A CASE REPORT OF SARCOIDOSIS CUTIS LICHENOIDES

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ABSTRACT:

Background: Sarcoidosis is a chronic multisystem granulomatous disease of unknown etiology, characterised by the formation of non-caseating granulomas in the involved organs, such as the skin, lung, lymph nodes, eyes, joints, brain, kidneys and heart. Cutaneous involvement is about 25% with different clinical presentations, including papules, nodules, plaques, and infiltrated scars. The lichenoid pattern is one of the rarest types of cutaneous sarcoidosis.

Purpose: To present 50-year-old woman diagnosed with Sarcoidosis cutis lichenoides.

Material and methods: We present a 50-year-old woman in good general condition, with skin rash localised on upper limbs. The disease started four months ago. From physical examination, pathological cutaneous lesions affected right and left forearms and cubital folds, presented by papules of various sizes, pink in colour, smooth and shiny. Complete blood count, biochemistry, urine analysis and microbiology were in normal ranges except elevated levels of alpha-1 globulin and gamma-globulin. X-rays examination and abdominal ultrasound were normal too. Mantoux test was negative. Histopathological examination demonstrated granulomas located in the papillary dermis and composed of epithelioid and giant cells, a few lymphocytes, without necrosis. Consultations with pulmonologist and ophthalmologist showed no abnormalities.

Results: The diagnosis Sarcoidosis cutis lichenoides was based on the medical history, general status, and morphological characteristics of skin lesions, histopathological examination and consultations. In the absence of organ involvement, systemic therapy with Methylprednisolone and topical treatment with Flumetasone pivalate 0.2mg/Salicylic acid 30mg/g was started. On the 5th day of treatment, we revealed initial resorption of papules. After discharging the patient from the hospital, systemic therapy with Methylprednisolone and topical treatment with Tacrolimus 0.1% ointment was conducted. There were no new lesions 3 months after the end of the therapy with Tacrolimus 0.1% ointment.

Conclusions: We present a rare case report of Sarcoidosis cutis lichenoides.

Keywords: Sarcoidosis cutis lichenoides, lichenoid pattern, tacrolimus.

INTRODUCTION:

Sarcoidosis is a rare systemic multi-organ disease with unknown aetiology. It is characterized by granulomas, without caseous necrosis, in the affected tissue of the lung, lymph nodes, brain, kidneys, heart, eyes and joints. Skin involvement is observed approximately 25% to 35% of cases and can be detected at any stage of the disease, most commonly occurring in the early stages. Because of the variety of lesions described in cutaneous sarcoidosis, diagnosis is a challenge. Exclusion of many other diseases is required as there is no single diagnostic test to prove this granulomatosis [1]. We present a clinical case of cutaneous sarcoidosis in a 50-year-old woman with lichenoid skin lesions. The final diagnosis of Sarcoidosis cutis lichenoides was based on the clinical and histological findings.

CASE REPORT:

A 50-year-old woman was observed in the Department of dermatology and venereology with a history of skin rash on her upper limbs. The skin eruption started 4 months before hospitalisation and number and size of the lesions gradually increased. The patient does not report itching. She is having pain from palpation of the rash. The patient has second degree chronic venous insufficiency, as an underlying disease. Physical examination does not reveal any pathological changes. Pathological skin changes have affected left and right forearms and cubital folds. They were presented by smooth and shiny, pink coloured papular lesions with a diameter of 1-3 mm (Fig. 1, 2). The size and number of the papules increased with diagnostic process. There were no pathological changes in the mucous membranes and skin appendages. Peripheral lymph nodes were not palpated. There was no evidence of synovitis and arthritis from the musculoskeletal system examination. Increased values of the following laboratory parameters were found: erythrocyte sedimentation rate (32/70 mm), C-reactive protein (6.98 mg/l), Alpha 1 globulin (8%), Gamma-globulin (21%) as well as the ratio of T-helper / T-suppressors. From total blood count Stm-granulocytes was elevated (77.4%) and lymphocytes were lowered (15%). There were no pathological findings from lung and heart X-ray and abdominal ultrasound. Mantoux test was negative. Histopathological examination of the skin revealed atrophy of the epidermis and multiple, superficially and deeply located, well-demarcated, non-caseating granulomas composed of epithelioid histiocytes,
eosinophils, plasma cells, lymphocytes and Langhans giant cells, in the dermis. (Fig. 3, 4). There were no pathological changes from the consultations with pulmologist and ophtalmologist. The dermatological, laboratory, radiographic and histological findings listed above have been consistent with the diagnosis of the skin sarcoidosis. In the absence of organ involvement, the patient initiated systemic therapy with Methylprednisolone at a dose of 40mg /daily for seven days and topical treatment with Flumetasone pivalate 0.2mg/ Salicylic acid 30mg/g. On the fifth day of treatment, we recorded initial resorption of the papules. After the discharge, the patient continued the following treatment: systemically with Methylprednisolone tablets (for 14 days) and topically with Tacrolimus 0.1% ointment. There were no new lesions after three months of treatment with Tacrolimus 0.1% and the old ones were completely resolved (Figure 5).

**Fig. 1 and Fig. 2.** Right and left forearms – lichenoid papules in cubital folds of right and left forearms.

**Fig. 3.** Skin sarcoidosis, Hematoxylin and eosin stain (HE) magn. x 25. Atrophy of the epidermis. Inflammatory granulomatous reaction in the dermis with multiple well-demarcated, uniform in size and shape, superficially and deeply located, noncaseating granulomas.

**Fig. 4.** Skin sarcoidosis, Hematoxylin and eosin stain (HE) magn. x 40. An epithelioid granuloma composed of epithelioid histiocytes, plasma cells, few lymphocytes and Langhans giant cells.
Fig. 5. Right forearm – completely resolving of the papules three months after treatment with Tacrolimus ung. 0.1%

DISCUSSION:

Sarcoidosis is a chronic granulomatous multisystem disease with unknown aetiology. The disease was described for the first time from Besnier in 1889. The author reports a patient in whom the relationship between red lesions on the face and nose and swelling of the fingers is examined [2].

This diagnosis occurs with a slightly higher incidence in women than in men. Usually, patients are between 20 and 40 years old [3]. Skin involvement was observed in 25% of cases with different clinical manifestations [4]. Skin rashes in sarcoidosis can be classified into two groups: specific and non-specific [2]. The lichenoid form of Sarcoidosis is in the group of specific lesions. This extremely rare skin manifestation of the disease occurs in 1%-2% of cases of skin sarcoidosis [5]. Clinically, the lichenoid type is presented by multiple 1-3 mm, erythematous or violet maculopapular lesions, covered with finesquams. Most common rashes affected extensor areas of the face, trunk and limbs and the lesions could be single or grouped, as in the case here presented. The dermoscopy usually revealed circular or oval yellowish-brown lesions with the absence of Wickham’s striae, even if this pattern is not specific for sarcoidosis. These homogeneous patches indicate for a granulomatous skin disease [6]. Lichenoid lesions have rarely been reported in young children, often in combination with ocular and joint complications, but respiratory system involvement is usually absent [7, 9].

The histological examination of Sarcoidosis reveals granulomas without caseous necrosis, with epithelioid histiocytes, a different number of Langhans cells and mature macrophages in the skin, lungs and lymph nodes [8, 10]. They are surrounded by a small number of lymphocytic infiltrates composed mainly of CD4 + T-lymphocytes and several CD8 + lymphocytes [11]. It is essential to use special stains in histological preparations and various cultures to exclude bacterial and fungal infections. The reason is that sarcoid granulomas have no unique histological characteristics to differentiate them from other non-caseating granulomatous diseases [12].

Various therapeutic approaches are described in the treatment of cutaneous sarcoidosis including local, intraleisional and systemic corticosteroids, anti-malaric treatment, Methotrexate, Tetracycline, Thalidomide, Allopurinol, Isotretinoin and Infliximab. In the last decade, interest in the use of topical calcineurin inhibitors in the treatment of cutaneous sarcoidosis has increased [7, 13, 14]. Tacrolimus, 0.1% ointment, is an immunosuppressive agent, a macrolide isolated from Streptomyces tsukubaensis. Tacrolimus has a modulating effect on the T cell-mediated response and has the ability to suppress both hapten-induced production of Th1 cytokines and the production of TNF-alpha [9, 15]. Several cases of cutaneous sarcoidosis treated with Tacrolimus 0.1% ointment have been reported in the literature [17, 18, 19]. Only one of them has reported a lichenoid pattern [16].

CONCLUSION:

In conclusion, we presented a rare clinical case of a 50-year-old woman with Sarcoidosis cutis lichenoides. There was no other organ involved in the patient. Treatment with Tacrolimus 0.1% ointment gave excellent results. The patient is under observation now.

REFERENCES:


Please cite this article as: Tsvetanova D, Yordanova IA, Pavlova V, Betova T, Gospodinov DK. A case report of Sarcoidosis cutis lichenoides. J of IMAB. 2017 Apr-Jun;23(2):1560-1563. DOI: https://doi.org/10.5272/jimab.2017232.1560

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