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CLINICAL CASE OF LYMPHOMATOID PAPULOSIS

Vasyl Tkach[1], Oleksandr Aleksandruk[1], Marian Voloshynovych[1], Galyna Girnyk[1], Ivan Kostyshyn[2], Nataliia Kozak[1]

[1]Department of Dermatology and Venereology
Ivano-Frankivsk National Medical University
[2]Department of Oncology
Ivano-Frankivsk National Medical University
mvoloshynovych@gmail.com

Abstract. Lymphomatoid papulosis is a rare skin disease. The incidence averages 1.5 cases per 1 million population. It is the most common in adults around the age of 40. It was previously considered as paraneoplastic dermatosis, but recent studies have confirmed that lymphomatoid papulosis is a primary CD30 + T-lymphoma of the skin. Despite the aggressive morphological features, it is benign. The disease is characterized by a tendency to spontaneous regression. Typically, skin lesions last 3-12 weeks, although in some severe cases they may last longer. Skin lesions may disappear or recur over decades. Patients require observation because a second lymphoproliferative disease develops in 10-40% of patients.

A case of lymphomatoid papulosis in a patient with diffuse astrocytoma is presented. The patient was treated with systemic retinoids. The results of differential diagnosis and additional research methods, including dermoscopy, are given. Although the use of dermoscopy was not crucial, the results of the research showed microscopic visual differences between the elements of the rash, depending on the stage of the disease. The picture was mostly typical, however, spots formed by the areas of central erythema on the background of light brown areas, with a predominance of the reticular vessels pattern were also noted in addition to the rash elements on the hairy skull. Clarification of this feature requires further study.

The purpose of our report is to draw the attention of dermatologists and family doctors to cases of rare dermatoses. The rareness of the disease, lack of knowledge on the clinical findings lead to diagnostic errors, "inadequate treatment".

Keywords: lymphomatoid papulosis, clinical findings, differential diagnosis, treatment.

Background

Lymphomatoid papulosis (LP) is a rare skin disease first described in 1968 by W. L. Mocaulay (1). The incidence averages 1.5 cases per 1 million population. It is the most common in adults around 40 years of age, but it may develop in all age groups (2,3). This disease belongs to the group of cutaneous T-cell and NK-cell lymphomas in the updated WHO-EORTC classification 2018 (4). It was previously thought to be paraneoplastic dermatosis, probably caused by oncoviruses, but recent studies have confirmed that LP is a skin T-lymphoma, including primary CD30 + lymphoproliferative disease (5,6). Despite its aggressive morphological features, it is benign, usually without subjective sensations.

We consider the term "papulosis" to be not quite accurate, because atrophic scars remain after the regression of these papules, and the disease is also manifested by nodules, ulcers in addition to the papules. Clinically papular form is similar to parapsoriasis guttata. Lesions ranging in size from a few millimeters to 2 cm, red with a bluish tinge, with noticeable necrosis in the center appear on the skin, mainly on the extremities, torso.

The disease is characterized by a tendency to spontaneous regression, which is crucial for establishing a clinical diagnosis. Typically, the lesions last 3-12 weeks, although in some severe cases they may last longer. They may disappear or recur over decades (7). Patients require a follow-up because a second lymphoproliferative disease, such as CD30-positive anaplastic large cell lymphoma, Hodgkin's lymphoma, or fungal mycosis (8) develops in 10-40% of patients.

Case Presentation

Patient K, born in 1995, came to the clinic with common rashes on the torso, limbs, hairy skull. The lesions did not bother the patient, sometimes he felt a slight itching. He has been ill for 10 years, since the first time when isolated rashes appeared on his limbs and hairy skull. At that time, the child suffered from dizziness, rarely general weakness. For a long time, he did not consult the doctors, the rash slowly progressed. At the age of 20, he periodically suffered from headaches, dizziness became more frequent, mild seizures were rare, and the number of rashes markedly increased. The patient consulted dermatologists who diagnosed allergic dermatitis, psoriasis, parapsoriasis, the prescribed treatment was not effective.

A seizure with loss of consciousness happened in 2020. The patient was hospitalized in the All-Ukrainian Center for Radiosurgery of the Feofania Clinical Hospital. The examination detected an intracerebral tumor of the temporal lobe of the brain. Bone-plastic trepanation in the right temporal area with removal of intracerebral tumor was performed. The histopathological picture and immunohistological data corresponded to diffuse

astrocytoma. During the treatment at the clinic, he was examined by a dermatologist who confirmed the diagnosis of psoriasis and prescribed traditional therapy. The number of rashes did not decrease, on the contrary, the number of rashes increased after the remote radiation therapy at the site of the removed tumor, on the background of dexamethasone and anti-edematous agents' introduction.

On examination, unaffected skin was of normal color, turgor and elasticity were preserved, perspiration and sebaceous excretion were moderate. Normotrophic postoperative scar was observed on the skin of the right temporal area (Figure 1). Multiple red papular elements with a bluish tinge of a lentil grain size, hemispherical were observed on the hairy skull (Figure 2), front and back of the torso (Figure 3), limbs, the gray crust was noted in the center of the individual elements. Spots and single stamped scars were found along with papular rashes. The rash did not bother the patient creating only cosmetic inconvenience. Peripheral lymph nodes were not enlarged.

The use of dermoscopy in case of LP is not crucial, but descriptions of research results indicating visual structural differences at the microscopic level between the elements of the rash, depending on the stage of development of the lesion can be found in the literature. In particular, homogeneous pink and light brown areas are observed at the initial stage, where pinpoint or tortuous vessels with irregular distribution, surrounded by whitish zones, are determined. The unstructured center, deprived of vascular elements, covered with scales or crusts, with a tendency to the appearance of peripheral hyperkeratosis is formed in more mature elements. In the future, the papules show a necrotic center with crusts and scales. Scars devoid of vascular pattern or brown spots remain in the place of the elements (9,10).

The disease manifestations of our patient mostly corresponded to the described morphology. However, spots formed by the areas of central erythema, on a background of light brown areas, with a predominance of reticular vessels pattern were also noted among the rash elements on the hairy skull (Figure 4). Single areas with a well-visible vascular pattern, similar in nature, but with less marked blood flow were detected between the spots. This picture suggested a possible active blood supply typical of the hairy skull skin of microcirculatory bloodstream in the areas of LP localization and can hardly be considered as a dermoscopic feature of the early stage of the disease in this location. Clarification of this feature requires further study.

The results of general and biochemical analysis of blood were within normal limits. Creatinine was slightly increased and constituted 120 mmol/l, AST – 50 U/l, cholesterol - 6.0 mmol/l, C-reactive protein ++.

A biopsy of the affected skin was taken. Para- and hyperkeratosis was found in the epidermis, leukocytes were observed in parakaryotic masses, cytoid cells and necrosis of keratinocytes were detected in the basal parts of the epidermis. Diffuse lymphocytic infiltrate of small and medium lymphocytes was found in the dermis. Large, atypical cells in the stage of mitosis, Reed-Sternberg cells were observed in the papillary and reticular layers. The vascular endothelium was swollen, the walls were thickened, the lumen was narrowed, obliterated in some areas. Perivascular tissue was swollen, infiltrated by eosinophils and neutrophils, histocytes, lymphocytes. Histochemical examination of infiltrate cells revealed CD30 + cells.

We diagnosed LP based on the clinical findings and the results of histopathological and histochemical studies. Considering the ineffectiveness of corticosteroids (administration of dexamethasone in the postoperative period), the patient was prescribed retinoids, namely isotretinoin 8 mg per day, vitamin therapy, hepatoprotectors, phototherapy. The process stabilized after a month of the treatment, no new elements appeared, a noticeable regression of papular elements was observed (Figure 5). The patient continued to take isotretinoin, complete blood count and biochemical blood test were monitored every 2 weeks.

Conclusions

Clinically, LP is similar to parapsoriasis, psoriasis, but the tendency of rashes to regression resembles lymphogranulomatosis.

The primary element in case of parapsoriasis guttata is papule of round or oval shape, light brown, sometimes brownish-red, similar to LP elements. The duration of the rash also brings these two dermatoses together. Parapsoriasis foci as wells as LP foci may exist during months. However, remissions occur in case of parapsoriasis, more often in summer. Parapsoriasis is characterized by three phenomena. A hidden peeling occurs in case of papules grattage, wafer symptom and purpura phenomenon are noted in case of more intensive scraping. The patient's general condition is affected, the body temperature rises in case of parapsoriasis acute form.

Psoriasis is characterized by a psoriatic triad: stearin stain, terminal film, pinpoint bleeding, as well as staging of the course. Stress, focal infection cause new rashes.

Lymphogranulomatosis is a malignant tumor of the lymphatic system. Both men and women at the age of 30-40 (age of our patient) can be ill. It usually begins with an increase in cervical, rarely mediastinal, retroperitoneal lymph nodes. In addition to papular elements similar to LP, there are erythematous spots on the skin, urticaria, later tumors, ulcers, possibly erythroderma.

The results of histological and histochemical studies are crucial in the differential diagnosis.

The purpose of our report is to draw the attention of dermatologists and family doctors to cases of

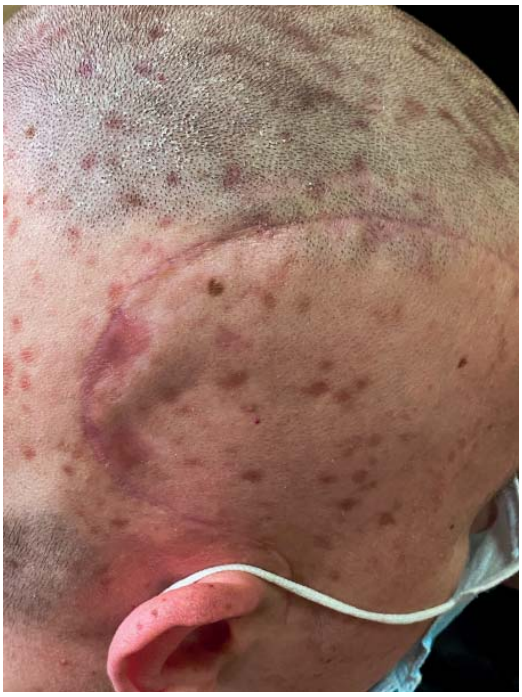

Figure 1. Normotrophic postoperative scar on the skin of the right temporal area. Multiple maculo-papular elements

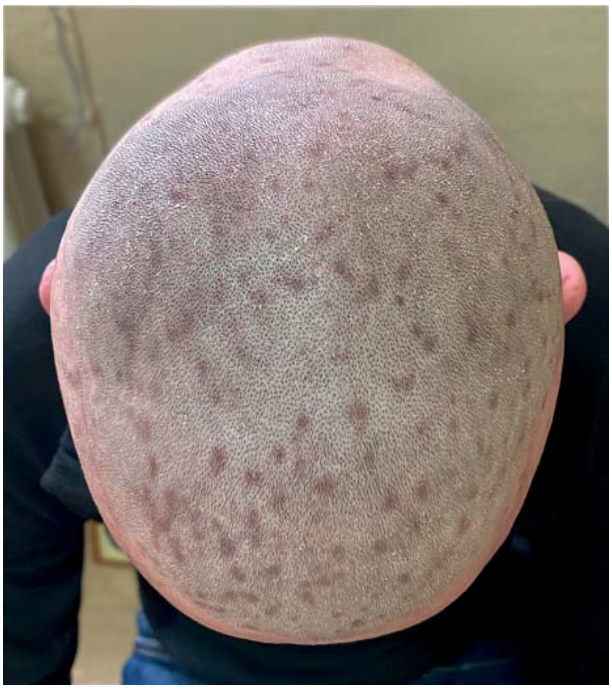

Figure 2. Multiple maculo-papular elements on the hairy skull

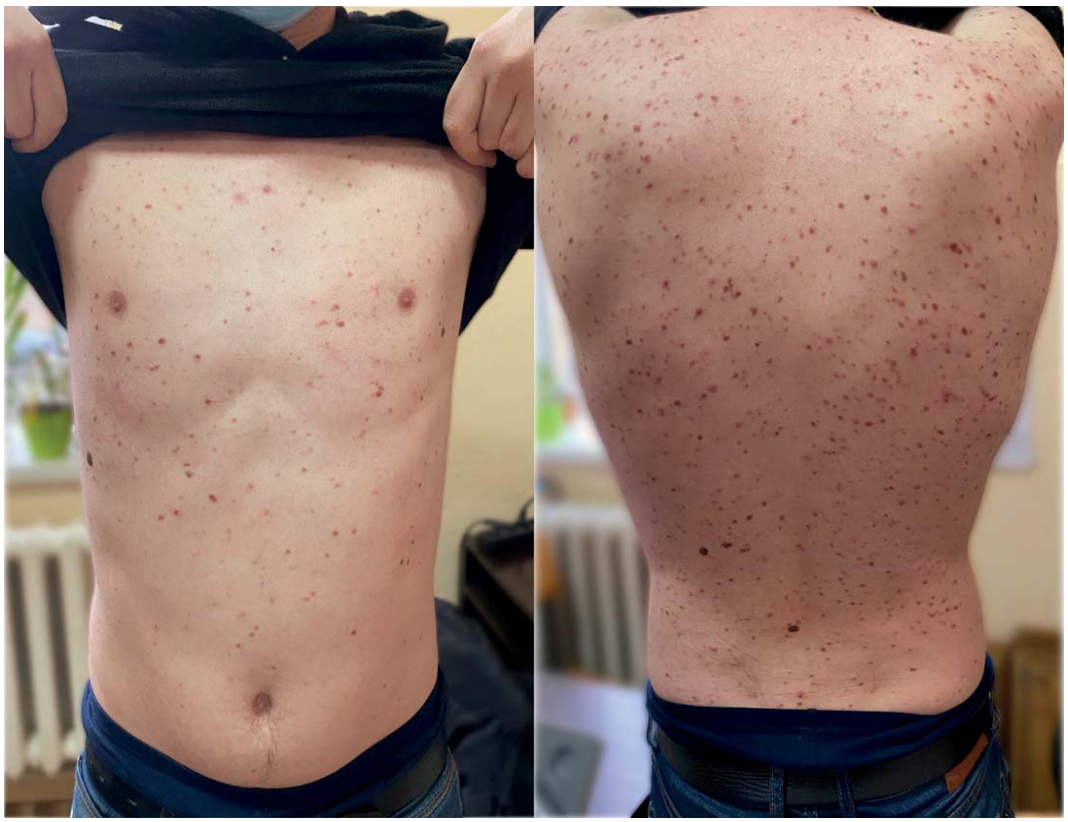

Figure 3. Multiple maculo-papular elements on the anterior and posterior surface of the torso

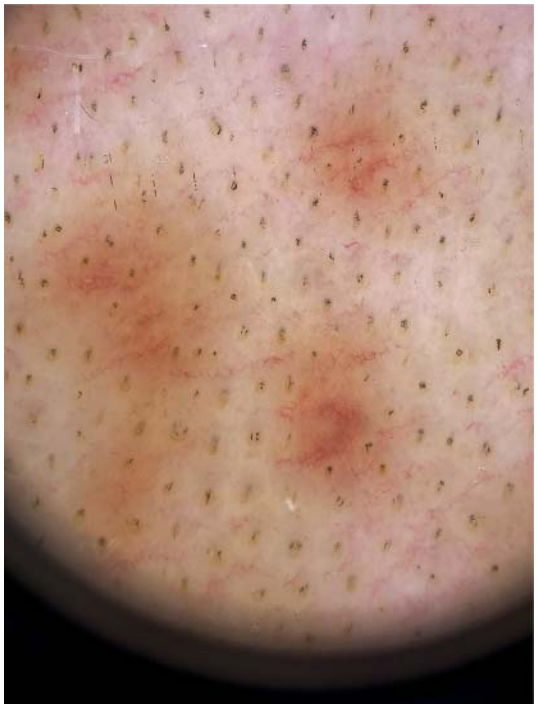

Figure 4. Dermoscopy using immersion fluid. The area of the hairy skull. Spots, central erythema, on a background of light brown areas, with a predominance of the reticular vessels pattern

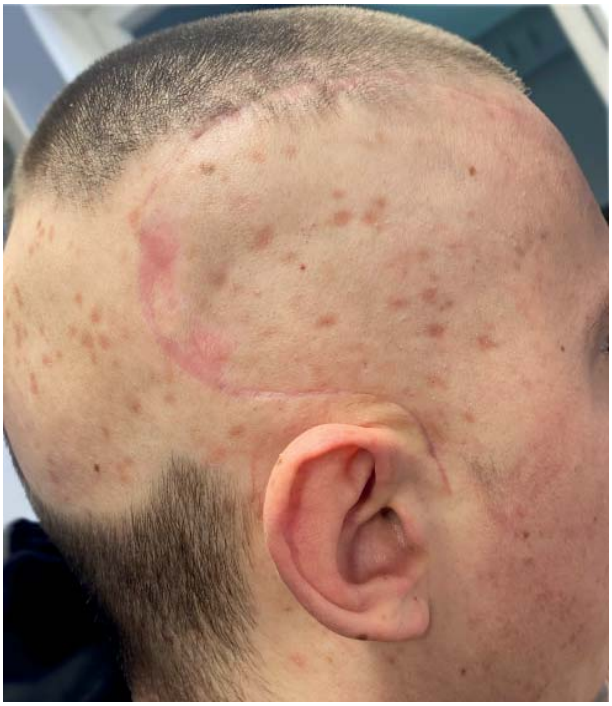

Figure 5. The condition of the skin of the right temporal area after a month of the treatment

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Inform consent of the patient is received at the start of contact and before any procedures.

Conflict of interest: The authors declare no conflicts of interest.

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