activity of the glutamic acid decarboxylase enzyme in the Krebs cycle.[8] The patient reported here presented, paradoxically, an accumulation of crises in the context of a DKA, with mild acidosis. The MRI findings correlated well with both the changes in the EEG and her clinical symptoms.

Neuro-radiological alterations have been described after isolated or recurrent epileptic crises. The MRI anomalies are typically hyperintense lesions in the white matter in the T2 or FLAIR sequences.[2,3] Some authors have recently described transient subcortical T2 hypointensities in patients with crises and NKHI in both retrospective and prospective studies.[4,9,10] There is one reported case of ketotic hyperglycaemia associated with partial continuous epilepsy with reversible hypointensity of the subcortical white matter in T2 sequences.[6] Attention has also been called to the existence of diffusion restriction in patients with visual crises in the course of a NKHI.[11] Diffusion restriction suggests the presence of cytotoxic oedema. Local cytotoxic oedema may be related to both the seizure itself and the existence of focal ischaemia or hyperviscosity.[12] The slight gyriform uptake of contrast has also been described in crises associated with NKHI. During the crises, metabolic changes such as hypoxaemia, oedema, acidosis and cell membrane alterations, associated with endothelial dysfunction in diabetic patients, may lead to a disruption of the blood-brain barrier.[7]

In conclusion, patients with seizures symptomatic of DKA may present focal hypointensity in T2-weighted white matter with diffusion restriction, as well as cortical hyperintensities in FLAIR with gadolinium uptake. The semiology of the seizure and post-crisis stages and the changes in EEG correlate with these findings. Acknowledging these changes will facilitate the differential diagnosis when studying these patients.

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Selective IgA deficiency and multiple sclerosis

Déficit selectivo de IgA y esclerosis múltiple

Dear Editor:

Multiple sclerosis (MS) is the most common demyelinating autoimmune disease of the central nervous system in young adults and is one of the leading causes of non-traumatic, neurological disability.[1-3]

Selective IgA deficiency is the most frequent primary immunodeficiency.[4] Generally speaking, this deficiency is not associated with disease and is only revealed when routine laboratory studies are performed. However, the deficit of IgA is usually associated with infections of the respiratory and gastrointestinal tracts, and less often, with allergic and autoimmune diseases; these latter associations are a bit hazy from a physiopathological point of view.[5-8] Specifically, the association between selective IgA deficiency and autoimmune phenomena has been reported in both systemic, as well as organ-specific processes, the most widely reported of which are haematological disorders (idiopathic thrombocytopenic purpura), diseases of the gastrointestinal tract (ulcerative colitis), endocrine diseases (autoimmune thyroiditis), and rheumatological diseases.[9] To date, there have been no case reports of the association between IgA deficiency and MS.

With this in mind, we report here the case of a female with selective IgA deficiency and MS, and we will review the possible physiopathology of this association between the two poles of immunity.

Twenty-year old female with a history of selective IgA deficiency (levels undetectable in blood), diagnosed at the age of 4 years after undergoing studies for repeated episodes of respiratory tract infections; she presented no family history of interest. At the age of 15 years the patient underwent evaluation due to a clinical presentation that was compatible with optic neuritis of the right eye (pain on moving, gradual decrease in visual acuity, desaturation of colours, and alteration of the afferent papillary reflex). A cerebral MRI scan performed at that time revealed hyperintense, periventricular lesions in both hemispheres and callososeptal interface in T2 and FLAIR compatible with demyelinating lesions. The bloodwork was completed, ruling out an autoimmune, toxic, and/or metabolic process and an anlaysis of the CSF was carried out in which the presence of oligoclonal bands was apparent. The picture was interpreted as an isolated, demyelinating syndrome; immunomodulating treatment was not initiated at that time. At 20 years of age, the patient began to notice paraesthesias on the left side of her face and left arm, which lasted for 20 days followed by full recovery. A brain MRI was performed (figs. 1 and 2), in which new, bilateral, periventricular lesions were apparent. The immunological lab analyses were repeated, revealing nothing more than the persistence of serum IgA deficit. The event was considered to be a second clinical, demyelinating episode, establishing the diagnosis of relapsing-remitting MS and immunomodulating treatment was considered at that time.

We present the case of a female patient with IgA deficiency who was later diagnosed with MS.

IgA is the most abundant isotope of all the immunoglobulins produced by the immune system. The IgA present in secretions is necessary to neutralize viruses, to bind toxins, agglutinate bacteria, and prevent the binding of bacteria to the cells of the epithelial mucosa, as well as to bind several food antigens to keep them from entering the general circulation,[4,8] comprising one of the most efficient mechanisms by which to control infection through the mucosal tissues.

Of all the primary immunodeficiencies, selective IgA deficiency is the most prevalent defect,[7,8,10] with an observed frequency of 1 in 600. The criteria for its diagnosis according to the European Society for Immunodeficiency are: IgA levels of less than 7 mg/dl with normal levels of IgG and IgM in a male or female over the age of 4 years, in whom other causes of immunodeficiency have been ruled out.[8,10]

Although it might seem paradoxical *a priori*, IgA deficit is associated with autoimmune phenomena and is considered to be a risk factor for the development of such processes (systemic lupus erythematosus, rheumatoid arthritis,[8,9,11] idiopathic thrombocytopenic purpura, autoimmune haemalytic anaemia, coeliac disease, and thyroid disorders). In Western countries, the prevalence of autoimmunity in patients with IgA deficit is approximately 3-5%;[10] however, this may vary from 7-36% and even 40% in symptomatic individuals.[8,10,12] Up until the present time, there have been

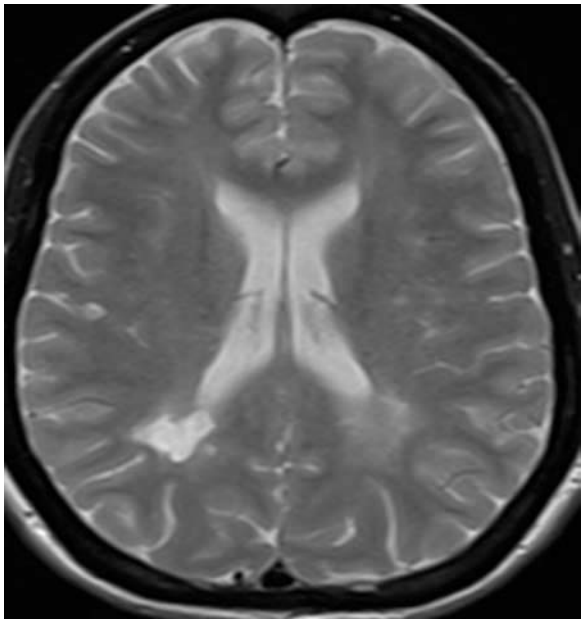

Figure 1 Axial MRI (T2).

different hypotheses that attempt to account for the association between IgA deficiency and the presence of autoimmunity.[8,9] One of these hypotheses maintains that because IgA is the first line of immune protection on the mucosal surface against potentially harmful external agents,[13] its absence might facilitate the absorption of a great many environmental antigens, with the possibility of these agents triggering cross reactions with their own antigens, and with the subsequent production of auto-antibodies and autoimmunity.[8] Another hypothesis postulates that the deficiency in the immune response for the eradication of microbial and viral pathogens and the persistent antigenic stimulation would bring about a compensating, exaggerated chronic inflammatory response that would lead to tissue damage and, consequently,

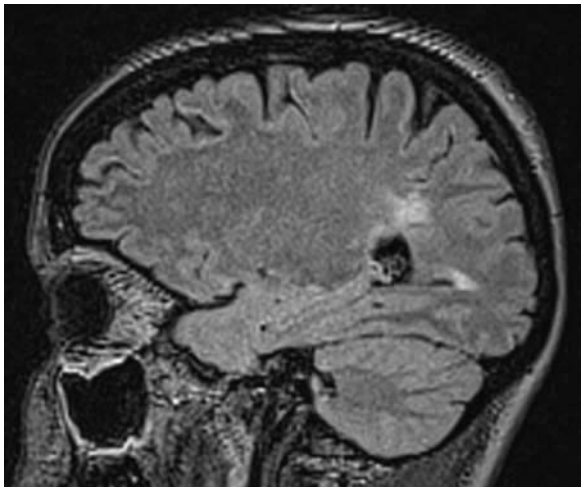

Figure 2 Sagittal MRI (FLAIR) that reveals hyperintense periventricular lesions in the white matter.

autoimmune phenomena.[5,8,9] Finally, another theory would establish an association between IgA deficit and alterations on the part of T cells in the regulation of peripheral tolerance, hence provoking the autoimmune process.[8,14] Regardless of the cause for the association, what is clear is that IgA deficit patients have a higher risk of associated autoimmune diseases.

This is the first case in the literature reporting an association between IgA deficiency and MS. Although there are several hypotheses, the exact role of IgA deficiency in the genesis of the autoimmune phenomenon has not been elucidated to date. Future research will clarify the precise role of this association.

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Diagnosis of delayed cerebral ischaemia and vasospasm in subarachnoid haemorrhage: How long should they be monitored?

Diagnóstico de la isquemia cerebral tardía y el vasoespasmo en la hemorragia subaracnoidea: ¿hasta cuándo monitorizar?

Dear Editor:

We have reviewed with great interest the article published by Rodríguez-García et al. regarding the diagnosis of delayed cerebral ischaemia and cerebral vasospasm in subarachnoid haemorrhage (SAH), which appeared in the previous issue of your journal (Neurología. 2010;25:322-330).[1]. We believe this to be a document of the greatest importance and extremely timely, as well as being very well developed, and we should like to contribute a few comments.

Rodríguez-García et al. report, on the basis of a review of the literature, that delayed cerebral ischaemia is a major cause of morbidity and mortality among patients presenting SAH and that this ischaemia is secondary to delayed cerebral vasospasm, therefore proposing monitoring, mainly through clinical and ultrasound examinations, for the first 10 days following SAH before reducing and eventually suspending monitoring.

We present here a case of delayed vasospasm and its clinical, radiological and ultrasound correlation, manifested on the 16th day after the SAH.

Female, 52 years old, hypertensive without treatment, presenting a diffuse SAH at basal cisterns, WFNS I, Fisher 3, with a Glasgow score of 14 on admission, without neurological focality. On the same day, a cerebral arteriography was performed and revealed a 3 x 2 mm aneurysm of the right posterior cerebral artery, a 3 x 4 mm aneurysm in the anterior communicating artery and an infundibular dilatation of the right posterior communicating artery. The first aneurysm was treated endovascularly. The following day, the aneurysm in the anterior communicating artery and the dilatation of the right posterior communicating artery were clamped.

The patient progressed favourably, with scant stiffness in the neck and a good level of consciousness (Glasgow 14,