Selective IgA Deficiency: Ruling out Coeliac Disease and Selective Antibody Deficiency to Polysaccharides

Ilenia Della Libera · Stefano Martellosi ·
Alberto Tommasini

Received: 1 March 2013 / Accepted: 24 June 2013 / Published online: 5 July 2013
© Springer Science+Business Media New York 2013

Dear Editor

We read with interest the report from Jorgensen on the February 2013 issue of this journal, showing a strong association between Selective IgA Deficiency (SIgAD) and respiratory infections.

An original point raised by the Authors is that the infection rate in patients with SIgAD detected during hospitalization wasn't different compared to a group of blood donors with SIgAD detected by screening. Thus, the Authors conclude that the clinical impact of SIgAD on respiratory infections may be higher than previously recognized. This may reflect low awareness of the problem and absence of clear guidelines for suspecting and diagnosing SIgAD.

We recently performed a similar study in paediatric patients, analyzing the results of IgA measured at the laboratories of our Hospital from January 2009 to January 2012.

We identified 50 SIgAD-subjects out of 6625 analyzed samples (cut off IgA <2 mg/dl under 4 years of age and <7 mg/dl above 4 years). Recurrent respiratory infections were found in 4 patients (8 %), one of whom, a 7-year-old girl, had repeated pneumococcal infections, including two pneumonia and one meningitis. All infections were due to pneumococcal strains present in the vaccines previously administered to her (conjugated 7-strain and polysaccharidic 23-valent). This behaviour prompted us to perform deeper investigations: IgG subclasses

resulted normal, but anti-pneumococcal antibodies remained undetectable after a further diagnostic vaccination. The girl was diagnosed with Selective Antibody Deficiency (SAD) and successfully treated with subcutaneous immunoglobulin replacement therapy.

SIgAD is an exclusion diagnosis. Normal values of IgM, IgG and subclasses aren't always enough to exclude a more serious condition, such as SAD (Sherkat R et al., Iran J Immuno 2013). With our case we would like to recommend performing pneumococcal antibody determination in all patients with SIgAD and recurrent serious bacterial infections.

In our experience, the more representative disorder associated to SIgAD was coeliac disease (CD) (13 patients=26 %). No patient had anti-tissue transglutaminase (tTG) IgA or EMA positive, but 11 patients presented high level of tTG-IgG and were diagnosed with CD after jejunal biopsy. Subjects with negative tTG results were further screened for HLA DQ2 and DQ8 and, if positive, they were selected for jejunal biopsy or follow-up based on the presence of CD-related symptoms. In our series, 2 tTG-IgG negative patients were finally diagnosed with CD. In all cases, intraepithelial lymphocytes were above the cut off of 25/100 enterocytes.

We would stress the diagnosis of CD in SIgAD patients can be difficult. It would be interesting to know how the Authors excluded the diagnosis of CD in their series.

I. Della Libera · S. Martellosi · A. Tommasini (✉)
Institute for Maternal and Child Health IRCCS Burlo Garofolo,
Trieste, Italy
e-mail: tommasini@burlo.trieste.it