





Case Report

Linear Immunoglobulin a Bullous Dermatitis in a Child

Bir Çocukta Linear İmmünoglobulin Büllöz Dermatöz

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ABSTRACT

A 22-month-old male patient had extensive itchy and firm bullous lesions on his entire body, including his scalp. First of all, infectious bullous diseases were considered, but infectious diseases were ruled out with tests. Skin biopsy was examined with immunofluorescence tests for a definitive diagnosis. The patient was diagnosed with linear IgA bullous dermatosis disease, and treatment was started. Follow-up was continued due to insufficient response to treatment. In pediatric patients, it is important to distinguish Linear IgA Bullous Dermatitis from other autoimmune bullous diseases and bullous infectious diseases. With the right treatment, the quality of life of the patient can be improved in the early period. In this case, we examined the way the patient was diagnosed with Linear IgA Bullous Dermatitis and reached the right treatment.

Keywords: Bullae, Bullous Disease, Linear IgA Bullous Dermatitis

ÖZET

Bu çalışmada 22 aylık erkek hastada kafa derisi de dahil olmak üzere tüm vücudunda yaygın kaşıntılı ve sıkı büllöz lezyonlar görüldü. Öncelikle enfeksiyöz büllöz hastalıklar düşünüldü fakat yapılan testlerle ekarte edildi. Kesin tanı için immünofloresan testleri ile cilt biyopsisi incelendi. Hastaya lineer IgA büllöz dermatöz hastalığı tanısı konuldu ve tedavisine başlandı. Tedaviye yanıtın yetersiz olması üzerine takiplerine devam edildi. Pediatrik hastalarda Lineer IgA Büllöz Dermatözün diğer otoimmün büllöz hastalıklardan ve büllü enfeksiyon hastalıklarından ayırt edilmesi önemlidir. Doğru tedavi ile erken dönemde hastanın yaşam kalitesi yükseltilebilir. Bu vakada hastanın Lineer IgA Büllöz Dermatöz tanısı almasına ve doğru tedaviye ulaşmasına giden yolu inceledik.

Keywords: Büll, Büllöz Hastalık, Lineer IgA Büllöz Dermatöz

INTRODUCTION

Autoimmune bullous diseases are a disease group that is rarely seen in the pediatric group age. Common pathogenesis for these cases are autoantibodies that target specific adhesion molecules of the skin that are

the cause of mucosal and/or cutaneous bullae formation (1,2). Linear IgA Bullous Dermatitis (LABD) is a disease characterized by subepidermal tight bullae and Linear IgA aggregation in the basal membrane zones(3).

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The disease has a bimodal age tendency, which is usually seen in children between the ages of six months and 10 years and adults after the age of 60. The cause of LABD cannot be identified for most patients. However, in some cases, LABD has been reported to occur after using antibiotics, antihypertensives or nonsteroidal anti-inflammatory drugs (4,5). It has been reported that LABD is associated with lymphoproliferative disorders, infections, ulcerative colitis, and systemic lupus erythematosus (4).

CASE REPORT

In this study, 22 month-old male patient with no known illnesses applied to our clinic with scratchy and sore tight bullous lesions filled with clear liquid, approximately half a centimeter in diameter, spread to the body and scalp, which has first seen in genital and perineal areas, then densely in bilateral lower limbs.

Vital findings of the patient were within the normal range for his age (Heart rate: 100/min, respiratory rate: 20/min, blood pressure: 90/60mm-Hg). During the appliance, the patient was agitated, but his overall condition was good. Bullous lesions of different sizes and widths were mostly found on the face, scalp and extremities and are present throughout the body. Physical examination of other systems was evaluated as normal. The lesions were burst in some places due to the scratching (Figure 1). Lesions were not present in the patient's oral mucosa, palms and soles. According to the patient's parents, the patient had no history of drug use, no history of inflammatory disease, and no history of drinking well water or raw milk. It was also ascertained that no other family members had any similar symptoms and no allergic reaction to vaccines or foods. On the 2nd and 3rd days after the appearance of the first lesion, itchy lesions burst, new lesions appeared and the patient was reluctant to urinate due to pain in the perineal region.

Laboratory parameters checked during the patient's hospitalization, white blood cell count was $17400 \times 10^9/L$ (N: $4.2-10.8 \times 10^9$), C-reactive protein was 3 mg/L (N: 0-5), and erythrocyte sedimentation rate was 10 mm/hour (N: 0-20). Transaminase levels and renal function tests were evaluated as normal. No evidence of hemolysis or atypical cells was found in the peripheral smear. Clindamycin treatment was initiated due to the growth of Methicillin Resistant Staphylococcus Aureus in the blood culture taken during hospitalization.



Figure 1. Before Treatment

LABD, Herpes Simplex Virus (HSV) infection, hypereosinophilic syndrome, and bullous mastocytosis were considered in the differential diagnosis of the patient. Tzanck smear was performed to rule out HSV/Varicella Zoster Virus (VZV) infections since the lesions of the patient on the trunk and scalp were scattered varioliform, some dried and some vesicle-shaped, and no pathology was found. No pathological result was found in the Francisella Tularensis agglutination test, HSV, VZV and Borrelia serologies, which were examined on the patient to investigate infectious causes. HSV PCR test performed from the patient's tissue fluid was negative. A punch biopsy sample was taken from one of the lesions in the patient's leg.

In the biopsy sample, moderate inflammation involving neutrophils was shown in the superficial dermis in the interstitial and perivascular areas. In the immunofluorescence examination, a linear positive, strong fluorescence was obtained with IgA along the basement membrane. In the light of these findings, the biopsy sample resulted in accordance with LABD. The patient was consulted to the dermatology department. Spontaneous LABD was accepted as there was no history of drug use and mucosal involvement stated before the

lesions appeared in the patient's history. After the patient was diagnosed with LABD, intravenous methylprednisolone was started at a dose of 1 mg/kg/day and oral hydroxyzine dihydrochloride treatment was started to prevent itching.

To investigate the relationship between LABD-IgA nephritis described in the literature, a complete urinalysis test was performed and the results were within normal limits. No protein excretion was observed in the spot urine in the complete urine test (6). Cases with inflammatory bowel diseases and LABD have been reported (7), but the patient did not have symptoms, such as chronic diarrhea, joint pain or conjunctivitis. The eye examination performed to evaluate the eye findings of LABD, such as vision loss, dry eye and irritation, was normal.

After the treatment was started, the patient's agitation decreased, his oral nutrition increased, and his itching complaints disappeared. A significant reduction in bullous eruptions was detected in the physical examination performed at the 48th hour of methylprednisolone treatment; it was observed that there were 1-2 bullous lesions in each region, bullous lesions regressed, leaving areas that were hypopigmented.

On the 6th day of the treatment, blisters in the patient's body disappeared completely (Figure 2). Physical examination performed on the 14th day of treatment revealed significant regression in the hypopigmented areas (Figure 3). The patient was discharged with oral methylprednisolone treatment.

The patient continued his outpatient follow-ups regularly for a while after discharge. During this period, new lesions were seen on the scalp and trunk. Therefore, oral prednisolone treatment was discontinued on the 28th day. Oral dapsone and topical steroid treatment were started. The patient stopped coming for follow-ups after one month. Therefore, we did not know our patient's response to oral dapsone and topical steroid therapy. The consent from the patients' parents was obtained for this case study.

DISCUSSION

LABD is a rare immunobullous disease with an incidence of 0.2-2.3/1,000,000 cases (9). On physical examination, erythematous or urticarial bullous lesions are seen. These lesions are generally tense and differ in size. Lesions are usually located in the lower abdomen, perineal region, inner thigh and scalp in children. Involvement of the face, hands and feet is rare. Bullae can be

broken up and crusted due to intense itching and disappear, leaving an erosive post-inflammatory depigmented area (4).



Figure 2. 6th Day of Treatment

Histopathological examination reveals an infiltration predominantly neutrophil in the superficial dermis and predominantly in the dermoepidermal junction, arranged in a characteristically linear array. The gold standard method for diagnosing LABD is direct immunofluorescence (DIF) examination of skin biopsy. Linear IgA deposits are seen along the basement membrane at the dermoepidermal junction with DIF. It can rarely be seen in Ig G, Ig M and C3. The indirect immunofluorescence (IIF) method is used to detect circulating IgA antibodies. Western immunoblot is a more sensitive method than IIF. The most common antigens detected in LABD by Western immunoblot are BP 180, LABD-97 and Ladinin-1(LAD-1). IgA accumulation under an immunoelectron microscope; it can be seen in lamina lucida and/or lamina densa/sublamina densa layers (9).

Our patients' lesions were intense, especially in the perineal region, inner thighs and scalp. Although facial rashes are rarely reported in the literature, there were also rashes on the patient's face.

The factors determining LABD treatment are the determination of the degree of involvement and triggering factors. If there is the use of triggering pharmacological agents, such as vancomycin and aspirin, discontinuation of these agents provides improvement of skin findings gradually in a few weeks (7). The patient did not have a history of using any drug. Clinical cases, such as Steven Johnson Syndrome and Toxic Epidermal Necrolysis (TEN), have been reported in the literature(8), but clinical findings similar to these were not observed in the patient. It has been reported that mucous membranes are also affected in forms triggered by drug use. In addition, it is stated that the drug-triggered form is more severe than the spontaneous form (10). From this point of view, the patient's history and physical examination conform to the spontaneous LABD form (9).



Figure 3. 14th Day of Treatment

Dapsone, used in leprosy treatment, is considered the first-line pharmacological agent in the treatment of LABD (4). Dapsone is recommended at a dose of 0.5-2 mg/kg/day (max: 100 mg/day) in children. Dapsone was not preferred in the first-line treatment in the patient because of its excessive side effects. However, it was

preferred due to the lack of response to steroid use in the future. The most common side effects of dapsone are anemia, leukopenia, agranulocytosis, hemolysis, hepatitis, cholestatic jaundice, peripheral neuropathy, psychosis, and cutaneous hypersensitivity. Other treatment options include topical and systemic corticosteroids, sulfonamides, immunosuppressants, intravenous immunoglobulin (IVIG), plasmapheresis, and extracorporeal photopheresis (11).

Autoimmune bullous diseases are rare in children; they are a group of diseases that should be kept in mind in the differential diagnosis of these lesions, showing significant improvement with medical treatment. In approaching the patient, it is important to first obtain a detailed anamnesis of the patient, learn about the recent drug use, and question whether there are similar diseases or accompanying symptoms in the family. The location, type and appearance of the fluid in the skin lesions are important in physical examination. Definitive diagnosis of these clinically similar diseases is possible with immune histochemical methods. Direct and indirect immunofluorescence methods are the gold standard for diagnosing this group of diseases.

Patient Consent Form / Hasta Onam Formu

The parents' of this patient consent was obtained for this study.

Conflict of Interest / Çıkar Çatışması

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