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CLINICAL REPORT

"Occult Cutaneous Lymphangiectasis": An Unusual Case of Cutaneous Lymphangioma

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An unusual case of cutaneous lymphatic abnormality is described and named as "occult cutaneous lymphangiectasis". A 26-year-old man had noticed pigmented maculae in the left inguinofemoral region and waist, which had gradually increased in number for as long as he could remember. The peculiar distribution of the eruptions and their transient saccular dilatation due to infection suggested that they were of lymphatic origin. Lymphangiography showed the presence of dilated lymphatics in the left inguinofemoral area and correspondence of the distribution of dermal backflow with the locations of the pigmented maculae. The histology is consistent with a diagnosis of lymphangioma. We could find no other reports of cases of cutaneous lymphangioma featuring pigmented maculae as the sole manifestation, although whether the pigmentation is an original clinical feature of this type of lymphatic abnormality is still an open question. ***Key words: cutaneous lymphangioma; occult; unusual, reflux; dermal backflow.***

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Lymphangioma is a hamartomatous malformation of lymphatic vessels that shows wide variations in clinical and pathological features, depending on the nature of the lesions and on their complications. Most cases are categorized into three types of lymphangioma (lymphangioma circumscriptum, cavernous lymphangioma and cystic hygroma). We report an unusual case of cutaneous lymphangioma, named as "occult cutaneous lymphangiectasis" because of its unique skin manifestations and difficulty in fitting to the ordinary classification of cutaneous lymphangioma.

CASE REPORT

A 26-year-old man was admitted with fever (38.8°C), general fatigue, diffuse erythema of the lower left trunk, the genital area and the left thigh, and left inguinal pain. The patient had noticed genital skin eruptions for as long as he could remember, and these had gradually increased in number, but he had not had any other skin complaints such as lymphedema of the leg, or fluid discharge from the skin. He had had three similar, but milder episodes of painful swelling in the same site when at primary school, secondary school, and university. During the second episode, at age 14, the patient was hospitalized and treated but was not found to have any hereditary abnormality or mental or physical disorder. On physical examination, the scrotum was found to be swollen and erythematous, with numerous blister-like eruptions, and several papules, measuring 5 mm in diameter, present on the shaft of the penis (Fig. 1). Diffuse

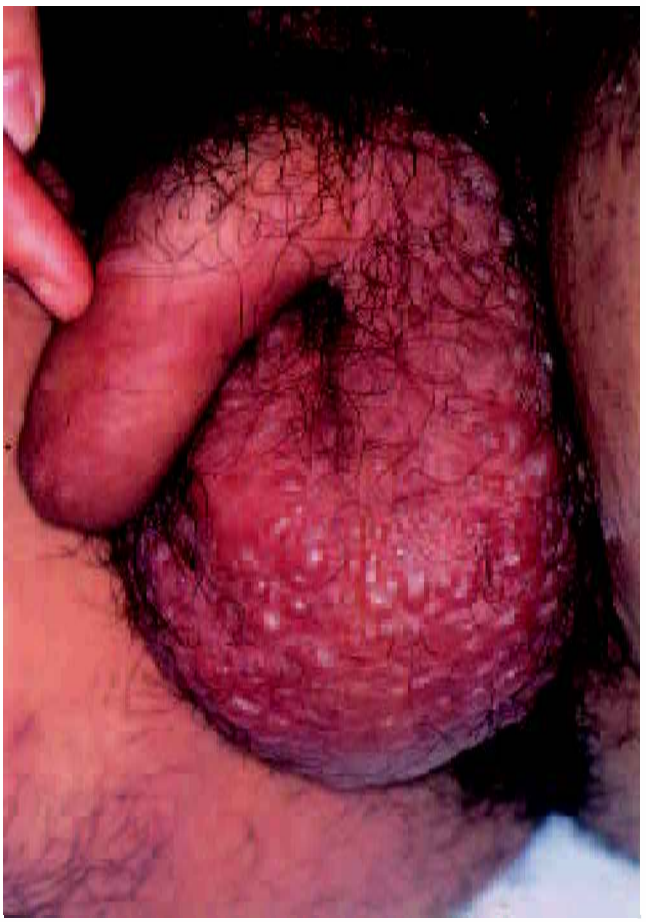

Fig. 1. Clinical appearance of the scrotum and penis at admission. The scrotum is swollen and erythematous, with numerous blister-like upheavals. Small similar papules can also be seen on the shaft of the penis.

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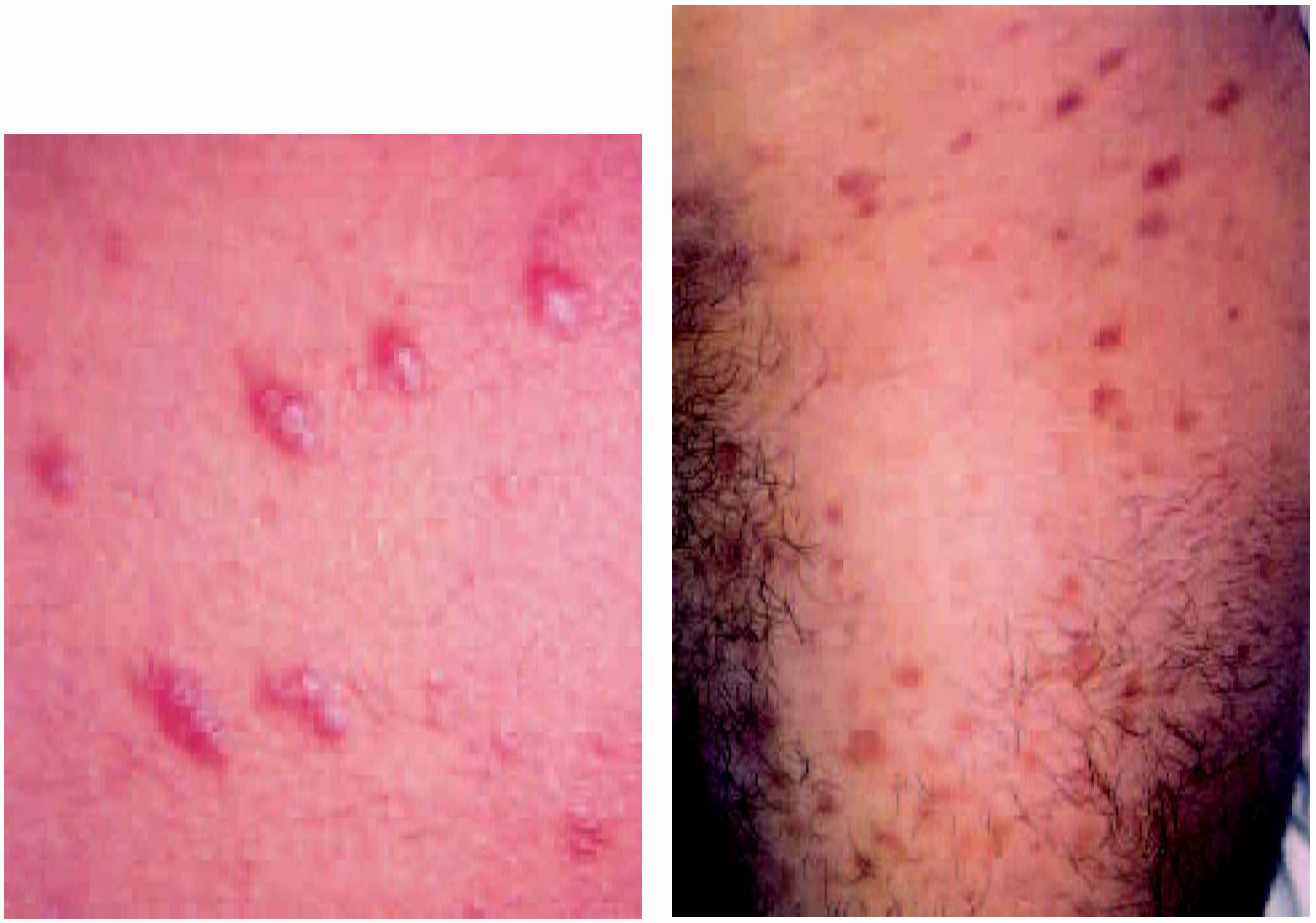
erythema extended over the lower abdomen, waist, genital area, and left thigh, with numerous skin eruptions resembling keloids, ranging from 5 mm to 20 mm in diameter (Fig. 2*A*). A slightly bloody yellow fluid was aspirated from the inguinal eruptions. No lymphedema or difference in size between the lower extremities was observed. The face, palms and soles were erythematous, as in scarlet fever. No skin trauma, ulceration, or suppuration was detected in the affected lesions or on the left lower limb.

The results of laboratory studies at admission (WBCs, 19,100/mm with 93% polymorphonuclear leukocytes; CRP, 19.2 mg/dl) indicated a bacterial infection. No subcutaneous transparent shadow or skeletal abnormality was observed on the radiographs of the affected areas. Chest X-rays did not show any abnormalities. Bacterial cultures were performed twice using the aspirated fluids of the eruptions, and also using the biopsy skin, pharyngeal mucosa, and blood. Alpha- and gamma-streptococcus, *Staphylococcus aureus*, *Neisseria* spp., and *Haemophilus parainfluenzae* were cultured from the pharyngeal mucosa, but the other sources were negative. Antistreptokinase was 320-fold elevate, and the antistreptolysin O level was 194 IU/ml on the 4th day of hospitalization, but these values rose 5,120-fold and to 228 IU/ml, respectively, on the 13th day of hospitalization. These results confirmed a diagnosis of recurrent cellulitis caused by a streptococcal infection.

The patient was treated with a combination of three types of intravenous antibiotics: piperacillin sodium 4.0 g/day, cefotiam hexetil hydrochloride 2.0 g/day, and minocycline hydrochloride 200 mg/day from the first day of hospitalization. After start of this treatment, the first clinical features that were observed (erythema, swelling, and pain) disappeared, leaving desquamation of the superficial skin layers of the palms and soles. The numerous keloid-like skin eruptions were left as scattered pigmented maculae (Fig. 2*B*), and several larger eruptions in the left inguinal region changed into soft, atonic bumps, and so minocycline and cefotiam were withdrawn, and piperacillin was continued at the reduced dosage of 2.0 g/day for 9 days. No fluid discharge was emitted from any of the skin lesions, as mentioned above.



(*a*) (*b*)

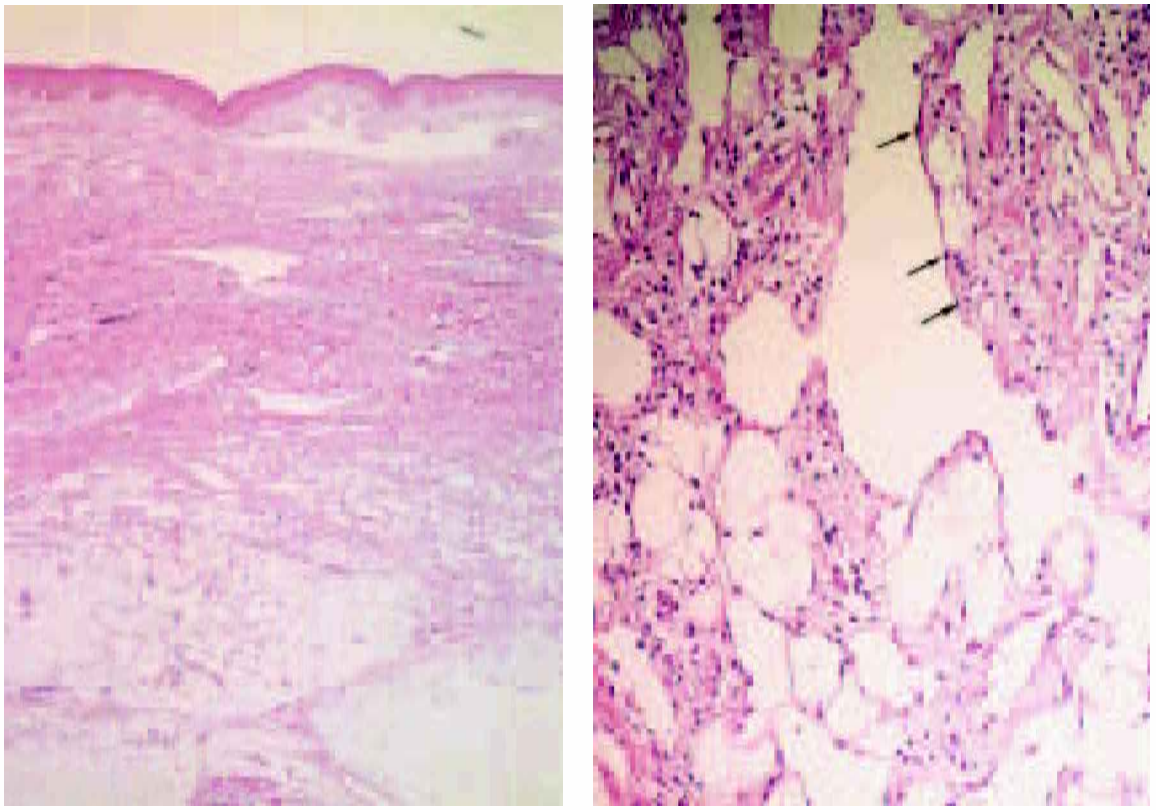
Fig. 2. A: Close-up view of the scattered skin eruptions observed on the left waist at admission. The appearance resembles that of keloids. *B*: Clinical appearance of the left aspect of the waist and thigh after treatment. Numerous scattered pigmented maculae are seen.

Histologic examination of the keloid-like lesions of the thigh on admission revealed many dilated lymphatic vessels of varying size, which increased in number throughout the edematous dermis and subcutaneous fatty tissue, especially in the upper dermis and in the transitional zone between the dermis and the subcutaneous fatty tissue. The dermis was markedly edematous and was infiltrated diffusely by inflammatory cells, chiefly neutrophils, in the stroma (Fig. 3). No protrusion of the lymphatic vessels into the epidermis was observed in any of the histological thin sections. The endothelial cells were negative for factor VIII-related antigen. Hyperpigmentation was recognized along the basal layer of the overlying epidermis. The histology of the scrotum was similar to that of the thigh excluding the inflammatory cell infiltration, and dilated lymphatic vessels were also found in the scrotal muscle tissue. Some parts of the walls of these vessels were thickened, but the dermis and subcutaneous tissues did not show any fibrotic changes. No bacilli were recognized in the sections stained with Giemsa and Gram stains.

Pedal lymphography of the left lower limb was performed. In the left groin and upper femoral area, tortuously dilated lymphatic vessels could be observed (Fig. 4*A*). The flow of the contrast material to the iliac and external iliac lymphatics could be roughly traced in the normal position along the left side of the pelvic space and of the vertebrae. After 24 h, images of the dilated lymphatic vessels had disappeared, but spotty stasis of the contrast medium (dermal backflow) could be recognized on X-ray films of both the area corresponding to the distribution of the skin eruptions and even the margin of the body, so the sites were thought to match the distribution of the skin lesions (Fig. 4*B*). Computed tomography of the lower trunk and thigh performed 11 days after lymphography showed that the contrast material used in lymphography was still present in the subcutaneous lesions of the left thigh and in the inguinal area, but revealed no obstructive tumor mass or other abnormal finding in the abdominal or pelvic cavities or the inguinal area.

DISCUSSION

Lymphangioma is observed in any site with a predilection along the courses of embryological development of lymphatics, mainly in the groin, neck and axilla. Multiorgan, multifocal cases, or diffusely distributed cases, even in a single area of the body, have been reported (1–6). In such cases, lymphedema, chylothorax, chylous



(*a*) (*b*)

Fig. 3. Histology of the biopsy specimen of the left thigh at admission. *A*: Many dilated lymphatic vessels of varying size are recognized through the edematous dermis and subcutaneous fatty tissue (left, hematoxylin-eosin, ×5). *B*: Close-up view of the dilated vessels of the subcutaneous tissue (right). The dilated vessels are lined by well-differentiated endothelium (arrows) and inflammatory cell infiltration is seen in the stroma (hematoxylin-eosin, ×25).

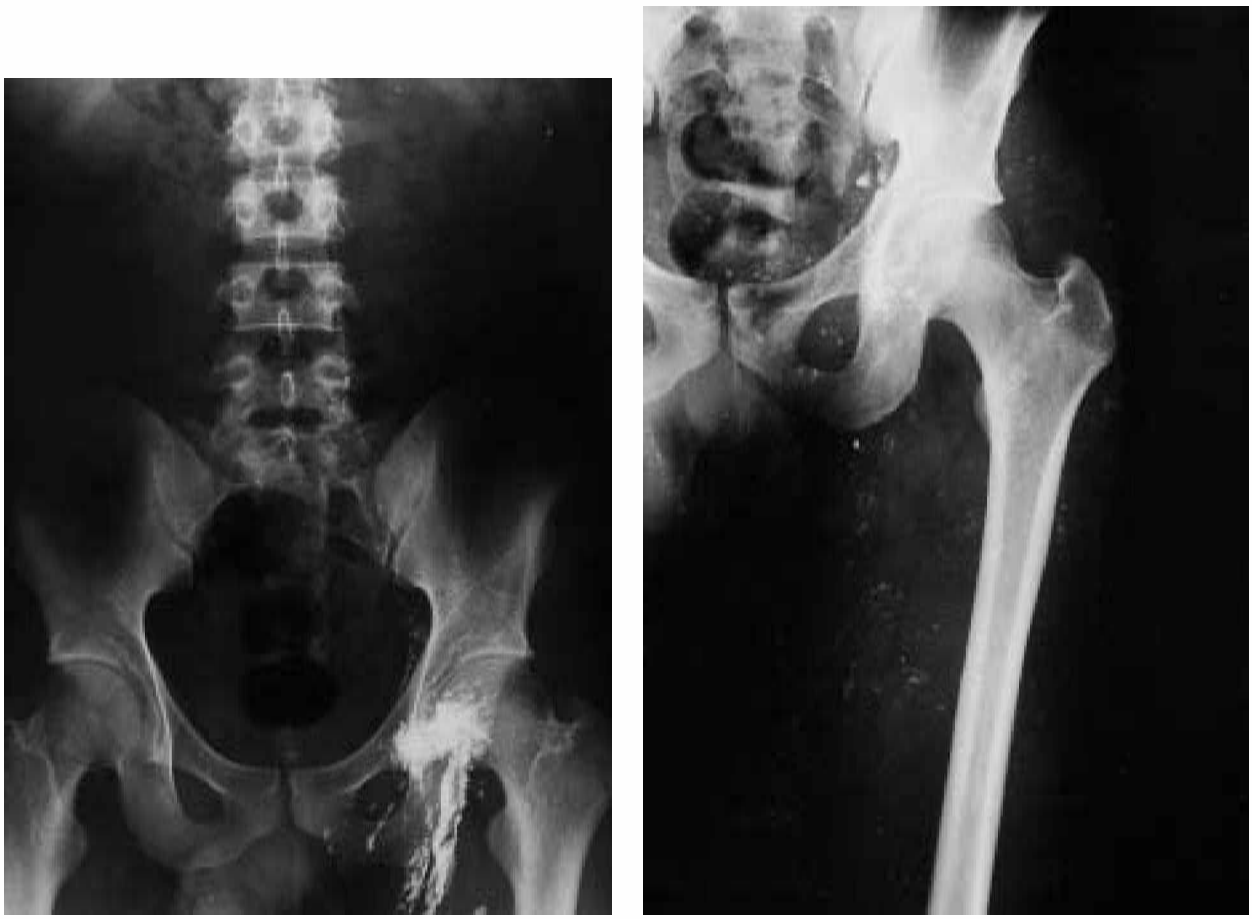


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Fig. 4. A: A lymphangiogram (immediately after injection) showed dilated lymphatic vessels in the left inguinofemoral area, and a poor flow of contrast medium in the normal sites of the iliac lymphatics. *B*: Spotty stases of contrast medium (dermal backflow) observed 24 h after injection were seen even at the surface of the body, and so their locations are thought to correspond to those of the skin lesions shown in Figures 1, 2, and 3.

ascites, localized gigantism, protein-losing enteropathy, weeping scrotum, vulvar and vaginal chylorrheas, xanthoma, and verrucous changes of the skin are associated with or result from inadequate lymphatic drainage (7–9). The peculiar keloid-like skin eruptions and marked swelling of the scrotum observed on admission of this patient were transient saccular dilatations of cutaneous lymphatic vessels. The expressions of bullae, vesicles, and chylous vesicles are not appropriate for these eruptions. Abnormal lymphatic flow including chylous reflux of the skin usually accompanies lymphedema, chylous vesicles, and chylous oozing from the skin. The dermal backflow observed in the lymphangiograms indicates an impairment of the flow of lymph along the subcutaneous lymphatics (10). In this case, correspondence between clinical skin eruptions and lymphographic dermal backflow was demonstrated. Recurrent infectious episodes are occasionally associated with lymphangioma or lymphedema, but the patient had no leg lymphedema or chylous skin disorder.

Pigmented macules as clinical manifestations of lymphangioma have not previously been reported, except in the case of a dog with generalized lymphangiectasis, in which the skin overlying the left axillar and inguinal swellings was hyperpigmented with multiple small vesicles (11). Fossum et al., who conducted this study of lymphangiectasia in a dog, did not mention the pathogenesis of the pigmentation. Although the hyperpigmentation along the basal layer of the overlying epidermis was histologically confirmed, the pathogenesis of the pigmented maculae corresponding to the saccular dilatations is unclear. These pigmented maculae may have been a manifestation of cutaneous chylous reflux or a sequela of inflammation. A long follow-up of the degree of these pigmented maculae may reveal whether these pigmented spots are likely to fade or increase. In patients with cutaneous chylous reflux, this condition is usually accompanied by leg lymphedema, and cases unassociated with lymphedema are rare (7, 12). The degree of inadequacy of lymphatic flow and the severity of chylous vesicle formation may depend on the degree and site of the lymphatic abnormality, on the remaining lymph flow, and on the collateral lymph flow in the region. No case has been recorded in the literature in

which vesicles have developed from saccular dilatation, but with time, stronger or prolonged, continuous lymphatic pressure may form chylous vesicles in this patient, and the possible future development of leg lymphedema cannot be ruled out (10, 13).

It was difficult to judge whether the primary disorder in this patient was dysplasia of the iliac lymphatic vessels (which were poorly traced on lymphangiograms) or dilatation of the inguinofemoral lymphatics, and indeed the latter could be described as a consequence of the former. In addition, cutaneous lymphatic dilatation could have been induced by the failure of the lymphatic drainage of the dilated inguinofemoral lymphatics. However, there is little doubt that the primary abnormality was in the iliac or inguinal lymphatics, beneath the level of the cisterna chyli; and an increase in the number of lymphatic vessels and of their anastomoses in the dermis was clearly recognized microscopically.

In cases of large abnormality of lymph vessels associated with lymphedema of the extremities, local surgical management and local excision are useless rather than difficult. Radical megalymphatic excision with ligation of the transected lymphatic ducts and redirection of the chylous flow toward the thoracic duct and away from the major trunk of caudal lymphatic bed may be helpful in improving the abnormal lymphatic flow (14–17). Prophylactic antibiotic coverage to protect against infection may prove to be useful in this case (18, 19).

The term "lymphangioma" is widely used to describe benign neoplastic or hamartomatous conditions of lymph vessels, but our case cannot be categorized as the generally accepted cutaneous lymphangioma: capillary lymphangioma (lymphangioma simplex or lymphangioma circumscriptum), cavernous lymphangioma, and cystic types of lymphangioma (cystic hygroma) (16). This case is difficult to match with the typical concept of lymphangioma, and there is some terminological confusion regarding lymphatic lesions, especially with the terms "lymphangiomatosis" and "lymphangiectasis". As Tazelaar et al. (1) stated, the term "lymphangiectasis" should be reserved to describe those extremely unusual congenital or secondary lesions in which the primary alteration is the dilatation of existing lymphatic channels, without any increase in their number or complexity; while the term "lymphangiomatosis" should be used for those diffuse lesions characterized primarily by an increased number of complex anastomosing lymphatic channels in which dilatation is a secondary phenomenon. We prefer the term "cutaneous lymphangiectasis" for our case, and the name "occult cutaneous lymphangiectasis", because our patient did not show any skin eruption, with the exception of pigmented maculae not obviously indicating lymphatic origin, and did not demonstrate any neoplastic tumor mass in the deeper lymphatic system by pedal lymphography. The affected skin area was relatively large.

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