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Case Report

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Delayed diagnosis of selective immunoglobulin deficiency: A case report

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ABSTRACT

Selective immunoglobulin A deficiency (SIgAD) is the most common of all primary immunodeficiency diseases; however, the pathogenesis has not been fully understood. Although most people with SIgAD are asymptomatic, some may present with recurrent infections; such as respiratory disorders, gastrointestinal tracts disorders, and allergic disorders. Herein, we report a case with SIgAD who presented with the complaints of pruritus, dental caries and chronic sinopulmonary infections.

1. Introduction

Selective immunoglobulin A deficiency (SIgAD) is defined as a serum Ig A level at less than 5 mg/dl. This ilness is the most common one which occurs in primary immuno deficiency but pathogenesis is not defined clearly (Phankingthongkum et al., 2002).

First described in serum in 1953, SIgAD has a worldwide prevalence that differs from one region to another; 1:143 in the Arabian peninsula, 1:163 in Spain, 1:252 in Nigeria, 1:875 in England and 1:965 in Brazil (Yel, 2010). A lower incidence of SIgAD has been reported from Asian countries; such as Japan, China, India, and Saudi Arabia (Phankingthongkum et al., 2002). In the USA, the prevalence of SIgAD is estimated to range from 1:223 to 1:1.000 in community studies and from 1:333 to 1:3.000 among healthy blood donors (Cunningham, 2001).

Although most people with SIgAD are asymptomatic, some may present with recurrent infections of respiratory and gastrointestinal tracts, allergic disorders and autoimmune manifestations (Yel, 2010).

Herein, we report a case with SIgAD who has presented to outpatient clinic with pruritus, dental caries and chronic sinopulmonary infections.

2. Case report

A 26-year-old female who admitted to our outpatient clinic with the complaint of itching who had suffered from it 15 years. In the past she had been diagnosed with acute gingivitis, pneumonia and a seasonal allergic rhinitis, and had been prescribed corticosteroids, antihistamines, analgesics and antibiotics. However, the itching proved to be persistent, her other symptoms recurred, and she developed several dental cavities. Information about the patient's family history, history of allergy, weight loss, occupation and hobbies was obtained. She had no prior illnesses. Her vital parameters were stable. Except for several scracth marks all over her limbs due to itching, there ısn't any physical examınaton (Fig. 1).

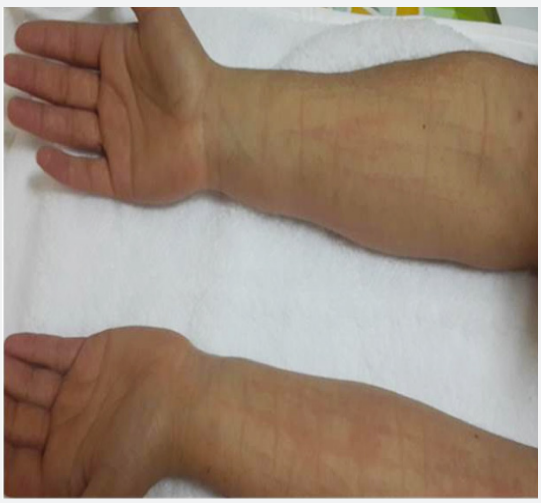

Fig. 1. There were skin abrasions on both arms, due to prolong period of scratching

A pulmonary function test was performed. Forced expiratory volume in 1 (FEV1) was measured as 85% and FEV1/forced vital capacity (FVC) was measured as 84%. Table 1 shows the laboratory parameters. Blood IgA level was measured to be 2.5 mg/dl, and IgG and IgM were normal. A preliminary diagnosis of IgA deficiency was made.

3. Discussion

There are two classified types of IgA deficiency: complete IgA deficiency and partial IgA deficiency. If blood IgA level is lower than 5mg/dL but IgG and IgM levels are normal, this condition is identified as a complete IgA deficiency. If blood IgA level is 5 mg/dl or higher, but the standard deviation of the age-specific mean value is below 2, then it is classified as a partial IgA deficiency (Conley and Nortarangeld, 1999).

Some patients with IgA deficiency frequently develop recurrent sinopulmonary infections, allergies, autoimmune conditions, and malignancies. Other

Table 1. Laboratory parameters of the patient

Variables	Level of laboratory parameters	Normal ranges
Immunoglobulin G	1480 mg/dL	650-1600 mg/dL
Immunoglobulin A	2.5 mg/dL	3.5-250 mg/dL
Immunoglobulin M	191 mg/dL	50-300 mg/dL
Immunoglobulin E	1060 mg/dL	0-100 mg/dL
Antinuclear antibodies	Negative	Negative
C3 (Complement)	124 mg/dL	79-152
C4 (Complement)	27.9	16-38
Salmonella	Negative	Negative
Brucella	Negative	Negative
C-RF	Negative	Negative
ASO	52.4 ıu/mL	0-200
Anti-ds DNA antibodies	Negative	Negative
HBsAg*	Negative	Negative
Anti HBs**	Negative	Negative
Anti HCV***	Negative	Negative
Aspartate aminotransferase	26 u/L	0-40 u/L
Alanine aminotransferase	13 u/L	0-41 u/L
Thyroid stimulating hormone	2 µıu/mL	0.2-4.2 µıu/mL
Free throxin	1.1 ng/dL	0.9-1.7 ng/dL
Carcino-embryogenic antigen	1.9 ng/dL	0-5.2 ng/dL
Anti thyroglobulin antibody	19.3 ıu/mL	0-155 µıu/mL
Anti TPO****	10.1 ıu/mL	0-34 µıu/mL
Carcinoma antigen-15-3	20.6 mg/dL	0-25 mg/dL
Carcinoma antigen-125	18.1 mg/dL	0-35 mg/dL
Carcinoma antigen-19-9	11.9 mg/dL	0-39 mg/dL

*: Hepatitis B virus-surface antigen; **: Anti hepatitis B virus-surface antigen; ***: Anti hepatitis C Virus antibody; ****: Antithyroidperoxidase antibodies; **ASO:** Antistreptolizin-O

associated diseases that are commonly seen in patients with IgA are gastrointestinal infections; such as giardiasis, malabsorption, lactose intolerance, celiac disease, ulcerative colitis, nodular lymphoid hyperplasia, and malign proliferation (Yel, 2010).

SIgAD can also be related with the increased frequency of allergic disorders. In one study, atopy was found in 58% of pediatric and adult patients with IgA deficiency (Buckley, 1975). Another study reported a history of allergy and asthma in 13% of patients with IgA-deficiency, which is probably not higher than the percentage of people with atopy in general population (Edwards et al., 2004).

In 2008, (Jacob et al., 2008), investigated IgA deficiency in 126 patients and found that 48% had respiratory allergies and atopic dermatitis. In a prospective study, IgA-deficient pediatric patients were found to be at an increased risk of pseudocroup at year one and parentally reported to have had food hypersensitivity at year four, both of which were possibly not IgE-mediated, as compared to children with normal serum levels of IgA (Janzi et al., 2009). In another study, 40.5% of the patients presented with the

symptoms of allergy. It was found that 25% of patients with IgA deficiency were diagnosed during screening for allergic disorders (Cunningham, 2001).

There are many factors involved in the formation of dental cavities such as; age, education level, eating habits, oral hygiene, frequency of dentist visits and fluoride intake. In addition, long-term antibiotic treatments for recurrent infections and immunoglobulin replacement therapy may also suppress microorganisms in dental plaque (Fernandes et al., 2012).

Legler et al. (1981), found that patients with immunodeficiency had a higher incidence of dental caries than the general population. In the case we presented, the patient had several dental caries and had undergone recurrent dental operations.

In conclusion, in patients with long term pruritus, tooth decay and sinopulmonary infections, oral examination should be performed at regular intervals in addition to the normal physical examination. In patients with recurrent dental caries, IgA deficiency should be considered and an early diagnose should be made.

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