compared with10.49±12.44 (pg/ml) in control group)(p<0.001) is higher. It was demonstrated that the average of INF-gamma/IL-13(0.3054in asthmatic group compared with1. 9334 in control group) in patients suffering from asthma, is lower. The size of BCG scar in Asthmatic patients was, significantly, smaller than the control group. Moreover, the average of Thelper1(INF-gamma) was lower and the average of Thelper2 (IL-4.13) was higher in asthmatic patients. Therefore, there is a correlation between the size of BCG scar and the levels of Thelper1(INF-gamma) and Thelper2 (IL-4.13) with asthma; thus, there could also be a correlation between the scar size of BCG, Thelper1 and 2.

474

HUMORAL IMMUNITY IN CHILDREN WITH CHRONIC TONSILLITIS

doi:10.1136/archdischild-2012-302724.0474

L Danilov, S Diacova, M Maniuc, P Ababii. Otorhinolaryngology, Pediatric Clinic, SMPhU, Chisinau, Moldova

Background Chronic tonsillitis is a common problem for pediatricians and otorhinolaryngologists.

The aim of our study was to evaluate and compare the humoral immunity in children and adults with chronic tonsillitis .

Subjects We examined 24 children and 13 adult patients with chronic tonsillitis.

Methods Immunologic examination included analysis of: total leukocytes; absolute and relative lymphocytes; C reactive protein (CRP), antistreptolysin O (ASO), rheumatoid factor; investigation of humoral immune factors: absolute and relative B lymphocytes, serum IgA, IgM, IgG, circulatory immune complexes, interleukin amount - IL-4, IL-8, IL-1β in blood serum.

Results Increasing level of the absolute lymphocytes amount (2.92 \pm 0.24; 2.2 \pm 0.11) (p = 0.04), interleukin - 8 (229.09 \pm 73.63; 35.05 \pm 14.64) (p= 0.047) and interleukin - 1 β level (191.19 \pm 68.44; 10.88 \pm 4.28) (p= 0.045) (p=0.045) were observed in children. Average indices of interleukin-8 and interleukin - 1 β in children is in 6.5 and 17.4 times higher than in adults.

Serum IgA level in adults was significantly higher than in children (1.36 \pm 0.16; 1.92 \pm 0.19) (p=0,038). An antistreptolysin-O titre in adults in comparison to children (162.5 \pm 45.53; 216.67 \pm 45.78) is higher in 1.4 times.

Conclusions Nonspecific defense factors play an important role in children with chronic tonsillitis, while in adults the main role is held by specific immune response. Comparative analysis of anti-inflammatory cytokine synthesis by immune competent cells in chronic tonsillitis proved the fact that in children it is much higher than in adults.

475

AUTOIMMUNE LYMPHOPROLIFERATIVE SYNDROME IB: IMPROVEMENT WITH RAPAMYCIN

doi:10.1136/archdischild-2012-302724.0475

G Lozano Sánchez, M Rivera Cuello, D Moreno Pérez, E Núñez Cuadros, VM Candón Moreno, A Urda Cardona. *Paediatric, Hospital Carlos Haya, Málaga, Spain*

Introduction Autoimmune lymphoproliferative syndrome (ALPS) is a disorder on a defect in the apoptosis of lymphocytes with linfoproliferation and immune dysregulation. Type Ib, defined by mutation in the gene that encodes the protein FAS-ligand, is a rare entity. **Case report** A 9-month-old male infant was referred because a failure to thrive and abdominal distention. Physical examination showed pallor, signs of severe malnutrition, axillary and inguinal lymph nodes, hepatomegaly and giant splenomegaly. Family background: parents, cousins of Moroccan origin, brother died at the age of 4when he was under study for a giant visceromegaly. Complementary tests highlighted severe anemia (Hb 7.30 g/dl) and thrombocytopenia (platelets 76000/mm3), paravertebral lymphoid

proliferation of 4 cm, elevation of IL10, soluble CD25, 28% T lymphocytes double negative (CD4 - and CD8-) in peripheral blood and cell culture with defect of apoptosis in one of the samples. The genetic study identified the mutation of the gene TNFSF6 which encodes FAS-ligand-protein. After the diagnosis of ALPS type Ib, treatment with rapamycin was started at doses of 2 mg/m2 diary. An optimal evolution was observed, with a reduction of visceromegaly size after 30 months of treatment, without adverse events by the time.

Conclusion We emphasize the importance of the suspicion of this entity in children with chronic visceromegaly, especially with family history. Despite the few existing data on treatment with rapamycin for this disease and children in general, we have seen an appropriate response and a good tolerance in this patient.

476

A CASE REPORT OF KARTAGENER'S SYNDROME ASSOCIATED WITH NASAL POLYPS

doi:10.1136/archdischild-2012-302724.0476

¹S Asilsoy, ²C Ozer. ¹Pediatric Allergy; ²Otolaringology, Baskent Universitesi, Adana, Turkey

Nasal polyps are benign nasal masses that can cause nasal obstruction, headache and snoring. The overall incidence or prevalence of nasal polyposis is unknown. They are diagnosed more frequently in men and during the third and fourth decades of life. Nasal polyps are rare in children. Most clinical data indicate that there is no greater prevalence of nasal polyps among atopic compared with normal populations.

A 14-year-old male patient was referred by an otolaryngologist to investigate the cause of nasal polyposis. There were otitis media, sinusitis and bronchial asthma to patient's history therefore he used inhale corticosteroid for control long term asthma, sometimes bronchodilatator to asthma attack and antibiotic treatment for sinusitis. No family member had similar respiratory complaints or any significant systemic illness.

His physical examination was found above 97% for weight and height difficulty breathing through nose, bilateral nasal polyposis, rarely sibilan ronchi on pulmonary auscultation and his heart sound was heard deeply. Laboratory findings; Hemogram was normal, IgA:189mg/dl, IgE 203 IU/L, inhalen sIgE and epidermal prick test were negative, Chest X-Ray showed dextocardia, A computed tomograph scan of the chest showed situs inversus with lateral segment of right middle lobe tubular bronchiectasis, peribronchial thickening and atelectasis, the right lower lobe tubular bronchiectasis. Pulmonary function testing demonstrated a mixed obstructive and restrictive pattern.

The pathogonomic findings of Kartagener Syndrome are dextrocardia, bronchiectasis and sinusitis. His findings were consistent with Kartagener Syndrome. This patient were presented due to nasal polyposis associated with Kartagener Syndrome and delayed diagnosis.

477

CHRONIC RECURRENT SEVERE LIP ANGIOEDEMA IN YOUNG CHILD SECONDARY TO ALLERGIC RHINITIS

doi:10.1136/archdischild-2012-302724.0477

¹D AlZahrani, ²M Al Shumrani, ³W Mansouri, ⁴J Yousef, ⁴S Abdulmalik, ⁵M Satti, ²S Al Mutairi, ²M Hasosah. ¹Immunology and Allergy, Pediatrics; ²Pediatrics; ³Pediatric Infectious Disease; ⁴Pediatric Surgery; ⁵Pathology Department, King Abdulaziz Medical City-WR, Jeddah, Saudi Arabia

Background and Aims Allergic rhinitis (AR) is a significant disease that affects children and adults and often under-diagnosed which has a deleterious impact on quality of life. Chronic recurrent severe lip angioedema secondary to AR was not previously reported.