaa S. Osman 2, Esraa M.A. Eloseily 2, Rasha Refaat 2, Amir M. Abo Elgheet 2, Mervat A. M. Youssef 2, Khalid I. Elsayh 2
1Department of Clinical Pathology, South Egypt Cancer Institute; 2Children's Hospital, Faculty of Medicine, Assiut University, Assiut, Egypt.

Background: Gaucher Disease (GD) is associated with stimulation of the immune system with production of pro-inflammatory cytokines that could alter the counts of immune cells. CD8 T cell and its subsets T cytotoxic 1 (Tc1) and Tc2 are cytotoxic cells important in the defence against intracellular pathogens and tumors. Natural Killer (NK) and Natural Killer T cells (NKT) are vital components of the innate immune system and they regulate many autoimmune and inflammatory responses. Objectives: We sought to assess the counts of CD8 T cells, Tc1, Tc2, NK cells and NKT cells in GD patients. Methods: This single-center prospective controlled study included 20 GD patients and 20 healthy subjects. The patients were recruited from the Hematology Unit, Children's Hospital, Assiut University. They had Low β-glucocerebrosidase enzyme activity and had been receiving 60 IU of Imiglucerase enzyme therapy every two weeks for at least 6 months. They were subjected to clinical evaluation and laboratory investigations in the form of complete blood count, C reactive protein assay and counting of CD8 T cells, Tc1, Tc2, NK, and NKT cells by flowcytometry. Results: Significant increase in CD8 T cells and Tc1 was demonstrated in GD as compared to controls (p-value = 0.004, 0.024 respectively). NK cells showed a significant reduction in GD patients than controls (p-value = 0.029). However, no significant difference was noted concerning NKT cells. Conclusion: Elevation in CD8 T cells and especially Tc1 and reduction in NK cells are observed in GD. These immunological changes may contribute to the pathogenesis and progress of GD.

Oral abstract: 03

Not every eczema is atopic dermatitis.

Roba M. Elgaweesh
Pediatric Allergy and Immunology Unit, Children's Hospital, Ain shams University, Cairo, Egypt.

Hyperimmunoglobulin E syndrome (HIES) is a primary immunodeficiency disorder characterized by severe eczema, staphylococcal abscesses of the skin, recurrent sinopulmonary infections and elevated serum IgE (sIgE). There are two types of HIES; autosomal dominant type (STAT3 mutation) and autosomal recessive type (DOCK8 and TYK2 mutations). We hereby present a case with clinical features of HIES, showing mainly the DOCK8 mutation phenotype.
An 8-year-old girl presented to the Pediatric Allergy and Immunology Unit, Children's Hospital, Ain shams University, with severe eczema all over the body dating since 2 months of age which was resistant to conventional lines of treatment. She had history of recurrent attacks of pneumonia and recurrent skin abscesses. Shortly after admission, the girl developed acute onset of aphasia and inability to walk together with very aggressive behavior and conduct disturbance. She also had viral warts all over her body that were poorly responsive to topical antivirals. Her blood picture revealed moderate eosinophilia (4500/cu mm) while her total sIgE was high (750 UI/ml) and serum IgM was 25 UI/ml (moderately low). MRI of the brain showed picture of small frontal lobe abscesses together with picture suggestive of bilateral vasculitis and the chest CT scan showed bronchiectasis. The patient is currently on prophylactic oral cotrimoxazole together with IV antiviral and antifungal treatment in addition to skin local measures and pulmonary care (physiotherapy and inhalation therapy) with obvious clinical improvement. Conclusion: This case represents a phenotype of DOCK 8 mutation; however, genetic testing is still needed for accurate diagnosis and determination of further management plan including bone marrow transplantation.