Hypertrophic Lichen Planus – a Case Report

Mirjana PARAVINA¹*

¹Medical Faculty, University of Niš, Clinic of Skin and Venereal Diseases, Clinical Center of Niš, Serbia

*Correspondence: Mirjana Paravina, E-mail: mirjanaparavina@gmail.com

Abstract

Lichen planus is an immune, inflammatory reaction with characteristic clinical and histological lesions. It is a benign disorder, often chronic or recurrent, characterized by flat-topped, pink to purple, shiny pruritic polygonal papules on the skin, or milky white reticular papules on the visible mucous membranes. Hypertrophic lichen planus is a chronic form of lichen planus with marked epidermal hyperplasia and intense pruritus. It is characterized by symmetrical hypertrophic plaques, usually located on the pretibial or perimalleolar regions. Lesions are often resistant to treatment.

This paper presents a patient with a giant form of verrucous lichen planus on the lower extremities, with a chronic course and resistance to various forms of therapy (keratolytics, local and intralesional corticosteroids, radiotherapy, systemic antibiotics, cryotherapy). Significant improvement was seen after 8-month treatment with etretinate (initial dose of 75 mg per day, with progressive reduction to 10 mg per day). Etretinate therapy resulted in a significant regression of the disease.

Key words

Lichen Planus + physiopathology + therapy; Etretinate; Treatment Outcome

Lichen planus is a rare, immune, inflammatory tissue reaction with characteristic clinical and histological lesions (1). Several other diseases, including graft-versus-host disease, may induce lichenoid tissue reactions. Lichen planus is the prototype of all lichenoid eruptions. Its prevalence in the general population ranges from 0.5% (6) to 1% (7), even to 4% (8). The disease is benign, often chronic or recurrent, and it is characterized by flat-topped, pink to purple, shiny, pruritic polygonal papules on the skin or milky white reticular papules on the visible mucous membranes (2, 3). Some describe lichen planus with the six “Ps”: planar, purple, polygonal, pruritic, papules and plaques (9).

Hypertrophic lichen planus is a less common form of lichen planus. It accounts for 4.7% of lichen planus cases, and 2.2% of all lichenoid tissue reactions (10). It is a chronic, hypertrophic form of the disease with severe epidermal hyperplasia and itching (7). It is characterized by symmetrical hypertrophic plaques, usually located on the pretibial or perimalleolar regions (4, 5). The disease has a chronic course, and it is often resistant to treatment. Upon remission, scarring, hypo- or hyperpigmentation may follow (7, 11).

This paper presents a patient with a giant form of verrucous lichen planus on the lower extremities, with favorable results after treatment with systemic etretinate.

Case report

We present a 47-year-old housewife in good general health, except for surgical treatment of uterine myomatosis and hypertension. At the beginning, the patient presented with itchy warty growths on the sides of the heels, rapidly growing and spreading to...
other parts of the foot. They appeared on the front of the right lower leg 2-3 years later, at the site of a previous skin injury. There were no relatives with a similar disorder.

The patient was repeatedly treated at the Clinic for Skin and Venereal Diseases in Niš and once at the Military Medical Academy in Belgrade. She was treated with keratolytics, corticosteroids under occlusion and intralesional, cryotherapy with liquid nitrogen, radiotherapy (a total dose of 20 Gy, 10 sessions, two opposing fields), systemic antibiotics due to a secondary infection, sedatives, etc.), but without satisfactory long-term results. Therefore, systemic etretinate therapy was initiated.

Dermatological status before etretinate therapy
Tumor-like infiltrations, individual or confluent, were found on the sides and back of the heel, forming spur-like growths around the heels (Figure 1). Their surface was keratotic, verrucous, dark gray, and they were interspersed with rhagades producing hemorrhagic-purulent discharge under pressure. On the front right leg there was a tumor, 5 cm in diameter, with similar characteristics (Figure 2). The lesions were extremely pruritic, often painful, making everyday activities such as wearing shoes and walking difficult. There were no other changes on the skin and visible mucous membranes.

Laboratory tests
All required laboratory test results were within reference limits: erythrocyte sedimentation rate, complete blood count, hemoglobin, leukocyte count, urea, creatinine, liver scan, transaminases, protein electrophoresis, acid and alkaline phosphatase, lactate dehydrogenase, cholesterol, triglycerides, blood glucose, serum electrolytes; serological tests for adenoviruses, cytomegalovirus, herpes simplex virus, and Treponema pallidum. Staphylococcus aureus was isolated from the lesions, pointing to a secondary infection.

Histopathological analysis
Light microscopy of the affected skin biopsy samples showed: marked hyperkeratosis; sporadic involvement in the granular layer; uneven acanthosis, partly in the form of elongated epidermal ridges; moderate dermal infiltrations consisting mainly of lymphocytes and some histiocytes which mostly penetrated into the lower layers of epidermis, making the dermal-
Figure 2. Skin lesions on the internal parts of the feet and on the right lower leg before etretinate therapy

Figure 3. Histopathological finding (moth-eaten appearance)
epidermal border unclear (moth-eaten appearance) (Figure 3).

Direct immunofluorescence analysis
Direct immunofluorescence showed findings typical for lichen planus: fibrinogen deposits with rough jagged edges along the epidermal-dermal junction and in the blood vessel walls in the papillary dermis; multiple clustered colloid bodies (IgM) beneath the dermal-epidermal border; lines of fibrinogen deposits in the border area between the dermis and epidermis.

Treatment
Etretinate was initiated at a dose of 75 mg per day (3 x 25 mg, ie. 1 mg/kg/day, with a gradual decrease to 10 mg per day). Topical keratolytics were used with boric lotion. The treatment lasted for 8 months. The patient reported the following side effects at the beginning of treatment: dry mouth, increased desquamation of the palms and soles, transient elevation of serum triglycerides, which was normalized after dose reduction. The therapeutic effects on skin lesions were remarkable (Figures 4 and 5). The tumefaction decreased, the heels regained nearly normal size, the patient could wear shoes, there were no secondary infections and relief of itching was significant.

During the further course of the disease, the patient suffered from a mild recurrence in the following year, so etretinate was initiated again at a dose of 0.5 mg/kg bw/day with a gradual reduction to a maintenance dose of 10 mg every other day. In the last two years, the patient's condition remained unchanged, and her quality of life has improved. Currently, the skin lesions are located at the same sites as during the worst stage of the disease, but the infiltrates are less prominent, clearly demarcated, uneven keratotic lesions, with scattered atrophic areas circumscribed by hyperpigmentation.

Discussion
Lichen planus usually occurs between 30 and 60 years of age (1), more frequently in women. It is a chronic mucocutaneous T-cell-mediated disease, the cause of which remains unknown (12). According to Boyd and Neldner (13), there are two main types of lichen planus: "classical" idiopathic type, and lichenoid reaction.
Viruses, bacteria, hormones, metal ions, drugs and physical factors are considered to be potential triggers (18, 19). The most accepted theory is that it is a cellular autoimmune response to a viral infection (20, 21). Cellular components of the immune system induce epidermal reactions, injury to basal keratinocytes, and secondary inflammatory reactions (22, 23, 24). In lichen planus lesions, CD8+ T cells infiltrate the epidermis while T cells, both CD4+ and CD8+, accumulate in the dermis. It has been suggested that CD8+ cytotoxic T cells recognize an antigen (currently unknown) associated with the major histocompatibility complex (MHC) class I on lesional keratinocytes and lyse them (24).

Malignancy developing in lesions of lichen planus is a rare phenomenon, except in oral lichen planus (chronic inflammation and accelerated cellular turnover in erosive lesions) (25), where the risk is estimated to range from 1.1 to 3.5% (26, 27, 28), which is the reason
why the WHO defines it as a pre-cancerous conditions (29). Among other localizations and clinical forms being complications of hypertrophic lichen planus of the lower extremities, the most common are neoplastic transformations and squamous cell carcinoma with a risk of metastasis (30), (25, 31, 32, 33, 34).

The diagnosis is made through clinical and histopathological findings. Differential diagnosis of various forms of lichen includes several conditions (lichenoid eruptions induced by drugs or color developer, eczematous eruptions with lichenification from scratching, lichen amyloidosis), whereas in hypertrophic lichen planus, chronic lichen simplex must be excluded (19).

The choice of treatment mainly depends on the clinical type of lichen planus and its localization. According to Oliver et al (1), general principles of treatment include rest, dressings, topical steroids (mild or potent), retinoids, treatment of oral lesions and treatment of secondary or primary infections. Topical calcineurin inhibitors, such as tacrolimus (35, 36), pimecrolimus (37) and cyclosporine (38) are second-line therapies. H

References

Conclusion
This case report describes a patient with a rare form of verrucous lichen planus on the lower extremities. Although lesions were resistant to treatment, systemic etretinate therapy resulted in a significant regression of the disease, leading to better quality of life.
Hipertrofični lihen planus – prikaz slučaja

Sažetak

Uvod. Lichen planus je inflamatorna imunska reakcija sa karakterističnim kliničkim i histološkim lezijama. Bolest je benignog, često hroničnog ili recidivantnog toka, a karakterišu je zaravnjene ružičaste do ljubičaste sjajne vrugoci bez višenaplatnosti na koži ili mlečnobele karakterišu je zaravnjene ružičaste do ljubičaste sjajne pruritične poligonalne papule na vidljivoj sluzokoži.\n\nLichen planus hypertrophicus je hronična hipertrofična forma bolesti, sa najčešćim smučenim epitelijskim, kožnim i perimalne larno. Lijekovi se često rezistentne na terapiju. Prikaz slučaja. Prikazuje se 47 godina stara domaćica, dobrog opšteg stanja; osim operativnog lečenja miomatoznog uterusa i hemoroida u drugoj bolesti bilo oboljelih i valjano života i mlečnobele retikularne papule na vidljivoj sluzokoži.

Hipertrofični lihen planus

Lichen planus hypertrophicus je hronična hipertrofična forma bolesti, sa najčešćim smučenim epitelijskim, kožnim i perimalne larno. Lijekovi se često rezistentne na terapiju. Prikaz slučaja. Prikazuje se 47 godina stara domaćica, dobrog opšteg stanja; osim operativnog lečenja miomatoznog uterusa i hemoroida u drugoj bolesti bilo oboljelih i valjano života i mlečnobele retikularne papule na vidljivoj sluzokoži.

Dozvoljeno je da se iz navedenog dokumenta izrade tradukcija, pozorišta, gledaljske radnje, ili drugi oblici izrade i distribucije, uz određenih uvjeta.
tečnim azotom, zračna terapija X-zracima – ukupna primljena doza od 20 Gy data u 10 seansi na dva suprotna polja, sistemski antibiotici prema antibiogramu zbog sekundarne infekcije, sedativi, itd.) nije dala zadovoljavajuće i trajnije rezultate. Zbog toga je odlučeno da se primeni etretinat na obliku kapsula. U momentu pregleda, na bočnim i zadnjim stranama peta bili su prisutni infiltrati tumoroznog izgleda, pojedinačni ili sliveni, koji su davali izgled mamuza oko peta. Površina im je bila keratočvrsna, vežak uključeno u glavnom delu, sivkasto mrko prebojena, ispresecana ragadama, iz kojih se na pritisak cedio hemoragično purulentan sadržaj. Na prednjoj strani desne potkolenice bio je prisutan tumefakt oko 5 cm u prečniku, sličnih osobina. Promene su bile jako pruritične, često bolne, onemogućavale su nošenje obuće i otežavale hod. Drugih promena na koži su bile jako pruritične, često bolne, onemogućavale su nositi cipele i otežavale hod. Drugih promena na koži su bile jako pruritične, često bolne, onemogućavale su nositi obuće i otežavale hod. Drugih promena na koži su bile jako pruritične, često bolne, onemogućavale su nositi obuće i otežavale hod. Drugih promena na koži su bile jako pruritične, često bolne, onemogućavale su nositi obuće i otežavale hod. Drugih promena na koži su bile jako pruritične, često bolne, onemogućavale su nositi obuće i otežavale hod. Drugih promena na koži su bile jako pruritične, često bolne, onemogućavale su nositi obuće i otežavale hod. Drugih promena na koži su bile jako pruritične, često bolne, onemogućavale su nositi obuće i otežavale hod. Drugih promena na koži su bile jako pruritične, često bolne, onemogućavale su nositi obuće i otežavale hod. Drugih promena na koži su bile jako pruritične, često bolne, onemogućavale su nositi obuće i otežavale hod. Drugih promena na koži su bile jako pruritične, često bolne, onemogućavale su nositi obuće i otežavale hod. Drugih promena na koži su bile jako pruritične, često bolne, onemogućavale su nositi obuće i otežavale hod. Drugih promena na koži su bile jako pruritične, često bolne, onemogućavale su nositi obuće i otežavale hod. Drugih promena na koži su bile jako pruritične, često bolne, onemogućavale su nositi obuće i otežavale hod. Drugih promena na koži su bile jako pruritične, često bolne, onemogućavale su nositi obuće i otežavale hod. Drugih promena na koži su bile jako pruritične, često bolne, onemogućavale su nositi obuć

Ključne reči

Lichen planus + fiziopatologija + terapija; Etretinat; Ishod lečenja