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Ulcerative Sarcoidosis: Confusing Case Report

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Abstract
Background: Along with the most frequent and characteristic (typical) forms of specific cutaneous sarcoidosis, there are a great variety of clinically atypical granulomatous cutaneous lesions have been described, accordingly, rarely diagnosed, which are usually observed in patients with visceral sarcoidosis. There are many atypical forms of the disease, which complicates the clinical diagnosis of sarcoidosis, the diagnosis of which can be made as a result of histopathological examination. The literature describes in sufficient detail the morphological criteria for the histopathological diagnosis of the granulomatous process of sarcoidosis, which will be identical both in typical and atypical forms of sarcoidosis. In our article, we present difficulties in the diagnosis of atypically occurring sarcoidosis of the skin even with a morphological study. Diagnosis was hindered by the clinical similarity of this disease to lupoid cutaneous leishmaniasis in a 58-year-old patient. The final diagnosis was made only after corticosteroid therapy.

Key words: Atypical cutaneous sarcoidosis, differential diagnosis, lupoid cutaneous leishmaniasis, treatment, observation.

Background

Sarcoidosis is a chronic systemic disease of unknown etiology, the pathomorphological basis of which is the epithelioid-cell granulomatous structure. Most often, the lungs are affected with this disease (up to 90%), however, skin manifestations are observed in about 1/3 -1/2 of patients with sarcoidosis, but a variety of organs and tissues can be involved in the pathological process [1,2]. From a diagnostic point of view, at present, “Sarcoidosis” is a clinical diagnosis, since there are no reliable (specific) diagnostic tests, as a result of which the diagnosis is often delayed for months or even years. Taking into account a very diverse clinical picture and variability of morphology, Sarcoidosis is deservedly called in dermatology “a great imitator”. To make a diagnosis, clinical, epidemiological, radiological, laboratory and histological criteria must be considered. Therefore, Sarcoidosis's possibility should be considered in all patients with skin manifestations and concomitant pulmonary symptoms, granulomatous inflammation, the cause of which has not been established (infection and foreign body are excluded) [3-9]. According to modern concepts, Sarcoidosis is a disease of immunoreactivity disorder with a special reaction of the body to various factors, an important role is played by heredity (association with HLA-1, HLA-A2, HLA-B7, HLA-B8 and HLA-DR3 alleles and, more recently, HLA-DRB1 and DQB18 haplotypes) [5,8].

Clinical features are determined by the nature of organ damage. Skin manifestations with sarcoidosis can develop before the appearance of systemic involvement, with it or after it. Traditionally, pathological elements are classified as specific, with a biopsy revealing granulomas without necrosis (non-caseating granulomas), and nonspecific, which are reactive changes (usually erythema nodosum). Specific skin manifestations are usually asymptomatic, most often located on the scalp and neck, however, they can occur on any part of the skin or mucous membranes and have a symmetrical or asymmetric distribution. Almost all morphological types of lesions are described (spots, papules, plaques and nodes, also scaly, telangiectatic or atrophic changes)[3]. Despite the diversity of appearance, several clinical options are distinguished. Typical forms include: dermal sarcoid Beck (cutaneous sarcoidosis Boeck in three of its varieties: large nodular, small nodular, and diffuse-infiltrative), lupus pernio (Besnier-Tenneson syndrome), angiolupoid Broca-Potrier, Darier–Roussy disease (subcutaneous sarcoids) [1,2,10].

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In addition to the listed typical clinical variants of cutaneous sarcoidosis, atypical forms are distinguished, which are extremely rare. Clinically, they usually simulate one or another dermatosis, which is reflected in their names: lichenoid sarcoid, resembling lichen planus; verrucous and papillomatous; ichthyosiform and psoriasiform; anular and flat circinar forms, clinically resembling basal cell carcinoma and lupus erythematosus, mycosis fungoides; a form clinically indistinguishable from papulo-necrotic skin tuberculosis; post-traumatic (cicatricial), hypopigmented, erythrodermic, morpheaform lesions [1, 3,6].
Among the atypical forms of skin sarcoids, an ulcerative variety is often described. Frequency occurs worldwide only in 5% of patients with cutaneous sarcoidosis. Ulcers form mainly within the primary small and coarse site. Pronounced soreness is almost always noted. A histological examination of ulcerated sarcoid papules reveals a superficially located nonspecific inflammatory infiltrate and typical epithelioid cell granulomas in the deep layers of the dermis. In the pathogenesis of the ulcerative form of sarcoidosis of the skin attach importance to cooling, pressure, venous stasis, hypertension, diabetes mellitus; however, these factors appear to play a secondary role [1,6,7,9].

Case Report

A 58-year-old man, an uzbek, turned to a consultative appointment, with a directive diagnosis of chronic ulcerative pyoderma. Considers himself ill for 5 years, does not associate his disease with anything. The disease began with a small formation on the right back side, which after 2 months began to ulcerate. He underwent treatment at the place of residence with a diagnosis of ulcerative pyoderma. He does not remember the prescription and treatment, but he did not notice any improvement after the therapy. The process began to progress, new elements began to appear on the body, which also began to ulcerate. In the summer of 2018, he received treatment at a dermatology center with a diagnosis of lupoid cutaneous leishmaniasis. The patient received specific treatment, however, as such, the result after treatment was not observed. The process began to spread further. Ulcer elements have increased in diameter. After which the patient in January 2020 came to us for an appointment.

Complaints: rash and severe soreness of the elements and general weakness. Objectively: the general condition of the patient is relatively satisfactory, the position is constrained. From the side of internal organs - breathing is hard in the lungs, isolated rales. The borders of the heart are deflected to the left. Status localis: a cutaneous pathological process is localized on the skin of the chest, back, abdomen, upper and lower extremities (Fig. 1a, b).

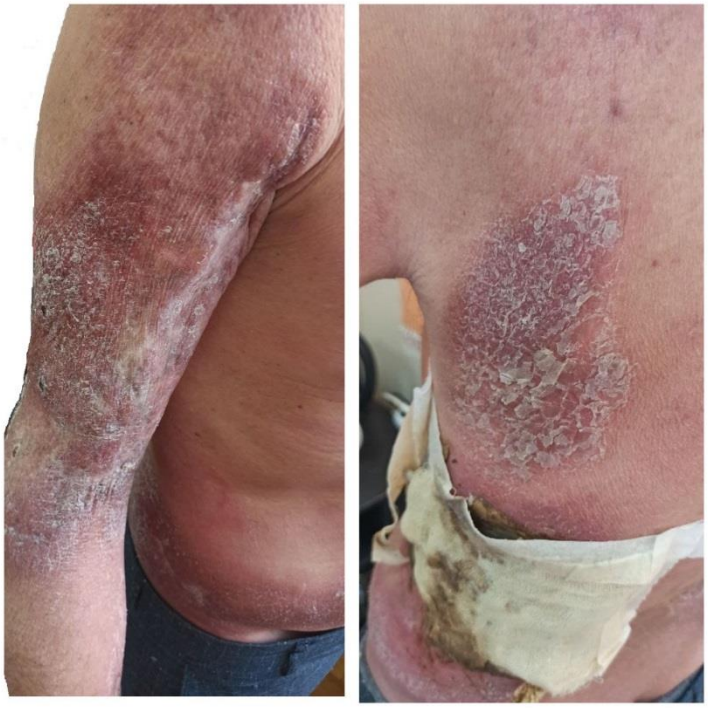

Fig. 1 a,b Plaques covered with scales

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Date of Publication:05-12-2021

In the area of the left lateral torso, a deep ulcer (15 * 20 * 20) of irregular round shape with distinct undercut edges, with serous-purulent discharge, and granulation tissue is noted at the bottom. The same ulcers are noted in the area of the right elbow joint, lower limbs, on the surface of some infiltrative foci (Fig. 1c).

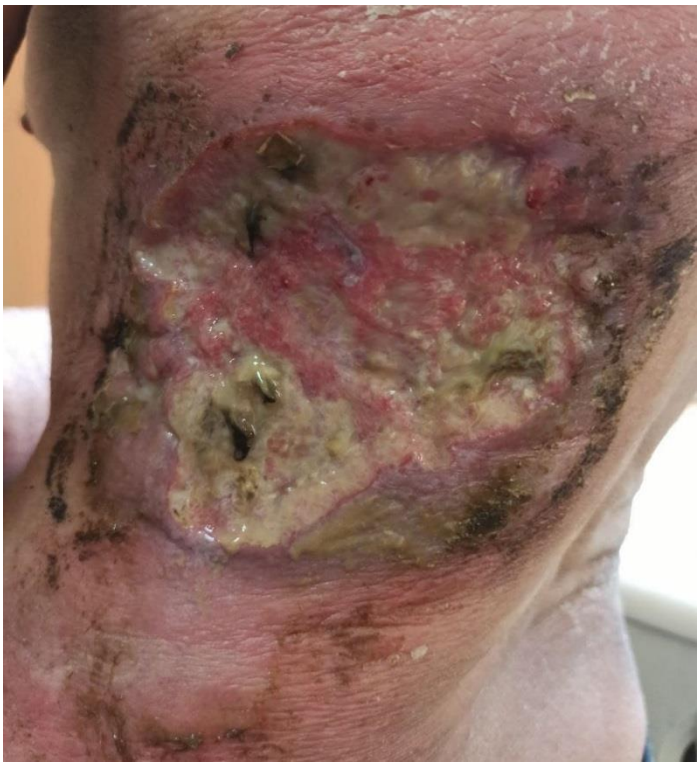

Fig 1c Ulcer on the left side of the torso

Infiltrative plaques up to 15 cm in diameter, irregularly oval in shape, with fuzzy borders of stagnant violet color, peeling on the surface, tight-fitting crusts are noted on the trunk and extremities. Around some plaques with lateral illumination, small tubercles are visible (Fig. 1d).

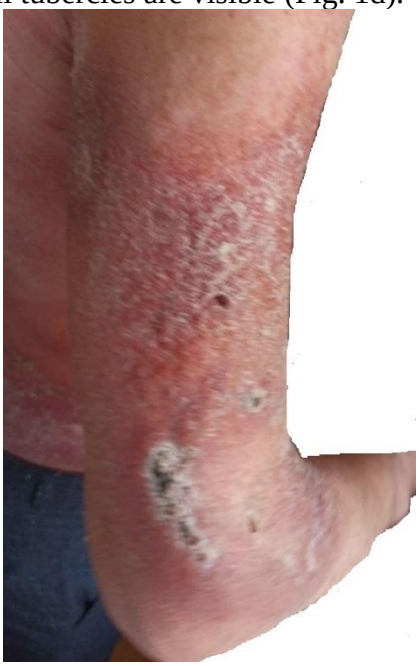

Fig 1 d Plaques and tubercles on the extensor surface of the right hand

Upon diascopy, the element acquires a yellow-brown color - the symptom of "apple jelly" is weakly positive, interspersed with yellow-golden dots are visible - the "dust particles" phenomenon. Multiple atrophic scars, secondary hypo- and hyperpigmentation, xerosis are also noted on the body.

ISSN NO: 2770-2936
Date of Publication:05-12-2021

The patient was hospitalized in a hospital with a diagnosis of Skin sarcoidosis. It was recommended: observation in dynamics and exclusion of diagnoses: lupoid leishmaniasis of the skin, mycosis fungoides, cutaneous tuberculosis.

The results of a biopsy of the plaques showed hyperkeratosis, acanthosis with densification of the epidermal processes, in some places atrophy of the epidermis. In the dermis, lymphohistiocytic infiltration around the vessels, appendages; in places with the penetration of cells of the infiltrate a epidermis. Infiltrate cells: lymphocytes, histiocytes, epithelial cells, neutrophils, in places Langerhans-type giant cells (Fig. 2a).

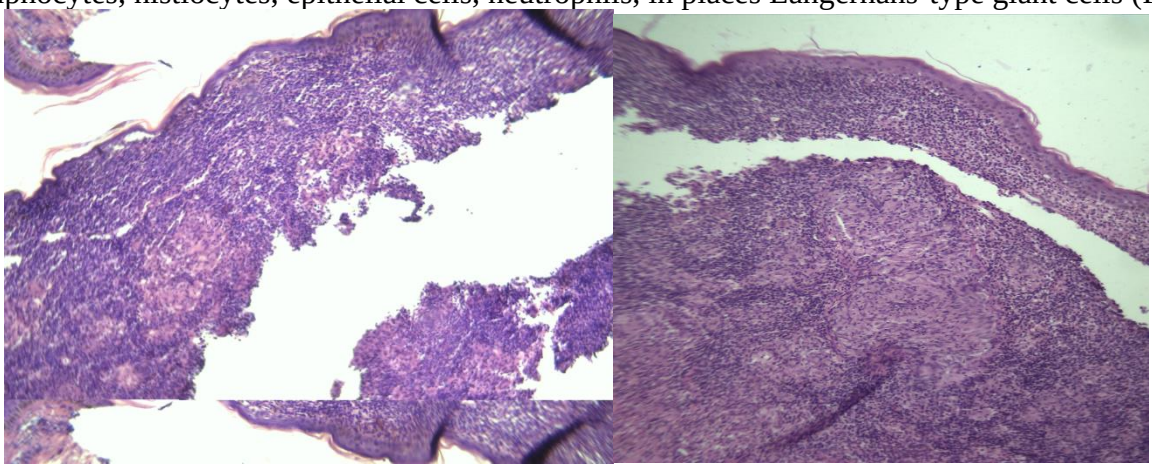

Fig 2 a Histologic picture of the lesion

The results of a biopsy of the ulcers: in the dermis there is dense lymphohistiocytic infiltration with multiple giant multinucleated cells. Epithelioid granulomas are noted in places (Fig. 2b).

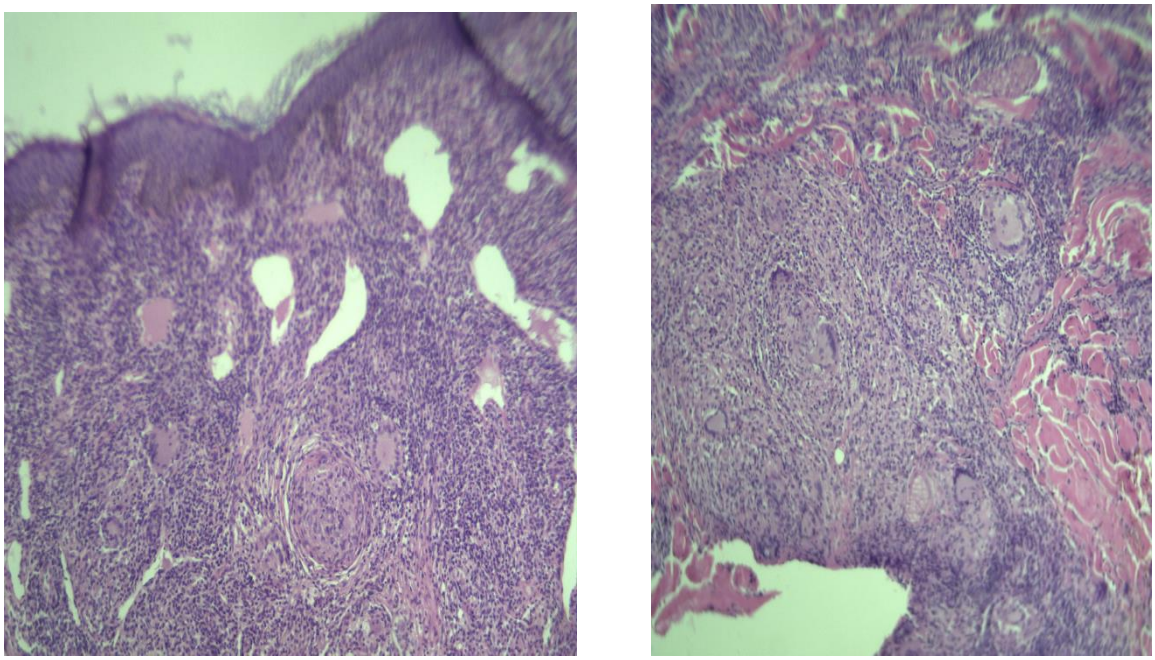

Fig 2 b Histologic picture of the ulcer

Results from laboratory test, including a full blood count, urinalysis, kidney and liver function tests, were normal except for slightly elevated CRP; HIV test negative; characteristic amastigote - not found; A chest X-ray revealed focal shadows in the upper parts of the lungs are against fibrosis, chronic bronchitis, bronchiectasis, diffuse pneumosclerosis.

The patient was started treatment with solution of dexamethasone (8 mg once daily), Stronger Neo-Minophagen C (20 ml once daily), Erbisol solution (2 ml once daily), Spiramycin (3 million twice daily). Locally: Levomycol ointment (chloramphenicol & methyluracilum) in the wound area, Dermovate Ointment on the foci of infiltration. During treatment, there was a visible positive trend (Fig. 3a, b)

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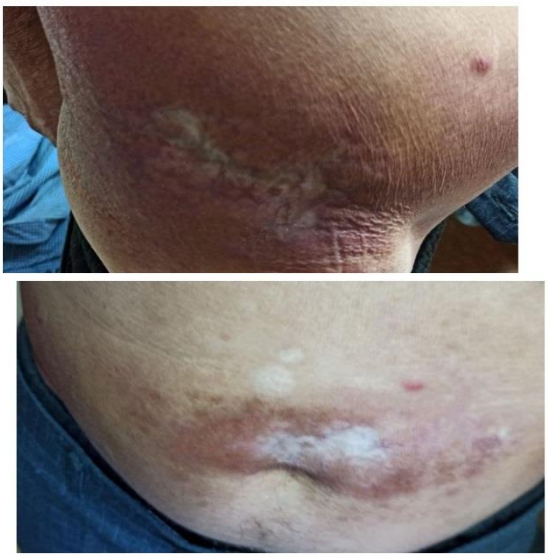

Fig. 3 a At the site of the absorbed plaques, athophy and hypopigmetation are determined

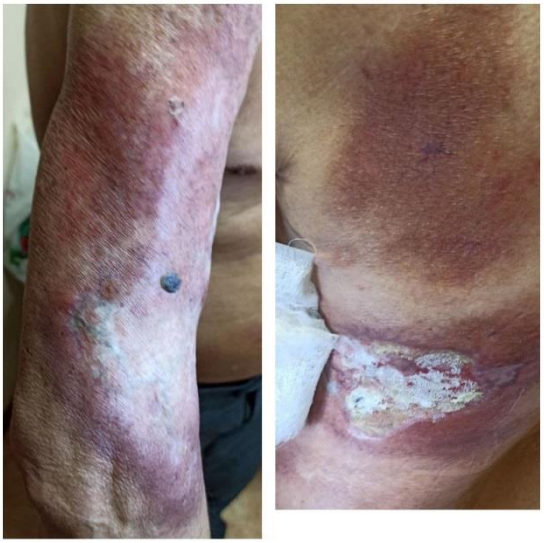

Fig 3 b The surface of the plaques is clear and ulcer in a stage of scarring

Differential diagnosis:

Sarcoidosis vs. tuberculosis

In case of sarcoidosis, small tubercles, an elastic consistency, the absence of specific lesions on the X-ray of the lungs, as well as a patient in the summer of 2018, and also in June 2020, were consulted by a TB doctor, were a diagnosis of skin tuberculosis is excluded.

Sarcoidosis vs. mycosis fungoides

Mycosis fungoides is indicated by the patient's age, as well as the presence of ulcerative, infiltrative plaques of bluish tinge in places, however, the patient has a weakly positive symptom of "apple jelly", as well as the absence of itching, which is one of the main symptoms of T-cell lymphomas. The patient experienced a rather rapid regression of rashes during therapy, which is not typical for mycosis fungoides. And most importantly, histological examination does not show specific changes corresponding to fungal mycosis.

Sarcoidosis vs. leishmaniasis

Lupoid leishmaniasis of the skin attracted much attention. Since, when clarifying the anamnesis, the patient could visit the epidemiological center (periodically went fishing in the lower reaches of the Syr Darya river). Moreover, from the anamnesis, the disease began with one element. Also, relying on published data, lupoid cutaneous leishmaniasis has a protracted course and, when examined, the contents of ulcers rarely find Amastigote. Also, the patient's clinical picture itself is very reminiscent of lupoid cutaneous leishmaniasis.

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The presence of small tubercles on the infiltrative foci, however, the tubercles are randomly located, there is no grouping. With lupoid leishmaniasis of the skin with dioscopy, the phenomenon of apple jelly is clearly traced, as in our case, where the phenomenon of apple jelly is weakly positive. According to published data, Dobrzhanskaya R.S. (1964, 1979) distinguishes the following varieties of lupoid cutaneous leishmaniasis: nodular, diffuse-infiltrative, ulcerative-deforming, erythematous, tumorous and mixed. The most frequent localization of the process is a face, up to 80% [1,2]. However, no lesions were found on the face of our patient. The histological picture with lupoid cutaneous leishmaniasis does not always help with the diagnosis. Histological signs of a specific inflammatory process. However, in rare cases, you can find the pathogen [1]. In our case, all the signs of a specific inflammatory process are present, but the pathogen was not found.

And most importantly, the patient received specific therapy for lupoid cutaneous leishmaniasis in 2018 (a year and a half ago). However, the effect after the therapy was not observed, the process gradually began to progress. After the appointment of corticosteroid therapy in February 2020, a good regression of rashes began to be observed a week after the start of treatment.

Atypical cutaneous sarcoidosis

The final diagnosis was made only during treatment. Given the patient's history, the variability of rashes. Everything indicated a sarcoidosis of the skin, an atypical form. According to literature data, there are many varieties of atypically occurring sarcoidosis of the skin. If we dwell on the ulcerative form of sarcoidosis of the skin, then ulcers form within the primary small or coarse site, marked soreness is noted. With almost all forms of sarcoidosis of the skin, dioscopy reveals small yellowish-brown spots (specks of dust), similar to the phenomenon of apple jelly with lupus [1].Histological examination reveals nonspecific inflammatory infiltrate and typical epithelial granulomas in the deep layers of the dermis [1-4].

In our case, the cyanotic infiltrative foci of a dense consistency are noted on the body, many of which have tight-fitting crusts, in many places the foci are ulcerated, sharp pain, lack of itching. On histologic examination there is granulomas from epithelioid cells and multinucleated giant cells. And most importantly, we noted a good regression of rashes against the background of corticosteroid therapy. However, in this case, we did not observe a typical course of the process in the patient. An atypical form of sarcoidosis, ulcerative type, was observed here.

Conclusion:

Considering the anamnestic data: A 58-year-old man, a long course of the skin process (5 years), lack of effect after specific treatment of leishmaniasis; clinical course of the process: cyanotic infiltrative foci of a denser consistency, in many places ulcerating, the presence of sharp pain, taking into account the results of a biopsy, as well as corticosteroid therapy and the presence of effect during and after treatment, the patient can be diagnosed with the final diagnosis: «Cutaneous sarcoidosis ulcerative form atypical course.»

Regression of rashes, reduction in size and epithelialization of ulcers occurred due to systemic corticosteroids therapy (dexamethasone, prednisone). The patient was prescribed a gradual decrease in corticosteroid according to the scheme. In the month of March after the patient went for a second appointment, a regression of rashes of more than 60% was observed. However, the patient requires further examination in a therapeutic clinic to exclude manifestations of sarcoidosis of the internal organs. The patient was recommended to take prednisone with a gradual decrease within 6 months.

Sarcoidosis of the skin is not yet a fully understood disease, which can mask as other skin pathologies and thereby mislead specialists. In the presented clinical case, the histopathological study was of great importance. The need for a biopsy followed by histopathological examination is shown in cases where it is important to conduct differential diagnostics for the correct diagnosis in complex clinical cases, which, in our opinion, is presented above.

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References

1. Kalamkaryan AA. et al. Rare atypical dermatoses. Clin dermatol. Yerevan: Hayastan; 1989. p.456–460. (In Russ).
2. Berenbejn BA, Studnicin AA. Differential diagnostics of skin disorders. 2. M: Medicina 1989 p.347–351. (In Russ).
3. Lowell A. Goldsmith et al. Fitzpatrick's Dermatology in General Medicine: in 3 volumes :per. from English.; Ls. ed. Potekayev NN. M.: publishing house of Panfilov; BINOM. 2016; 2:2060–71. (In Russ).
4. Tchernev G, Patterson JW, Nenoff P, Horn LC. Sarcoidosis of the Skin–A Dermatological Puzzle: Important Differential Diagnostic Aspects and Guidelines for Clinical and Histopathological Recognition. J Eur Acad Dermatol Venereol. 2010;24(2):125-37. doi: 10.1111/j.1468-3083.2009.03396.x.
5. Jean L Bolognia et al. Dermatology. 3rd ed. 2012, Elsevier. p.1557–63.
6. Mañá J,Marcoval J. Skin manifestations of sarcoidosis. Presse Medicale (Paris, France : 1983), 09 May 2012, 41(6 Pt 2):e355-74. DOI: 10.1016/j.lpm.2012.02.046. PMID: 22579238
7. Powell E, Rosen T. Ulcerative Sarcoidosis: A Prototypical Presentation and Review. Cutis. 2017;100(5):312-316. PMID: 29232421
8. Nefedyeva JV. et al. Case Report of Sarcoidosis of the Skin. Difficult patient. 2018; 5(16):38-40. (In Russ).
9. Yoshiro Ichiki, Yasuo Kitajima. Ulcerative Sarcoidosis: Case Report and Review of the Japanese Literature. Acta Derm Venereol. 2008;88(5):526-8. doi: 10.2340/00015555-0525.
10. Sycheva NL, Faustov LA, Osmolovskaya PS. Problems of clinical diagnosis of atypical skin sarcoidosisn. Klinicheskaya Dermatologia I Venerologia . 2016, 15(2), p19-24. 10.17116/klinderma201615219-24. (In Russ).