ANNALS OF ALLERGY, ASTHMA, & IMMUNOLOGY

January, 1996
Volume 76, Number 1

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Official Publication of the American College of Allergy, Asthma, & Immunology

Abstracts: Poster Sessions

P45 HYPOGAMMAGLOBULINEMIA IN DIABETICS WITH NEPHROTIC SYNDROME, SR Lane MD; SJ Bigelsen MD Moorestown, N.J.

Nephrotic syndrome (NS) results when the glomerular filtration system becomes damaged by disease leading to complications including proteinuria, hyperlipidemia, edema and hypogammaglobulinemia. We report 2 cases of diabetics with NS, low IgG and severe infections.

The 1st is a 56 y/o woman with frequent bouts of severe bronchitis with various organisms (including Pseudomonas aeruginosa) as well as sinusitis requiring endoscopic surgery. She has developed chronic bronchitis and steroid dependent asthma and is currently responding well to therapy with IVIG. Her IgG is 548mg/dl (IgG1=375, 2=80, 3=21, 4=17) and urine protein is > 7gms.

The 2nd is a 69 y/o woman who requires prolonged courses of IV antibiotics to clear her bronchial infections. She becomes severely dyspneic with thick purulent sputum and has once required mechanical ventilation. Her IgG is 530 mg/dl (IgG1=386, 2=86, 3=48, 4=44) and urine protein is > 9gms/24 hours.

NS with low IgG occurs more frequently than reported and should be considered in diabetics and renal patients with recurrent infections. IVIG may be of benefit.

P46 SEVERE COMBINED IMMUNODEFICIENCY; A CASE REPORT S GONZALEZ MD., G GALINDO MD., G ALCALA MD., M GARZA MD., J ELIZONDO MD., H MORENO MD., C CANSECO MD. MONTERREY, NUEVO LEON, MEXICO

WE REPORT A 4 MONTH OLD LATIN BOY WITH VERY LOW VALUES OF IMMUNOGLOBULINS AND FAILURE TO THRIVE. SINCE THE FIRST MONTH OF LIFE HE HAD INTRACTABLE DIARRHEA, CHRONIC ORAL MONILIASIS AND GROWTH RETARDATION HE HAD SEVERAL HOSPITALIZATIONS, MEDICAL HISTORY: BREAST FED EXCLUSIVELY FOR THE FIRST MONTH OF LIFE, AT THE END OF THIS PERIOD HE HAD DELAYED SEPARATION OF THE UMBILICAL CORD AND GRANULOMA IN THIS AREA, CHRONIC DIARRHEA WITH 6-8 EVACUATIONS PER DAY AND ABDOMINAL DISTENTION, THAT IMPROVED WITH SOY MILK FORMULA TO 3 EVACUATION PER DAY. CONTINUED IN THIS WAY FOR THE NEXT 3 MONTHS AND AT THE AGE OF 4 MONTHS HE HAD GROWTH RETARDATION, (5.900 KG. , 63 CM) AND AN ERITEMATOUS AND DISEMINATED RASH:, HE WAS HOSPITALIZED FOR 15 DAYS, AND HIS LABORATORY DATA REVEALED: PERIPHERAL EOSINOPHILIA, ABSENCE OF IgA AND IgE LEVELS, VERY LOW LEVELS OF IgG AND IgM , A T CELL CD3COUNT OF 24 (59-90), CD2 22(62-92), CD4 8 (42-58), CD4/CD5 1 (10-23), CD20: 42 (0-10), CD56 :39(3-7). A BIOPSY OF GASTROINTESTINAL TRACT SHOWED CHRONIC INFLAMATION- AND A SKIN BIOPSY WITH RECENT HEMORRAGIC AREAS. HE RECIVED IgG IV (GAMAGARD) AND ANTIBIOTIC THERAPY FOR 14 MONTHS WHEN HE DIED.

P47 LATE DIAGNOSIS OF X-LINKED AGAMMAGLOBULINEMIA WITH NEUTROPENIA IN A 7 YEAR OLD BOY, M Garcia, MD; C Oh, MD; and C Song, MD, Torrance, CA.

The mean age of diagnosis of new XLA in previously unaffected families is 3½ years. We report a boy diagnosed with XLA at 7½ years of age. He presented with draining otitis media, pneumonia and neutropenia. He had history of recurrent otitis media since 8 months of age, pneumonia at 8 months and skin infection at 5 years. Laboratory results at the time of diagnosis: IgG < 33mg/dl, IgM 17mg/dl, IgA < 6.7mg/dl, IgE undetected, total B cell < 1%, absolute B cell < 1% total T cells 91%, CD8 49% CD4 41%, WBC 7.1 with absolute neutrophil count of 207. Patient responded to IVIG and systemic antibiotics. Neutropenia resolved within 2 weeks. The recurrent infections have led this patient to several outpatient medical services which failed to identify his primary immune problem. We conclude that an index of suspicion regarding XLA should be maintained in boys with recurrent infections despite normal appearance and growth parameters.

P48 SPECTRUM OF IMMUNODEFICIENCY IN THE FAMILY OF AN IgA DEFICIENT CHILD K T Hovanky MD, T P Atkinson MD, PhD, M L Johnson MD, H W Schroeder Jr MD, PhD, Birmingham, AL

IgA deficiency (IgAD), characterized by the absence of serum IgA in the presence of sIgA+B cells, is the most common primary immunodeficiency in the United States. We have previously proposed that IgAD and Common Variable Immunodeficiency (CVID), characterized by panhypogammaglobulinemia and normal numbers of peripheral B cells, represent the polar ends of the same spectrum of immunodeficiency. These disorders often occur in members of the same family. In order to better define the spectrum of immunodeficiency within families of IgAD patients, we are in the process of extensive family studies. We now report a family wherein the 6 y/o proband and his mother both have selective IgAD complicated by recurrent sinopulmonary infections. The proband's grandmother is clinically asymptomatic, but has borderline IgM, IgG, and IgA serum levels. She responded adequately to tetanus and pneumovax. The proband's maternal greataunt suffers from recurrent sinopulmonary infections, including pneumonia. Her IgG (719 mg/dl), and IgG1 (352 mg/dl) levels are just below the normal range, her IgG2 level is normal (323 mg/dl). However, she failed to respond to pneumovax and tetanus. Isolated IgG1 deficiency and poor antipolysaccharide antibody responses may lie within the spectrum of immunodeficiency characterized by familial IgAD and CVID.