



A Clinical Case of Pityriasis Lichenoides Chronica Presenting with Palpable Purpura after Streptococcal Infection

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Abstract

Pityriasis lichenoides is a rare inflammatory skin condition presenting with diffuse red-brown papules with evolution polymorphism and mica-like crust on older skin lesions. We present a 60-year-old female patient with pityriasis lichenoides chronica that manifested ten days after streptococcal pharyngitis. Initially, palpable purpura appeared on the lower extremities and later, erythematous-squamous papules and plaques appeared at the site of the palpable purpura and on the upper limbs and trunk. The patient had no history of hematological malignancy, viral hepatitis, kidney involvement, systemic rheumatic disease, or ANCA-associated vasculitis. After administration of methylprednisolone 20 mg for one month and an antimalarial agent (hydroxychloroquine 200 mg, 1 tablet bid) for three months, the skin lesions subsided without recurrence.

Keywords

palpable purpura, pityriasis lichenoides chronica, streptococcal pharyngitis

INTRODUCTION

Pityriasis lichenoides (PL) is a group of inflammatory skin disorders manifesting with erythematous macular lesions on the skin of the trunk and extremities and subsequent evolution to papules with hemorrhagic necrosis and the appearance of skin ulcerations^[1,2], causing intense itching and burning sensations. It encompasses several conditions, including pityriasis lichenoides et varioliformis acuta (PLEVA) with its subtype febrile ulceronecrotic form (Mucha-Habermann disease), and pityriasis lichenoides chronica (PLC).^[1] PL affects all age groups and races, and

can develop in both males and females, with slightly higher prevalence in males of young age.^[1,2] The prevalence of PL is considered up to 0.5%.^[2]

The etiology of PL is not well understood. It has been associated with many provocative factors and agent, including infections (herpes viruses – Epstein-Barr and varicella-zoster, herpes simplex 2, HIV, streptococci, toxoplasma, etc.), drugs, vaccines and xenobiotics, malignancies, especially lymphoma.^[1-6] The pathogenesis of PL is unclear, but immune-complex mediated hypersensitivity reaction has been suspected, along with cell-mediated hypersensitivity response to exogenous or endogenous antigen(s)^[7] with

formation of vascular wall inflammation, extravasation of red blood cells, fibrin and plasma component, release of lysosomal enzymes and reactive oxygen species from the clustered neutrophils, and further damage of the vascular wall. The described vascular wall changes in combination with the altered blood flow may lead to the formation of palpable purpura.^[8]

In 2023, we observed a patient with PLC developing after streptococcal pharyngitis, manifesting with palpable purpura of the lower extremities. Written informed consent was obtained from the patient prior to any diagnostic or therapeutic procedure.

CASE PRESENTATION

A 60-year-old female patient was referred to the Clinic of Rheumatology of Sofamed University Hospital in January 2023 for joint pain and palpable purpura on lower extremities. She reported having streptococcal pharyngitis in November 2022, with high fever, cervical lymphadenopathy, sore throat, and elevated antistreptolysin titer (537.40, normal <200 Todd units). She saw an ENT specialist, who prescribed amoxicillin and clavulonate (875/125 mg bid) for 10 days, but she took the antibacterial treatment for four days and stopped it without further consultations and despite the good tolerability. Ten days after she stopped the antibiotic, she had one episode of diarrhea followed by pain in the knee and ankle joints with palpable purpura in the thighs and lower legs (**Fig. 1**).

On December 23, 2022, she consulted an allergologist, who diagnosed skin vasculitis and prescribed treatment with methylprednisolone 16 mg/24 h and gradually decreased the dose. By the time of hospitalization, she was taking 4 mg/24 h. For six days, she also received 0.4 ml of fraxiparin subcutaneously. The laboratory tests in outpatient settings (January 4, 2023) revealed increased leukocyte count (12.8 G/l) with high lymphocyte count (5.6 G/l), normal ESR (16 mm/h) and C-reactive protein (2.5 mg/l), normal urinalysis, and increased rheumatoid factor (21.6 IU/ml, normal <14). The patient had a past history of arterial hypertension and aller-



Figure 1. Palpable purpura on the lower extremities.

gy to tetracycline. She reported having taken penicillin antibiotics in the past with good tolerability of the treatment. The patient had been on corticosteroid treatment from December 23, 2023, until the admission to the Rheumatology Clinic at the end of January 2024.

The physical exam revealed multiple rounded erythematous-squamous plaques on the lower extremities and dorsal surface of the palms, single erythematous-squamous papules and plaques on the trunk and upper limbs, yellowish squamous plaques on the neck and the occipital part of the capillitium (**Fig. 2**). The patient had spontaneous and provoked pain in ankles and knees, and mild hydrops in left knee. No



Figure 2. Rounded erythematous-squamous papules and plaques.

other pathological findings were present.

The clinical-laboratory investigations revealed normal ESR, complete blood count, fibrinogen, and biochemical investigations; positive ASO: 331.8 Todd units (normal <200), and positive rheumatoid factor: 38.4 IU/ml (normal <20); cryoglobulins, ANA, ANCA, anti-MPO, and anti-PR3 were negative; serum immunoglobulins (IgG, IgA and IgM) and C3 and C4 complement fractions were within the normal limits; HIV, VDRL, HBsAg and anti-HCV were negative; normal urinalysis, and negative urine culture. Chest X-ray examination was normal.

On January 30, 2023, while on 4 mg methylprednisolone, the patient underwent punch skin biopsy (3 mm) from erythematous-squamous papule on the left shoulder. The pathohistological investigation revealed irregularly expressed perivascular lymphocytic inflammatory infiltrate, containing here and there single macrophages and segmented nuclear leukocytes in the upper dermis. The infiltrate was more intense in part of the edematous dermal papillae, accompanied by small erythrocyte extravasates, adjacent to basal vacuolar changes, discrete spongiosis of the epidermis with cap-shaped parakeratosis above these changes. The histological findings were compatible with PLC. The patient was started on intravenous methylprednisolone 20 mg a day for 5 days and the skin lesions gradually faded and subsided with disappearance of erythematous-squamous papules and plaques (Fig. 3), and joint pain gradually subsided.



Figure 3. Fading of erythematous-squamous papules and plaques after five days of treatment with 20 mg intravenous methylprednisolone.

After 5 days of intravenous corticosteroid treatment and the described beneficial clinical evolution of the condition, the patient was discharged and was given oral methylprednisolone 8 mg a day (2 tablets) with gradual dose reduction and withdrawal within one month, and the patient was switched to hydroxychloroquine 200 mg bid for three months without recurrence of symptoms.

DISCUSSION

Pityriasis lichenoides is a rare inflammatory skin condition that can develop after infection, vaccination or other contact with foreign antigens.^[1,2] PL should be differentiated from cutaneous vasculitis, Gianotti-Crost syndrome, lichen planus, secondary syphilis, skin lymphoma and histiocytosis, exantematous drug eruptions, urticaria pigmentosa, viral infections of the skin and all conditions associated with erythematous macular lesions, papules with hemorrhagic necrosis and skin ulcerations.^[1] The course of the disease is usually mild and self-limiting, and therefore, in the majority of cases, no specific treatment is required, besides anti-inflammatory agents and antibiotics, if infection is suspected or present. The retrospective studies on the systemic treatment of PLC have shown the most effective strategy as first line treatment is the administration of corticosteroids in combination with antibacterial agents (especially macrolide antibiotics) and/or phototherapy^[9], and supportive treatment – antihistamines to relieve itching, local and topical treatment of ulcerative changes^[10]. Phototherapy is indicated when anti-inflammatory + antibiotic treatment is ineffective^[9,11], and also as a first-line therapy^[9,12]. Other medications, used for PL treatment, are: methotrexate, cyclophosphamide, tacrolimus, cyclosporin, etanercept, dupilumab, oral retinoids, vitamin D3 analogues, diamino diphenyl sulfone, amitriptyline, pentoxifylline, colchicine, pyrimethamine and trisulfapyrimidine, and topical tar coal preparations, topical capsaicin, topical menthol, hydrocolloid dressings.^[1,2,9,12-17] The course of PL is usually beneficial, although rare cases of fatal outcome have been reported, mainly with the febrile ulceronecrotic form (known as Mucha-Habermann disease).

In our patient, the diagnosis PL was based on the typical clinical manifestations preceded by streptococcal infection, the biopsy findings, and the beneficial effect of corticosteroid treatment. Viral hepatitis, HIV and syphilis were ruled out, and there was no data for skin vasculitis or ANCA-associated vasculitis, no sign of hematological malignancy, serum immunoglobulins and complement fractions were normal. The provoking factor in our patient was streptococcal pharyngitis, proven both clinically and serologically. It is well known that streptococci are a frequent provoking factor for the development of PLC, proven in 79% of the cases, mainly using serological markers.^[18] Therefore, antimicrobial treatment (especially with antistreptococcal agents) is advisable as treatment of first choice, in combination with anti-inflammatory agents. Our patient received a

short course of oral antibiotic (ceased by the patients herself, without consulting physician) and oral corticosteroid treatment was initiated prior to the admission to the Rheumatology Clinic. Corticosteroids were continued during the hospital stay in higher doses and had beneficial effect on skin changes and symptoms. Therefore, phototherapy was not undertaken, and no additional anti-inflammatory medications were given. In order to withdraw safely the corticosteroid treatment, we decided to switch the patient to an antimalarial agent (hydroxychloroquine) for three months that proved to be effective to prevent disease recurrence. The patient is still followed up and has shown no further recurrence of PL.

In conclusion, we present a female patient with PLC, developing after streptococcal infection of the throat and resolving at the background of low/medium dose of corticosteroid treatment followed by antimalarial agent without further relapses.

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Competing Interests

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Author contributions

All authors have equally contributed to the diagnosis and treatment of the patient. All authors have contributed to the development of the manuscript. All authors have approved the manuscript.

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Клинический случай хронического лихеноидного лишая с пальпируемой пурпурой после стрептококковой инфекции

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Резюме

Лихеноидный петириаз — редкое воспалительное заболевание кожи, проявляющееся диффузными красно-коричневыми папулами с эволюционным полиморфизмом и слоистой коркой на старых поражениях кожи. Мы представляем 60-летнюю пациентку с хроническим лихеноидным петириазом, который проявился через десять дней после стрептококкового фарингита. Первоначально на нижних конечностях появилась пальпируемая пурпура, а затем на месте пальпируемой пурпуры, а также на верхних конечностях и туловище появились эритематозно-сквамозные папулы и бляшки. У пациентки не было в анамнезе гематологических злокачественных заболеваний, вирусного гепатита, поражения почек, системного ревматического заболевания или ANCA-ассоциированного васкулита. После приёма метилпреднизолона 20 мг в течение одного месяца и противомаларийного средства (гидроксихлорохин 200 мг, 1 таблетка два раза в день) в течение трёх месяцев поражения кожи исчезли без рецидива.

Ключевые слова

пальпируемая пурпура, хронический лихеноидный петириаз, стрептококковый фарингит