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LEUNER-MOUSSOUX DESQUAMATIVE ERYTHRODERMA, A RARE AND SEVERE DISEASE OF NEWBORNS

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Desquamative erythroderma of Leiner-Mussu, a rare and severe disease of newborns. The description of clinical case, course, complex of therapeutic and prophylactic measures in dexvamatic erythroderma Leiner-Mussu is presented. This is a rare severe disease of children of the first 3 months of life, which in case of untimely diagnosis and inadequate treatment can be fatal, and therefore it is important to timely recognize the dermatosis and refer sick children for treatment in hospital, which will significantly increase the chances of recovery. The unclear etiology, pathogenesis and rarity of occurrence presents difficulties in diagnosis and treatment. Erythroderma Leiner-Mussouw, diagnosis, clinic and treatment.

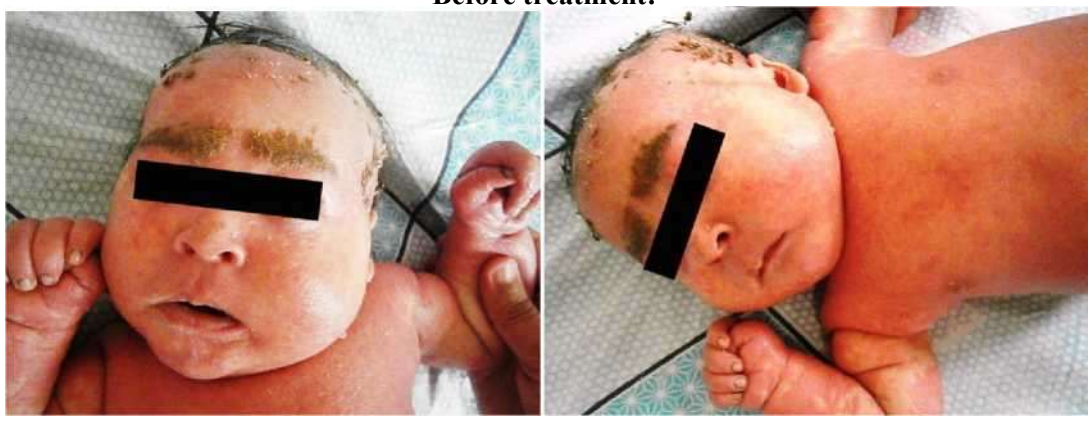
Erythrodermia desquamotiva Leiner-Mousslous - was first isolated as a separate nosological unit by French pediatrician Mousslous in 1905 and described by Austrian pediatrician Leiner in 1907.

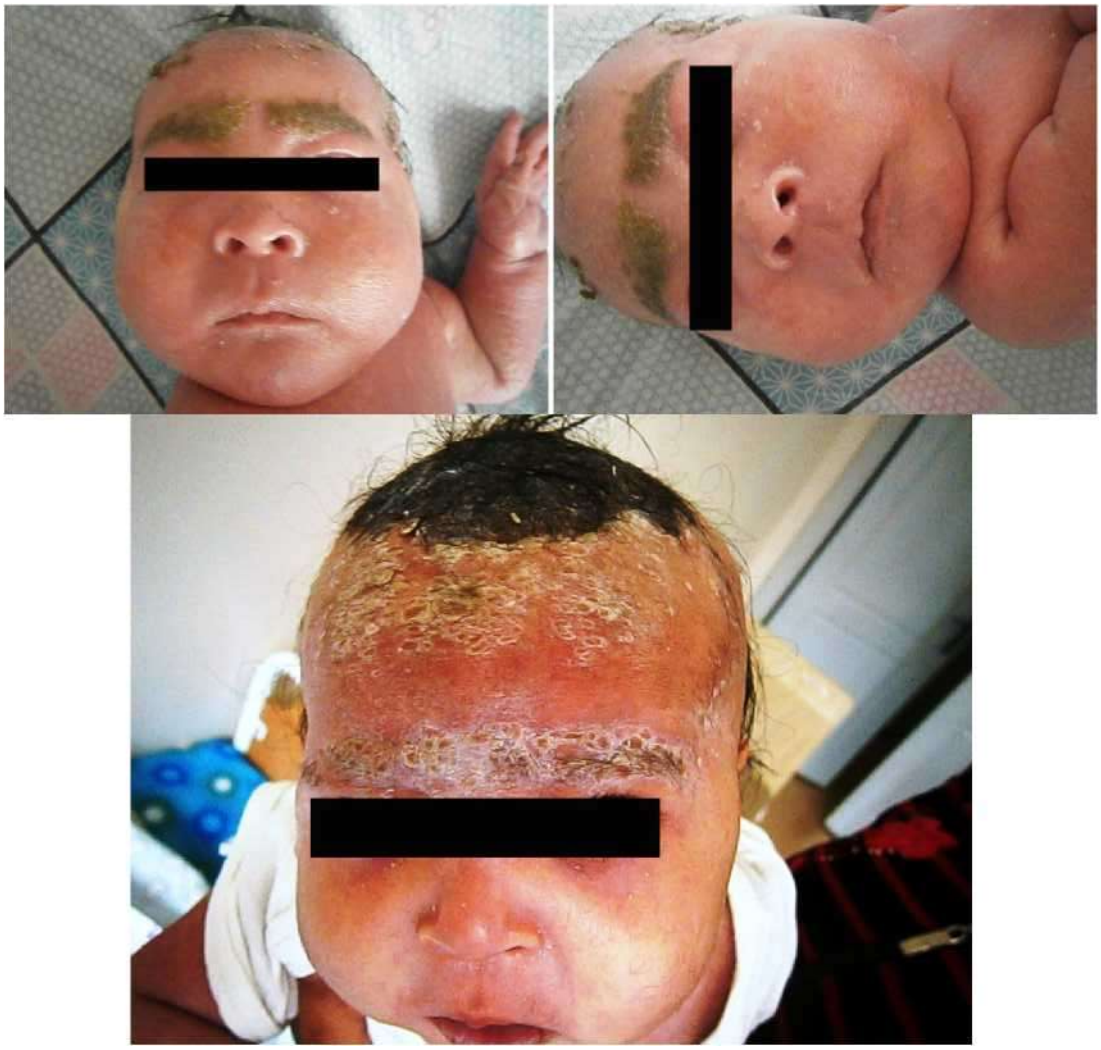
Leiner-Mousslous dexvammatory erythroderma (DELM) is a rare severe disease of children in the first 3 months of life. The etiology of the pathogenesis of DELM is not completely clear. The disease is considered as a generalized form of seborrheic dermatitis, its erythrodermic variety. The prevailing opinion is that seborrheic dermatitis under unfavorable conditions can transform into DELM [1,2,4,5]. One of the main causes of DELM development is considered to be auto-intoxication, because skin lesions are aggravated with the severity of gastrointestinal tract disorders [3], associated with a decrease in the enzymatic activity of the intestine and wall digestion leading to a violation of protein and lipid metabolism. It should be noted that in the causes of DELM besides intoxication with products of disturbed metabolism, an important role is played by feeding with qualitatively inferior mother's milk, with a reduced content of biotin, vitamins of the B group (B2, B6, B12), A, E, C [1,5,6]. It should be noted that the disease is more characteristic of breastfed children, more often girls. The disease is aggravated by pyococcal and candida infection, which have a sensitizing effect on the body [6,7]. Some researchers consider DELM an independent disease [6].

The main skin signs of DELM are flaky erythroderma, and diarrhea and nutritional disorders (hypotrophy) in the GI tract. The disease more often begins in the 3-4th week of life with dyspeptic disorders (frequent regurgitation, vomiting, frequent liquid stools). The skin process more often begins in the buttocks, groin and hip folds with the appearance of bright red flaky foci, and sometimes the pathological process can begin with the scalp, moving to the neck, upper torso, axillae, descending into the anogenital area. Within a few days, most of the skin becomes brightly hyperemic, covered with papillary and lamellar abundantly flaky che-shuiki, except for small islets of unaffected skin. In the area of cervical, axillary and inguinal-femoral folds there are skin maceration with formation of cracks and wetting. The scalp is covered with multilayered thick scales - crusts, grayish-yellow and brownish color in the form of a shell, descending on the forehead, supraorbital arches with spreading to the occiput. Temporary baldness is possible. Scales can be removed, but soon they reappear. The skin under the scales is hyperemic with infiltration. Pathologic changes in the skin may persist for an average of 1 month. Mucous membranes, hair and nails are not affected.

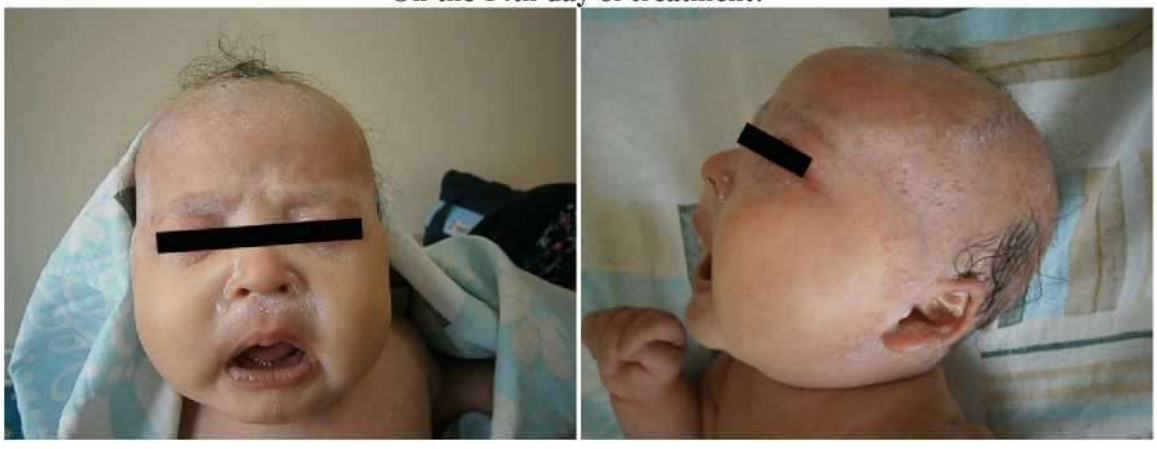
Photos:

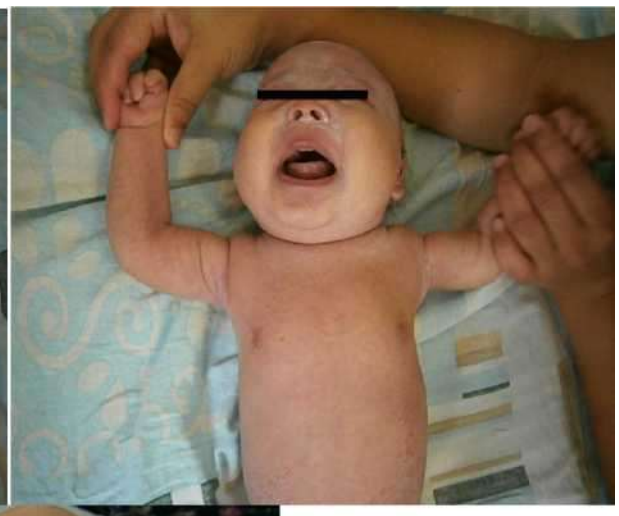
Before treatment:

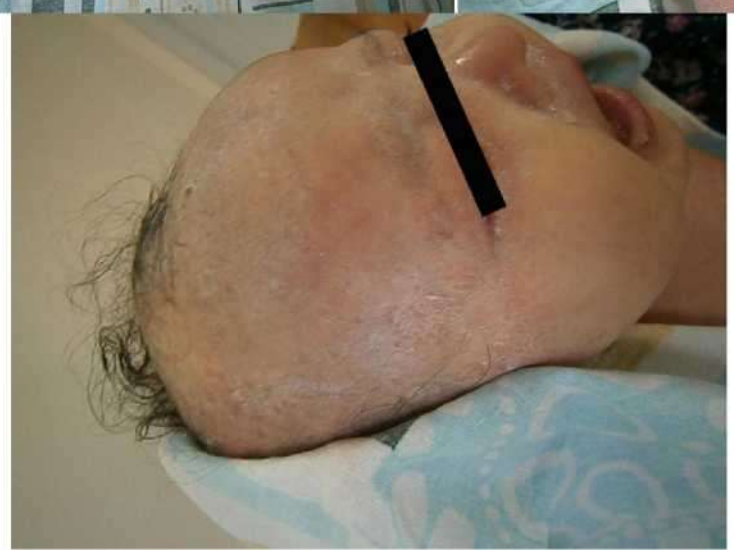




On the 14th day of treatment:







Timely adequate treatment in the hospital provides cure within 2-4 weeks. The complex of treatment and rehabilitation measures includes normalization of feeding of the child (caloric vitaminized food of a nursing mother, additionally vitamins C, B, A), broad-spectrum antibiotics (lincomycin, gen-tamycin, ampicillin, cephalosporins), glucocorticosteroids, desensitizing drugs, hemotransfusions, immunocorrective drugs, pro and prebiotics, enzymes that help and improve digestion. Properly conducted topical therapy is of great and key importance, it should include combined ointments and creams with anti-inflammatory, antibacterial, antifungal and epithelializing action.

Here is our observation: Patient A. I/B #230/82 was born on 06.06.23. From Kashkadarya region, admitted to the infant department of pregnancy Tashkadarya PMI on 22.09.23. child from the 2nd

child was born on time, physiologically with weight of 2 births, pregnancy was toxicosis, the of 3,500 grams. The child grew and developed normally until 25 days of age, breastfed, skin was clear. At the age of 25 days, the child had green, frequent, liquid stools and at the same time in the area of large folds appeared pronounced redness with wetting. We applied to the pediatrician in the SVP at the place of residence, who recommended topical ointment sinoflan, for 3-4 days erythema seized almost the entire skin, after which the child was referred to a dermatologist in the district clinic, where the child was diagnosed: Congenital ichthyosis with hospitalization in the hospital, but the child's condition did not improve and the parents arbitrarily took the child home and turned to the local "tabib", which as a treatment recommended to smear the whole body with plain sour cream with the addition of red ground pepper, which the parents did. The next day the child developed high fever, adynamia, erythematous areas covering the whole body became wet and covered with layered crusts. Due to the severity of his condition, the child was hospitalized in the children's somatic department of Chirchik city**.** Where, according to the mother, the child received benzylpenicillin every 6 hours for several days and topical ointment Dermoveit. Again, the parents took the child away from the medical institution of their own volition due to further deterioration of the child's condition without discharge and applied to the clinic of TashPMI, where the child was hospitalized in the infant department in an emergency order on 22.09.23.

At the time of admission, the general condition of the child due to general intoxication is extremely severe, passive and tearful, there was an affection of almost the entire skin in the form of continuous erythroderma covered with large scales. In the area of cervical, axillary and inguinal folds there were wet erosions and linear fissures, the scalp and supraorbital arches were covered with multilayered serous-purulent crusts in the form of a "helmet". Breathing through the nose was difficult, rapid with signs of dyspnea and pronounced perioral cyanosis. auscultation in the lungs hard breathing. After a review radiograph of the lungs, ultrasound, laboratory tests, examination of allied specialists: pediatrician, surgeon and dermatologist, the diagnosis was made: Desquamative erythroderma Leiner-Mussu, complicated by pyoderma with concomitant acute bilateral focal bronchopneumonia.

General therapy: cefatoxime 200mg per 50.0ml of saline solution x 2p.v.d v/v; 30% sodium thiosulfate solution 1.0 ml per 30.0ml of saline solution v/v; dexamethasone 4% 0.3 ml v/v; Drotaverine 2% 0.2 + dimedrol 1% - 0.2 per 2.0ml of saline solution v/v; albumin 10% of saline solution v/v #1; plasma transfusion #1 Per.os: fenistil 0% 0.2 + dimedrol 1% - 0.2 per 2.0ml of saline solution v/v. v/v; albumin 10% solution v/v №1; plasma transfusion №1 Per.os: phenistil 0.1% 2 drops x 2p.v.d.; bifobalance - baby 1 sachet x 2 p.v.d.; Creon 1/3 cap. x 3p.v.d.; asparkam % tablet x 1p.v.d.; aevit 1 drop x 1p.v.d. Inhalations and sanitation of the upper respiratory tract.

Topical combined therapy: methylene blue 1% (for cracks and wetting), "candida" powder (in the area of large folds), ointment "levamikol" + l-t "Vishnevsky" (1:1) for crusts under / bandage, cream "triderm" + zinc ointment (1:1) for redness and erosions. On the 7th day from the beginning of intensive therapy the general condition of the child began to stabilize, signs of dyspnea disappeared, redness and swelling of the skin decreased. On the 11th day the general condition became satisfactory, the scalp and supraorbital arches were significantly cleared of crusts. On the 17th day hyperemia in the lesions was gone, erosions and fissures were covered with epithelium, the child gained weight and was discharged home in satisfactory condition. It was recommended to continue supportive therapy and follow-up with the pediatrician and dermatologist at the place of residence.

The present publication and the described clinical observation will be useful for physicians, it will help them to be prepared to meet this rare, severe and complex nosologic form for timely and adequate treatment in order to achieve the fastest clinical effect and to avoid many complications and unjustified therapeutic measures. Prevention of DELM should start from the antenatal period and include rational nutrition of the pregnant and later lactating mother (complete, balanced, rich in vitamins), adequate regimen and proper care of the newborn.

Literature

1. Zverikova F.A. Diseases of the skin of children. St. Petersburg: Sotis. 1994:172-175.
2. Popkhristov P. Skin diseases in childhood. Sofia. 1963:236-238.
3. Atlas Ya. E. Skin diseases of the early infantile age. M 1961.
4. Studenikin M. A. Handbook of pediatrician. M: Ellis lac. 1994;313- 314.
5. Reference book. Edited by O.L. Ivanov. Skin and venereal diseases. Moscow: Medicine. 1997;331-332.
6. Skripkin Y.K., Zverikova F.A., Sharapova G.Y., Studnitsin A.A. Manual of pediatric dermatology. Moscow: Medicine. 1983;88-92.
7. Mannanov A.M., Khaitov K.N. Bolalar teri va tanosil kasalliklari. Toshkent. 2016:560.
8. Ashurova, D. T., and D. I. Ahmedova. "Hormonal adaptation features and neonatal period of newborn children depending on the body weight at birth." World Healthcare Providers Multidisciplinary medical journal USE (2012): 41-44.
9. Акрамова, Х. А., and Д. И. Ахмедова. "Характерные особенности плацентарного фактора роста при задержке внутриутробного развития плода." Педиатрия 3-4 (2014): 29-31.
10. Ahmedova, D. I., and Rahimjanov Sh A. Growth. "development of children. Methodical recommendation." (2006): 3-82.
11. Хакимова, Г. Г., А. А. Трякин, and Г. А. Хакимов. "Применение ниволумаба при раке толстой кишки с синдромом Линча. Клиническое наблюдение." Злокачественные опухоли 10.1 (2020): 41-48.