ABSTRACT:
IgA linear bullous dermatosis, also known as chronic bullous dermatosis of childhood, is an autoimmune disease which may be idiopathic or drug-induced. The disease affects children and adults. We present a 4 years old girl with itchy polymorphic eruptions. The skin rash was presented by bullous-erosive rosette-like lesions with reddish-brown crust in the center, distributed on the skin of the face, trunk and extremities. The vesicles were filled with serous and hemorrhagic content. Laboratory examinations were within normal values according the age. Histopathological examination of the lesional skin revealed sub epidermal blister. Direct immunofluorescence of perilesional skin demonstrated linear deposition of IgA in the basement membrane. Systemic treatment with Methylprednisolon and Claritromycin was applied with satisfactory effect. The patient is under observation.

Key words: IgA linear bullous dermatosis, direct immunofluorescence, Indirect Immunofluorescence, Methylprednisolon, Claritromycin.

INTRODUCTION:
IgA linear bullous dermatosis is a rare acquired autoimmune vesiculobullous disease characterized by linear IgA deposition on dermo-epidermal basement membrane observed with direct immunofluorescence. The characteristic clinical lesions are vesicles and tense bullae, which most often are grouped, giving a “cluster of jewels” appearance. The disease affects children and adults. It may be idiopathic or drug-induced.

CASE REPORT:
We present a case of 4 years old female child with tense serous content-filled vesicles and bullae, distributed mainly on the skin of the face around the mouth. The disease started two months ago with intense itching. With a diagnosis Impetigo the child was treated with systemic and topical antibiotics with slight improvement. After the end of the therapy the rash spread to trunk and extremities. From medical history according the mother the rash appeared immediately after a consumption of fruit yogurt. The child suffers from frequently episodes of spastic bronchitis.

Physical status: a child in good general condition, without fever, there were no pathological changes from general status.

Dermatological status: there were multiple bullous lesions grouped in rosette-like pattern with reddish-brown crusts in the center. The eruptions involved the skin of the face, trunk and extremities /Fig 1a, 1b, 1c/. Mucous membranes were not affected by pathological changes. Nikolsky sign was negative. Routine laboratory examinations were within normal values. Microbiological culture from bullous exudates and culture examination from feces, for Candida albicans, were negative. Parasitological examinations for intestinal parasites and protozoa and serological tests for toxoplasmosis and echinococcosis /ELISA/ were negative also. “Tzanck” test did not reveal acantholytic cells. Histopathological examination of the lesional skin showed subepidermal blister, filled with fibrin and neutrophilic inflammatory infiltrate. Direct immunofluorescence (DIF) of the perilesional skin revealed linear deposition of IgA (+++) at the basement membrane zone. Indirect immunofluorescence showed IgA-anti-basement membrane zone antibodies (human esophagus substrate) /Fig 2a, 2b/. A diagnosis IgA linear dermatosis was accepted. Systemic treatment with Claritromycin, 125 mg/5 ml suspension 2x5 ml/daily, Methylprednisolone 1mg/kg/daily i.m. and Choropyramine hydrochloride 10 mg/ml solution/daily i.m. was conducted. Topical therapy with Methylprednisolone aceponate 0.1 % cream was administrated. After 10 days treatment significant improvement with partial epithelization of the erosive lesions occurred /Fig 3a, 3b/. The child was released from the hospital in good condition, without bullous lesions. It was recommended to continues the treatment at home with Claritromycin, 125 mg/5 ml suspension, 2x5 ml/ daily for a month. One month later the patient achieved complete remission. A control microbiology investigation for Candida albicans from feces was negative. The child is under observation.
**Fig 1a, 1b, 1c.** Multiple bullous lesions grouped in rosette-like pattern with reddish-brown crust in the center on the skin of the face, trunk and extremities.

**Fig 2a.** Direct immunofluorescence (DIF) of the perilesional skin - linear deposition of IgA (+++) at the basement membrane zone.

**Fig. 2b.** Indirect immunofluorescence (IIF) of the human esophagus substrate - IgA-antibasement membrane zone antibodies.
DISCUSSION:
Immunoglobulin A (IgA) linear bullous dermatosis is an autoimmune bullous disease affecting primarily young children and adults and characterized by subepidermal blister formation and linear homogenous deposition of IgA at the basement membrane zone (1). Although it is a rare dermatosis, it is the most common autoimmune bullous dermatosis in children, the disease usually begins after the age of six months, with a peak incidence at 4 to 5 years. The etiologic factor triggering the autoimmune mechanism of the disease remains unknown (2). In IgA linear bullous dermatosis, autoantibodies recognize multiple basement membrane zone antigens, which are responsible for the different clinical aspects of the disease (3, 4). The characteristic lesions are vesicles and tense serous bullae, which most often are grouped giving a “cluster of jewels” appearance. They are mainly located on the lower abdomen, perineum and perioral area. Immunofluorescence testing is an important tool for diagnosing IgA linear bullous dermatosis and indirect immunofluorescence has a higher sensitivity when salt-split skin technique is performed (5). In the presented case the direct immunofluorescence of perilesional skin revealed linear deposition of IgA at the basement membrane zone and indirect immunofluorescence showed IgA-antibasement membrane zone antibodies in human esophagus substrate, which confirmed the diagnosis IgA linear bullous dermatosis. Differential diagnosis must be established with other autoimmune dermatoses, such as dermatitis herpetiformis and bullous pemphigoid. Remission in IgA linear bullous dermatosis occurs in most cases until puberty, and spontaneous remission is often observed within two years. The most commonly used treatment modalities are corticosteroids, dapsone and sulphapyridine, with excellent response in a short period (6). There are also reports of the use of antibiotics such as doxycycline, erythromycin, and oxacillin, as in the case presented (7).

CONCLUSION:
We report a case of a 4 year old girl presenting with bullous lesions, being diagnosed as IgA linear bullous dermatosis by direct and indirect immunofluorescence, with good response to systemic glucocorticosteroids and antibiotics (Clarithromycin) in less than 2 weeks.

REFERENCES:

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