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Atypical juvenile type of the Devergie disease

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ABSTRACT

Devergie disease or pityriasis rubra pilaris (PRP) — is an idiopathic hyperproliferative dermatosis. Clinically it is characterized by follicular papules, usually disseminated reddish orange scaling plaques with areas of normal skin around; palm and feet keratoderma. Onset occurs in the first or the fifth decade of life.

From the five clinical types of PRP, according to Griffiths W.A. Classification (1980), type IV (atypical juvenile) is rare. It develops during the first years of life. PRP is characterized by acquired well-defined plaques of follicular hyperkeratosis located on variably distinguished erythematous areas on knee and elbow joins, less often on other areas of the skin. Sometimes hyperkeratosis or cracks on palms and feet may be present. In some cases, onychodystrophy may be noted.

We followed up a five-year-old female patient A. that in 4 years of the disease was diagnosed by different dermatologists with various conditions: Atopic dermatitis, psoriasis, follicular keratosis, xerosis, streptodermia Treatment that followed brought only temporary effect.

Biopsy from the lesions was performed in daytime hospital of «Kolomenskiy» branch of State Budgetary Healthcare institution «Moscow Scientific and Practical Center of Dermatovenerology and Cosmetology» of the Healthcare Department of Moscow. Histologically diagnoses of psoriasis and Devergie disease were not excluded.

During treatment (2% salicylic ointment, «Topikrem», «Pimafucort», «Losterin») we noted positive trend of skin condition, its stabilization.

Keywords: atopic juvenile type of Devergie disease, clinical presentation, course, treatment.

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Pityriasis rubra pilaris (PRP) has been known for about 200 years. The disease was first mentioned by C. Tarral (1829), then described in detail by M.G. Devergie in 1857, and was named by E. Besnier in 1889 [12,14,25].

Devergie's disease (DD) (also known as pityriasis rubra pilaris) is a rare idiopathic hyperproliferative dermatosis characterized by follicular keratotic papules, keratoderma, erythema of varying severity, and scaling [4,6,7]. Pathogenesis of the disease is associated with abnormal keratinization in the setting of keratinocyte hyperactivity as well as with dysfunction of vitamin A metabolism and weakening of the protein-binding liver function [3,4,8].

The etiology of PRP still remains elusive. The development of PRP is affected by autonomic and hormonal dysfunctions, hepatoses, infections, insolation, vaccinations, etc. [1,4,5].

According to various data, the disease accounts for 0.03–1.3% of all skin diseases [5,8,28]. Its onset occurs in the first or fifth decade of life [4,14,21]. The congenital form of PRP has an autosomal dominant type of inheritance, and the acquired (sporadic) form is not genetically determined [3,4,5].

The disease first affects the follicular apparatus. Initially, in the presence of erythema, isolated miliary follicular-hyperkeratotic and cone-shaped papules appear in various parts of the skin. Grouped rash elements increase in the number, merge, and form plaques of various sizes [9,20,26]. The primary morphological elements of PRP are yellowish-orange or pinkish-brown spiky follicular papules covered with tightly attached whitish bran-like or plate-like scales with fluffy hair in the center [7,14,24]. Large plaque patches are surrounded by single miliary nodules [13,18]. Provoking factors can cause worsening of the disease and generalization of the skin lesion.

In 1980, W.A. Griffiths proposed a PRP classification based on the clinical features and course of the disease [1,3,4,11]. Five clinical types (forms) were distinguished:

Type I: classical adult;
Type II: atypical adult;
Type III: classical juvenile;
Type IV: circumscribed juvenile;
Type V: atypical or nevoid juvenile.

DD is characterized by pronounced clinical polymorphism.

Classical adult type I PRP usually develops acutely in adults and presents with erythematous-squamous rash and multiple follicular papules. Their color is reddish-yellow or brown-red. Initially, the rash is located on the scalp, neck, upper half of the body, and upper and lower extremities. The rash is typically localized on the posteri-

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or aspects of the proximal phalanges, knee and elbow joints, and extensor aspects of the extremities. Palpation reveals the "nutmeg grater" symptom caused by follicular keratosis. The skin lesion spreads from the head to the feet. There may be erythroderma with pathognomonic islands of unaffected skin, which are rounded and strip-like in shape and 1 to 3 cm in size [3,5,8,9]. The clinical picture may also include palmoplantar hyperkeratosis and onychodystrophy (yellow nail plates, transverse or longitudinal striation, subungual hyperkeratosis).

Atypical adult type II PRP is characterized by a chronic course, prolonged exacerbations, and short-term remissions. This type occurs in 5% of DD patients. It can begin with erythematous-squamous and hyperkeratotic lesions on the extensor aspects of the wrist and elbow joints. In some cases, there are primary changes in the nail plates that become dull, striated, with onychogryphosis. Sometimes, there are eczematous skin lesions and hair thinning on the head. In this form, scales are larger and more ichthyosiform. Squamous plaques of various sizes are often formed from perifollicular papules. There may also be follicular keratosis lesions. This form is especially resistant to therapy.

Classical juvenile type III PRP develops during the 1st or 2nd year of life. Clinically, it presents similarly to classical adult type I, differing only by the age of patients [1,29]. This form differs from the adult type by a more frequent (2-fold) onset in the lower half of the body and a slow spread. There may be follicular hyperkeratosis in the phalanges of the fingers. Compared to the classical adult type, the clinical picture is less pronounced.

Circumscribed juvenile type IV PRP occurs during the first years of life, accounts for 25% of PRP cases, and is characterized by acquired sharply demarcated areas of follicular hyperkeratosis and infiltration on the skin of the knee and/or elbow joints, less often on other skin areas. On the palms and soles, there is thickening of the stratum corneum, sometimes cracks. In some cases, onychodystrophy occurs [10,30].

Abortive (weakened) type V of the Devergie's disease presents in many ways, in particular with palmoplantar hyperkeratotic plaques, rare follicular papules on the face, and, more often, squamous erythema [13,21,26]. Mucosal lesions are rare and similar to those of lichen planus [13,15]. The scalp, with rare exceptions, is not involved in the pathological process; in this case, rash appears as whitish scales as well as dystrophic hair changes. Sometimes, the clinical picture involves palmoplantar keratoderma [2,5,10]. Color of the affected skin varies from an orange to pink-red-brown hue [3,5,17,19].

Skin lesions in childhood can exist for years, without a tendency to spread. There is no seasonality in their chronic course [12,13]. There are spontaneous remissions and exacerbations [3]. General condition is not affected. Subjective sensations include itching, burning, and skin tethering of varying intensity [14]. The prognosis is uncertain. Sometimes, the disease resolves spontaneously in late adolescence. Histologically, this type is similar to psoriasis but without Munro microabscesses.

Atypical juvenile type V PRP develops in the first years of life and has a chronic course. The clinical picture is characterized by follicular hyperkeratosis associated with very slight erythema. In some patients, there are scleroderma-like (nevoid) changes in the skin of the upper and lower extremities. Often, this pathology is masked by other diseases, and the correct diagnosis can be established only after prolonged observation of the patient as well as based on the results of pathomorphological studies [6,16,31].

The variability of DD manifests as typical (classical), atypical (circumscribed), and erythrodermic forms [4]. Clinically, dermatosis is characterized by follicular hyperkeratosis, perifollicular erythema able to transform into erythroderma with islands of unaffected skin, scaly desquamation, and palmoplantar hyperkeratosis [10,12,24].

The differential diagnosis of the circumscribed DD form includes seborrheic dermatitis, psoriasis, atopic dermatitis, follicular ichthyosis, lichen planus, variable erythrokeratoderma, lichenoid parapsoriasis, T-cell lymphoma, HIV infection, and nonbullous congenital ichthyosiform erythroderma [1,2,4,27].

Although DD is not associated with pathognomonic histological changes, the pathomorphological picture is characterized by certain features. Granulosis, focal parakeratosis, and acanthosis are found in the epidermis. There may also be hydropic degeneration of the basal cell layer and keratotic plugs in the follicular openings; in the dermis, there may be moderate infiltration of melanocytes and histocytes located around the follicular blood vessels [2,4-6]. Some authors note certain discrepancies in the clinical and histological picture.

Treatment of DD is a challenge. It is advisable to use comprehensive treatment including a wide range of drugs. The first choice drugs are systemic retinoids and methotrexate. The treatment also includes vitamins A, E, and group B, glucocorticosteroids (local and systemic), keratomycotics, emollients, 2% sulfur-tar and salicylic acid ointments, and urea ointments as well as heliotherapy and thalassotherapy [2,13,23]. Sometimes, the disease resolves spontaneously [1,3,5]. In about 1/3rd of cases, dermatosis resolves in the period from several months to 5 years [10,13,21]. Warm full bath with sea salt or starch, followed by application of keratolytic ointments and phonophoresis of hydrocortisone cream with addition of aevit is useful. The prognosis for life is favorable; the prognosis for treatment is uncertain [5,22,25].

Clinical case

A 5-year-old female patient A. presented with a 4-year history of the disease. During the treatment, the following diagnoses were made: atopic dermatitis, psoriasis, follicular keratosis, xerosis, and streptoderma. The provided treatment resulted in temporary improvement (decreased

severity of clinical signs). In August 2017, the patient first presented to the Kolomensky Branch of the Moscow Scientific and Practical Center of Dermatology, Venereology, and Cosmetology of the Moscow City Health Department with complaints of rash on the skin of the buttocks and extremities, without subjective sensations. The patient's parents used topicrem and some cosmetics with a temporary positive result. No concomitant somatic diseases were detected. There was no dermatological history; *IgE* – 8.8 IU/ml. A Wood lamp examination revealed no pathologic fluorescence; the psoriatic triad was not detected; a test for mycelial fungal pathogens was negative; analysis of feces and scraping for helminths/enterobiasis revealed no pathology. Clinical parameters of blood and urine were within reference values.

Local changes: the pathological process had a chronic subacute inflammatory character, occurred on the extensor aspects of the elbows, knees, and buttocks **(Fig. 1, 2, 3)**, and was represented by follicular papules, sometimes merging into plaques with scaly-cortical layers, 3.5 to 5.5 cm in size, rounded-oblong in shape, brownish-brown in color with a faint yellowish hue, with a firm consistency on palpation. Dermographism was red.

The treatment included topical application of salicylic acid ointment 2%, topicrem, and pimafucort for one week. There was improvement in the affected areas. Subsequent therapy including topical corticosteroids, emollients, and salicylic acid/urea creams resulted in clinical remission of the disease.

The patient underwent a diagnostic pathomorphological examination at the day hospital of the Moscow Scientific and Practical Center of Dermatology, Venereology, and Cosmetology (Kolomensky Branch): epidermis with layered and compact ortho- and parahyperkeratosis areas alternating in vertical and horizontal directions, single neutrophils in parakeratotic masses, agranulosis areas, and minor acanthosis **(Fig. 4, 5)**. In the upper dermis, there were perivascular lymphohistiocytic infiltrates and plethora of blood vessels. Conclusion: histological findings suggest psoriasis; Devergie's disease cannot be ruled out either.

The presented case is of particular clinical interest due to the rarity and variety of clinical signs of the observed dermatosis.

The authors declare no conflicts of interest.

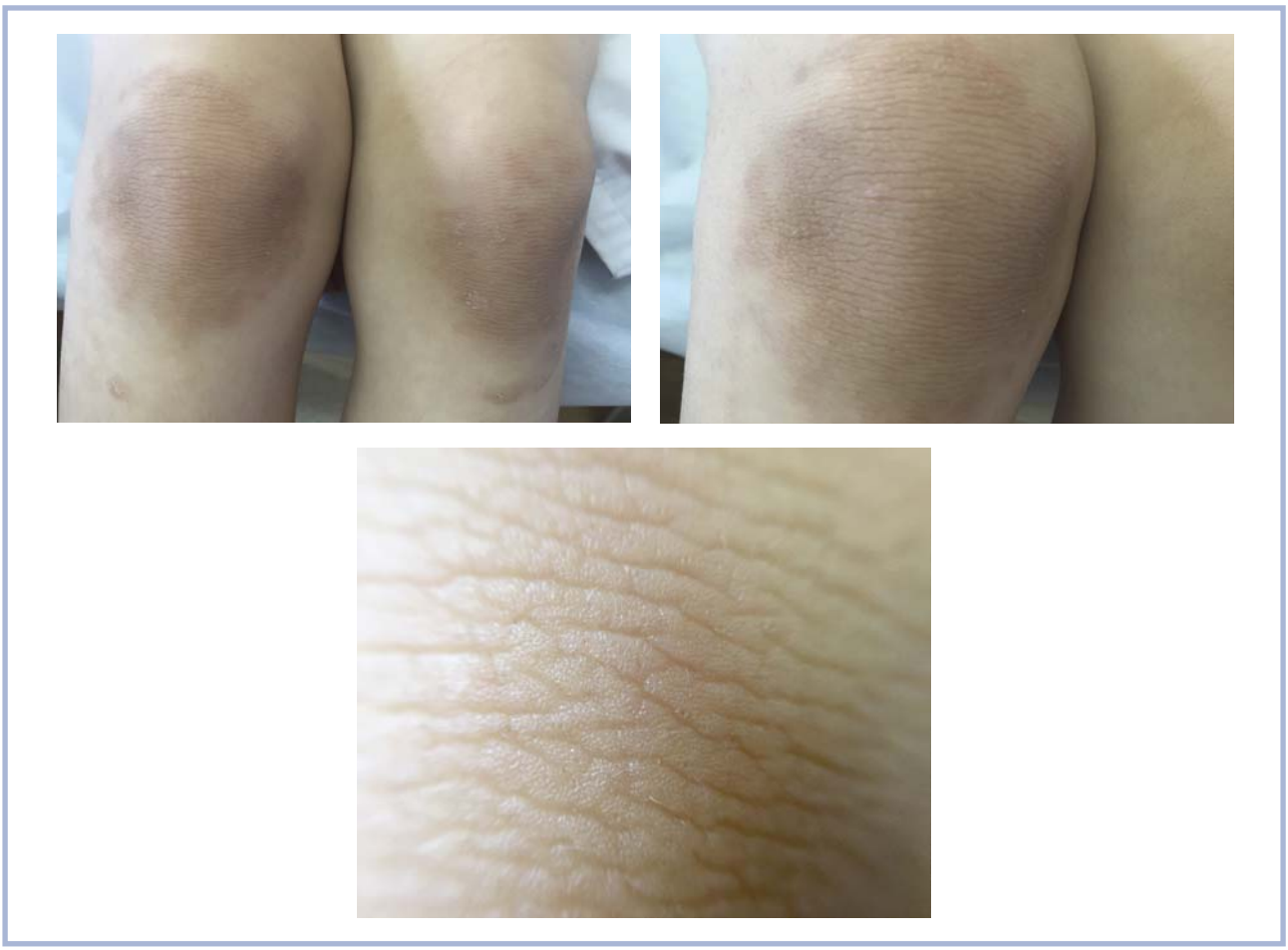

Рис. 1. **Очаги поражения кожи коленных суставов.**

Fig. 1. **Lesions on the skin of knee joints.**

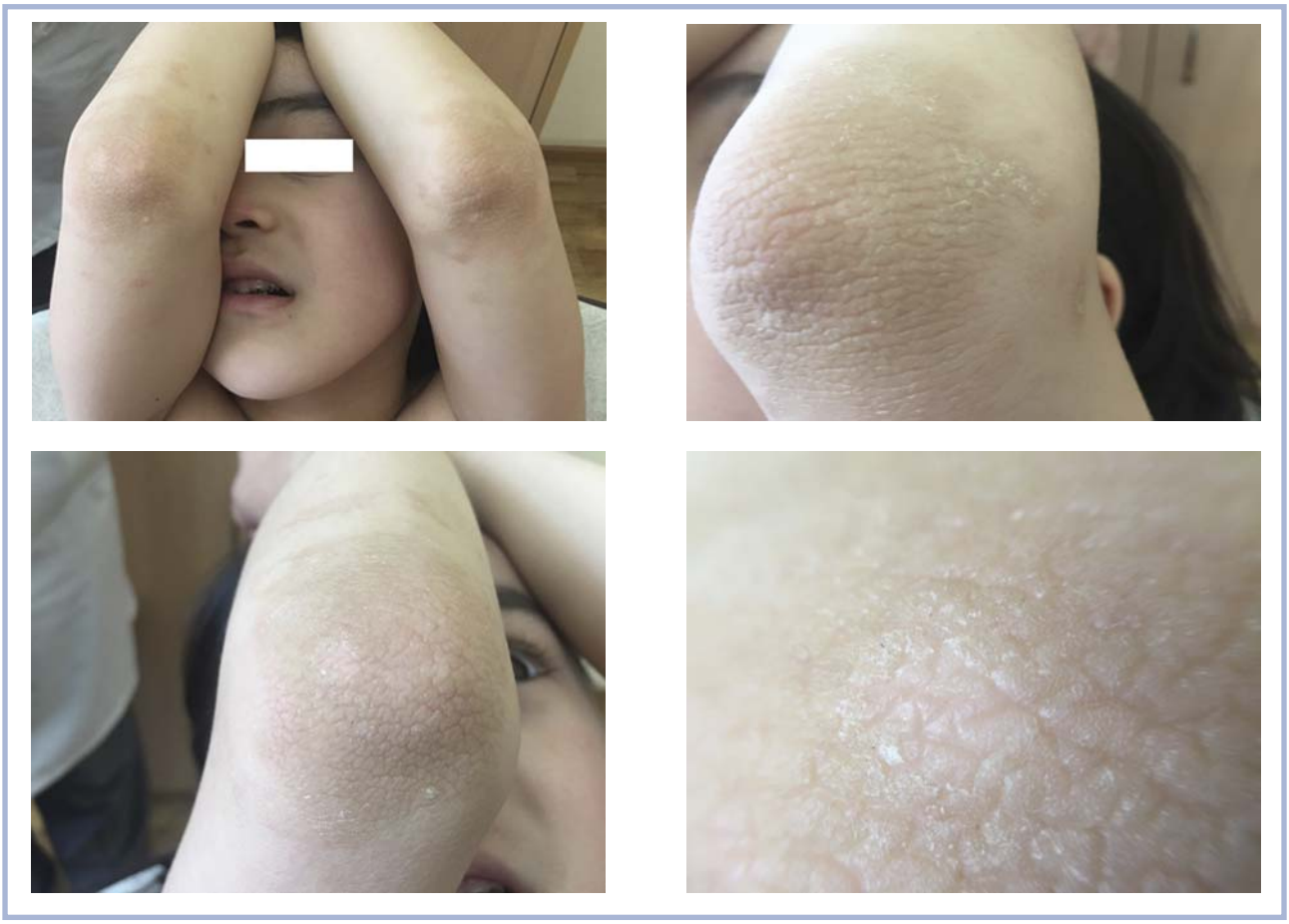

Рис. 2. **Патологический процесс на коже локтевых суставов.**
Fig. 2. **Pathologic process on the skin of elbow joints.**

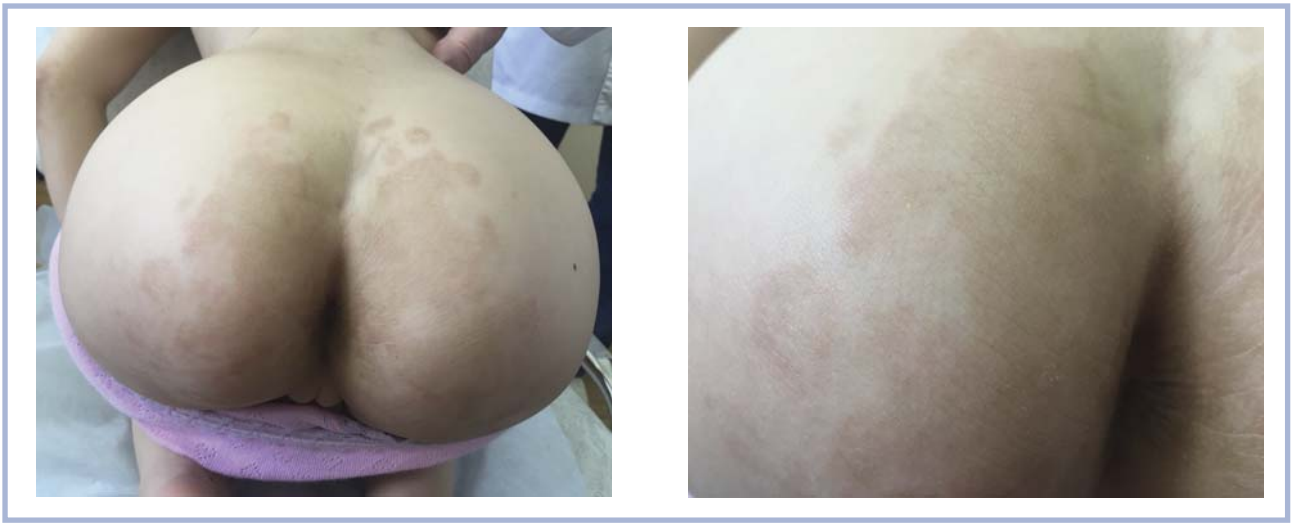

Рис. 3. **Редко наблюдающиеся поражения кожи ягодиц.**
Fig. 3. **Rare lesions of buttock skin.**

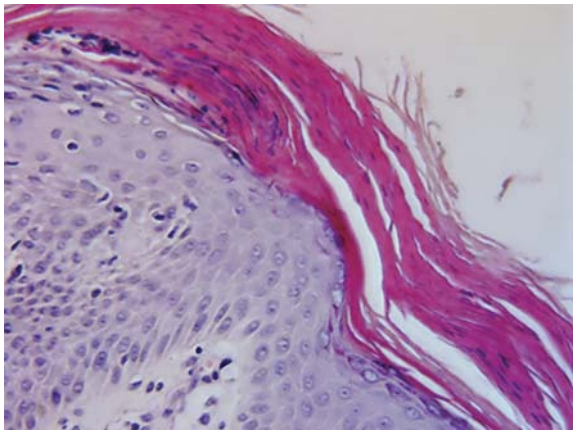

Рис. 4. **Участок с агранулезом, паракератозом и нейтрофилами в роговом слое по краю биоптата (окраска гематоксилином и эозином, ув. ×400). Изменения характерны для псориаза, но также нельзя исключить вторичный характер изменений (травма, присоединение вторичной инфекции). Рекомендована повторная биопсия для исключения/подтверждения псориаза.**

Fig. 4. **Area of agranulosis, parakeratosis and neutrophils in stratum corneum on the edge of biopsy specimen. These lesions are more characteristic of psoriasis, but secondary nature of these lesions cannot me excluded (trauma, secondary infection). Another biopsy is recommended to exclude or confirm psoriasis.**

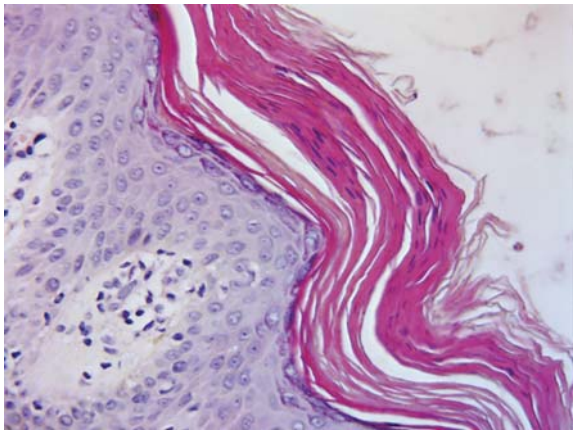

Рис. 5. **Характерный для болезни Девержи морфологический признак — чередование участков орто- и парагиперкератоза в вертикальном и горизонтальном направлениях (окраска гематоксилином и эозином, ×400).**

Fig. 5. **Morphological sign that is characteristic of the Devergie disease — alternation of areas of ortho- and parahyperkeratosis in vertical and horizontal directions (hematoxylin, eosin, ×400).**

ЛИТЕРАТУРА/REFERENCES

1. Иванов О.Л. *Розовый лишай Девержи. Кожные и венерические болезни*. М., 2007:154-155.
   Ivanov OL. *Pityriasis rubra pilaris Devergie. Cutis and venerol diseases*. M., 2007:154-155. (In Russ.).
2. Wolff K. *Pityriasis rubra pilaris*. Fitzpatrick's Dermatology in General Medicine.7th edition, McGraw. New Jork, Chicago, Lisbon, London, Moscow, 2012:251-254.
3. Hurwitz S. *Devergie's disease. Clinical pediatric dermatology*. WB Saynders company, Philadelphia, London, Toronto, Sydney, Tokyo, 1993:118-120.
4. Суколин Г.И. *Болезнь Девержи. Клиническая дерматология*. СПб., 1997:101-102.
   Sukolin GI. *Devergie's disease. Clinical dermatology*, St. Peterburg, 1997:101-102. (In Russ.).
5. Елькин В.Д., Митрюковский Л.С., Седова Т.О. *Отрубевидный волосяной лишай. Избранная дерматология*. Пермь, 2004:386-388.
   El'kin VD, Mitrukovskiy LS, Sedova TO. *Pityriasis rubra pilaris. Selected dermatology*. Perme, 2004:386-388. (In Russ.).
6. Cohen PR. Clinical current of Pityriasis rubra pilaris. *J. Am. Acad. Dermat.*1989;24:801-807.
7. Сафонова А.В. Острый вариант болезни Девержи у взрослого. *Рос журн. кож. вен. бол.* 1999;5:59-60.
   Safonova AV. Acute variant of Devergie disease in adult. Rus. *J. cutis vener. diseases*. 1999;5:59-60. (In Russ.).
8. Заварина И.В., Гришко Т.Н. Болезнь Девержи у детей. *Вестн дермат венерол*. 2003;1:10-12.
   Zavarina IV, Grishko TN. Devergie disease in childhood. *Vestn. Dermat. vener.* 2003;1:10-12. (In Russ.).
9. Молочков В.А., Сапронова Т.И. Лечение тигазоном. *Вестник дерматологии и венерологии*. 1999;3:27-29.
   Molochkov VA, Sapronova TI. Tigazon therapy *Rus. J. Derm. vener. diseases*.1999;3:27-29. (In Russ.).
10. Хлебникова А.Н. Увлажняющие средства в комплексном лечении красного волосяного лишая Девержи. *Вестник дерматологии и венерологии*. 2010;1:45-50.
    Khlebnikova AN. Moisturizing agents for combined treatment of Devergie disease. *Klin. Derm. vener.* 2010;1:45-50. (In Russ.).
11. Griffiths WA. Pityriasis rubra pilaris: the problem of its classification. *J. Am. Acad. Dermat*. 1992;1:140-142.
12. Скрипкин Ю.К. и др. *Красный волосяной отрубевидный лишай. Руководство по детской дерматовенерологии*. Ленинград, 1983:209-215.
    Skripkin YuK, et all. *Pityriasis rubra pilaris. Guide on children dermatology*. Medicine, Leningrad, 1983:209-215. (In Russ.).
13. Скрипкин Ю.К., Бутов Ю.С. *Красный волосяной лишай. Клиническая дерматология*, М., 2009;т.2:740-742.
    Scripkin YuK, Butov YuS (eds). *Pityriasis rubra pilaris. Clinical Dermatology*. M., 2009;v2:740-742. (In Russ.).
14. Braun-Falco D et al. Pityriasis rubra pilaris: clinicopatological and therapeutic study. *Arch.derm.res*. 1983;275,5:287-295.
15. Katsambas AD. *Pityriasis rubra pilaris*. European Handbook of Dermatological Treatments. 2008:267-270.
16. Shore K, et al. *Pityriasis rubra pilaris*. Color atlas and Synops. of pediatric dermatol. M., 2011:88-90.
17. Галямова Ю.А. *Красный волосяной лишай Девержи. Избранные лекции по дерматологии*. М., 2007; т.1:143-156.
    Galjamova YuA. *Pityriasis rubra pilaris, Devergie. Selected lections on dermatol*. M., 2007;v1:143-156. (In Russ.).
18. Мордовцев В.Н., Суворова К.Н. *Красный волосяной лишай. Наследственные заболевания кожи*. 1995:347-355.
    Mordovcev VN, Suvorova KN. *Pityriasis rubra pilaris. Genetic cutis diseases*. 1995:347-355. (In Russ.).
19. Levene GM, Calman CD. *Pityriasis rubra pilaris*. A color atlas of dermatology. Wolfe medical publications. 1974:200-201.
20. Darier J. *Pityriasis rubra pilaris*. Precis de Dermatologie. Moscow-Leningrad. 1930:511-514.
21. Cohen BA. *Pityriasis rubra pilaris: a review of diagnosis and treatment. Pediatric Dermatology*. 4th edit. M., 2015:115-116.
22. Griffiths AD. Pityriasis rubra pilaris. *Clin.Exp.derm*.1980;5:105-112.

23. Каламкарян А.А., Мордовцев В.Н., Трофимова Л.Я. *Красный волосяной отрубевидный лишай Девержи. Клиническая Дерматология.* Ереван, Айстан, 1989:284-289.
Kalamkaryan AA, Mordovtsev VN, Trofimova LYa. *Pityriasis rubra pilaris Devergie Klinical dermatol*. Yerevan, Aistan, 1989:284-289. (In Russ.).

24. Мордовцев В.Н. и др. *Красный волосяной отрубевидный лишай Девержи. Наследственные болезни и пороки развития кожи*. М., 2004:21-22.
Mordovtsev VN et al. *Pityriasis rubra pilaris Devergie. Genetic diseases and malformations of the skin*. M., 2004:21-22. (In Russ.).

25. Романенко И.М. и др. *Красный волосяной отрубевидный лишай Девержи. Лечение кожных и венерических болезней*. Медицинское информационное агентство. М., 2006:429-431.
Romanenko IM, et al. *Pityriasis rubra pilaris Devergie, Treatment of skin and venereal diseases*. M., 2006:429-431. (In Russ.).

26. Гребенюк В.Н. и др. Красный отрубевидный волосяной лишай у девочки 6 лет. *Клин дерматол венерол*. 2018;2:38-40.
Grebenjuk VN, et al. Pityriasis rubra pilaris in 6 years old girl (case study). *Russian Journal of Clinical Dermatology and Venereology = Klinichaskaya dermatologiya i venerologiya*. 2018;2:38-40. (In Russ.).

27. Elenor ES. *Devergie's disease in 3 years old boy*. Dermatology pearls. St. Petersburg, 2001:192-194.

28. Кубанова А.А., Арифов С.С. Особенности клинического течения красного отрубевидного волосяного лишая. *Вестн дерматол*. 1990;7:59-61.
Kubanova AA, Arifov SS. Particularities of clinical current of Pityriasis rubra pilaris. *Vestn.dermatol*. 1990;7:59-61. (In Russ.).

29. Олесова О.Ю., и др. Эритродермическая форма болезни Девержи. *Рос журн кож вен бол*. 2014;1:18-20.
Olisova OYu, et al. The erytrodermal form of Devergie's disease. Rus. *J. cutis and veneral diseases*. 2014;1:18-20. (In Russ.).

30. Кряжева С.С., и др. Особенности клинического течения и лечения красного отрубевидного волосяного лишая. *Вестн дерматовенерол*. 1992;8:4-9.
Kryazheva SS, et al. Particularities of clinical current and therapy of Pityriasis rubra pilaris Devergie. *Vestn. Dermatol. vener.* 1992;8:4-9. (In Russ.).

31. Чистякова И.А., Альбанова В.И. Клинико-морфологическая характеристика красного отрубевидного волосяного лишая Девержи. *Вестн дерматовенерол*. 1992;8:4-9.
Chistjakova IA, Albanova VI. Clinico-morphologic characteristic of Pityriasis rubra pilaris Devergie. *Vestn. dermatol. vener.* 1992;8:4-9. (In Russ.).

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