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Juvenil Dermatomyositis Associated to Selective IgA Deficiency: A Case Report and Literature Review

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Absctract

Selective immunoglobulin A deficiency (sIgAD) is the commonest primary immunodeficiency worldwide. Autoimmunity is common manifestation, few cases of cases of juvenile dermatomyositis associated to sIgAD are published. We present a case with both conditions.

Keywords: *Selective immunoglobulin A deficiency, primary immunodeficiency, juvenile dermatomyositis*

Introduction

Selective immunoglobulin A deficiency (sIgAD) is the commonest primary immunodeficiency worldwide, it is defined as decreased serum level of IgA in the presence of normal levels of other immunoglobulin isotypes. (Leman, 2010) Patients with sIgAD may present asymtomatic or with recurrent infections (most of them of respiratory and gastrointestinal tract), allergic disorders and autoimmune manifestations. (Jacob, Pastorino, Fahl, & Carneiro-Sampaio, 2008) (Leman, 2010)

Among the autoimmune associated disorders, the hematologic anomalies are the most common. There are few cases of Juvenile Dermatomyositis (JDM) associated to sIgAD.

A case of an 11-year old girl diagnosed with JDM and selective IgA deficiency is described.

Case Summary

An 11-year-old asthmatic girl consulted to the ER of our center, with one-month history of muscle weakness, associated with recurrent falls. She needed help to dress and perform daily activities. Her mother noted an erythematous rash in her eyelids. At examination she presented heliotrope rash, proximal symmetrical muscle weakness and Gottron´s papules. Laboratory findings revealed elevated CPK 25 247 IU/L, AST 908 U/l, ALT 619 U/L, LDH 1994. Hemogram showed Hb 13g/dl, Platelets 522/mm^3 and discrete leucocytosis: WBC 13 040/mm^3, with polymorphonuclear predominance. Serum immunoglobulin levels: IgA undetectable [less than 23mg/dL (below the lowest detectable value)], IgM 88.2mg/dl, IgG 1410mg/dl, positive ANA 2531 units, negative dsDNA and negative AntiSm. Electromyography revealed an inflammatory myopathy pattern. She was diagnosed with juvenile onset Dermatomyositis and was started on steroids (Prednisone 2mg/Kg/d).

Nevertheless, she continued to be symptomatic (with muscle weakness) after one week of treatment, so she was readmitted to receive intravenous metilprednisolone (30mg/Kg/d) for 3 days. Oral methrotexate (12.5mg/Kg/d) was initiated and she discharged after considerable improvement on her symptoms.

She had a relapse seven months later, at this hospitalization a second measurement of IgA was undetectable, so the diagnosis of sIgAD was made. She was started con Azyotropine and subcutaneous methrotexate with improvement.

Discussion

Juvenile dermatomyositis (JDM) is a rare, autoimmune illness characterized by muscle and skin involvement, with less frequent involvement of other systems, including the gastrointestinal tract and lungs (Bitnum, 1964). It has an incidence of approximately 2-3 cases per million children per year in some places. The mean age of onset is 7 years-old. (Martin, Li, & Wedderburn, 2012) (Modesto, 2014)

The diagnosis of JDM is established with the Bohan and Peter criteria, described in 1975 (Leman, 2010). Our patient presented with the most frequent clinical features of JDM: the heliotrope rash, the Gottron`s papules and the proximal muscle weakness. (Wedderburn & Rider, 2009) She fullfilled 4 out of 5 criteria for the diagnosis. The muscle biopsy was not needed, in fact, nowadays, the criteria of muscle biopsy and electromyography is under discussion, and other methods have begun to replace them, as is proposed by some important investigations groups. (Juvenile Myositis Carra Subgroup, 2014)

The diagnosis of selective IgA deficiency was made according to clinical and laboratory criteria (PAGID-ESID), as listed on Table 1. The serum IgA level of less than 7mg/dL is used because this concentration is the lowest detectable limit established by most laboratories. The age of 4 years is used to avoid premature diagnosis of sIgAD in younger children in whom there may be a delay in ontogeny of IgA system after birth. (Leman, 2010)

Table 1. *IgA Deficiency diagnostic criteria according to PAGID and ESID*

Definitive	Probable
Male or female greater than 4 years of age	Male or female greater than 4 years of age
Serum IgA less than 7mg/dL	Serum IgA at least 2 SD bellow normal for age
Normal serum IgG and IgM	Normal serum IgG and IgM
Other causes of hypogammaglobulinemia excluded	Other causes of hypogammaglobulinemia excluded
Normal IgG antibody response to vaccination	Normal IgG antibody response to vaccination

(PAGID: Pan-American Group for Immunodeficiency and ESID: European Society for Immunodeficiencies; 1999)

The decreased IgA levels should be confirmed in at least 2 consecutive tests, and other causes of immunodeficiency must be ruled out. (Abolhassani, Gharib, & Shahinpour, 2015). Our patient had undetectable levels in two different measurements. Although, for the second measurement she already had 6 months on steroids and methrotexate.

sIgAD patients may course asymptomatic in 85 to 90% of cases. The symptomatic patients develop sino-pulmonary infections, gastrointestinal infections, allergies, autoimmune conditions, and malignancies. (Leman, 2010)

Autoimmune diseases are among the most important clinical manifestations of IgAD. (Leman, 2010) This phenomena has been reported since the late 60s. (Amman & Hong, 1970). Among the most frequent associations are autoimmune thyroiditis, vitiligo, autoimmune hemolytic anemia, chronic arthritis and Celiac disese. (Abolhassani, Gharib, & Shahinpour, 2015), (Fahl, Silva, Pastorino, Carneiro-Sampaio, & Jacob, 2015)

The association of sIgAD with JDM however is very rare. In a recent study that evaluated 57 symptomatic patients with sIgAD (37 males, 20 females) and their relatives, 2 out of 17 patients with concomitant autoimmune disorders had Dermatomyositis. The first case was a female that had sIgAD associated to DM and thyroiditis. The second case was a male with sIgAD, autoimmune hemolytic anemia, Dermatomyositis and juvenile rheumatoid arthritis. (Abolhassani, Gharib, & Shahinpour, 2015)

Lisak and Zwiman analyzed the serum immunoglobulin levels in 30 patients with myasthenia gravis, polymyositis and dermatomyositis. They found one patient with polymyositis who had a low IgA level (42 mg/dL) with no other abnormalities on IgG or IgM levels. Instead of finding sIgAD, there were five patients with polymyositis or dermatomyositis with elevated levels of IgA. (Lisak & Zweiman, 1976)

Several hypothesis have been proposed to explain the increased risk of autoimmunity in patients with sIgAD (Abolhassani, Gharib, & Shahinpour, 2015): (a) secretory IgA plays an important role in mucosal surface`s protection, so in a deficiency state, environmental antigens can easily penetrate the

mucosa and cause molecular mimicry that leads to a cross-reaction with self-antigens with the production of auto-reactive antibodies formation; (b) sIgAD could result from shared genetic factors, such as common human leukocyte antigen alleles or haplotypes, which can predispose the affected individual to both diseases and (c) IgA has a role in anti-inflammatory processes and in pathogens clearance, so there may be defective antigen clearance leading to immune complex deposition in the inflamed tissue of various organs.

Conclusion

The association of JDM with sIgAD is very rare. However, being the most common immunodeficiency worldwide, it is important to screen asymptomatic patients with IgAD for autoimmune conditions, specially the hematologic anomalies and consider malignancy as an alternative in this population. Patients with autoimmune conditions could be screen for IgA levels.

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