Quiz-1 & 2

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***Key words**: cutaneous infiltrate, leukemia.*

QUIZ - 1:

Pruritic Generalized Rash in a Middle Aged Man

Case History:

A 45-year-old Pakistani male patient was admitted in the medical ward with history of itchy generalized rash of 4 months duration and fever of 2 weeks duration.

General examination revealed a well built man, with generalized lymphadenopathy. Lymph nodes in the cervical, axillary and inguinal groups were enlarged, as firm discrete non-tender nodes. Rest of the general examination was within normal limits.

Cutaneous examination revealed diffuse erythema, with fine scaling over chest, upper back, neck and upper limbs. In addition, there were erythematous infiltrated papules, over face, forehead, eyelids, ear lobes (Fig.1, 2).

Perepheral blood examination showed a total WBC count of 56,000/cm, with a lymphocyte differential count of 81% and blast cells 6%. Other investigations including blood chemistry, autoantibodies, x-rays ad ultrasound of chest, pelvis, abdomen were within normal limits.

Skin biopsy was performed (Fig.3, 4).

What is your diagnosis?

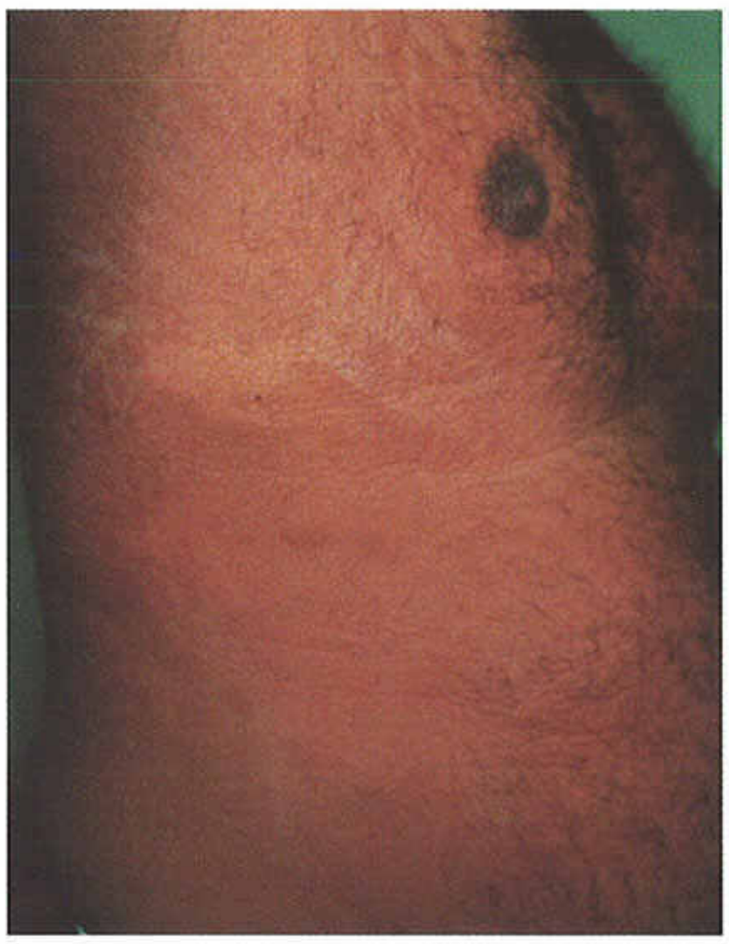
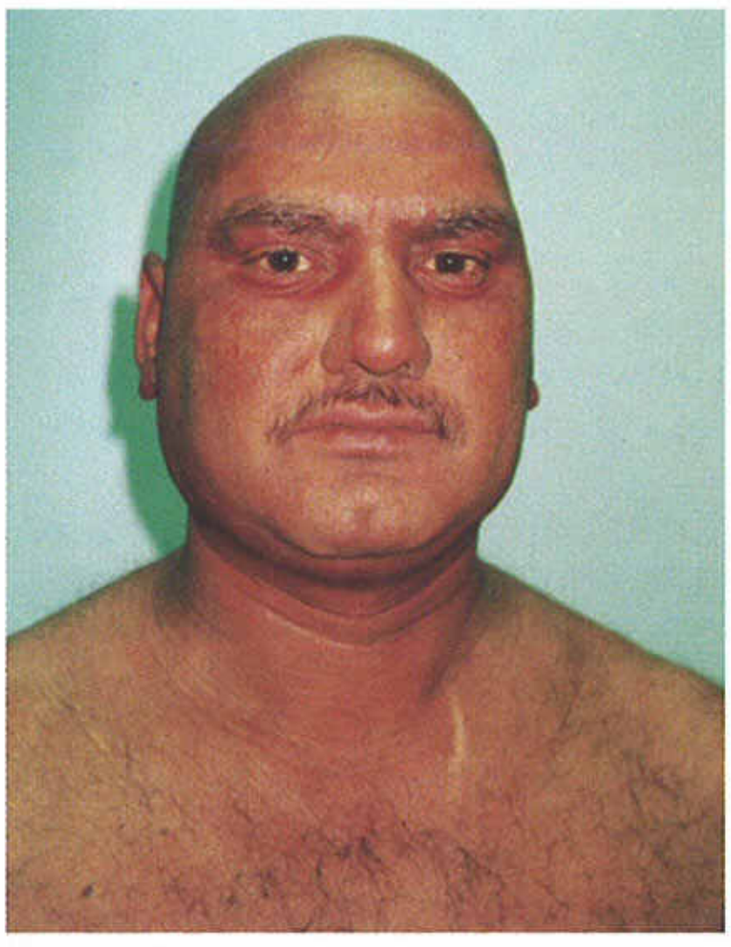


Figure 1



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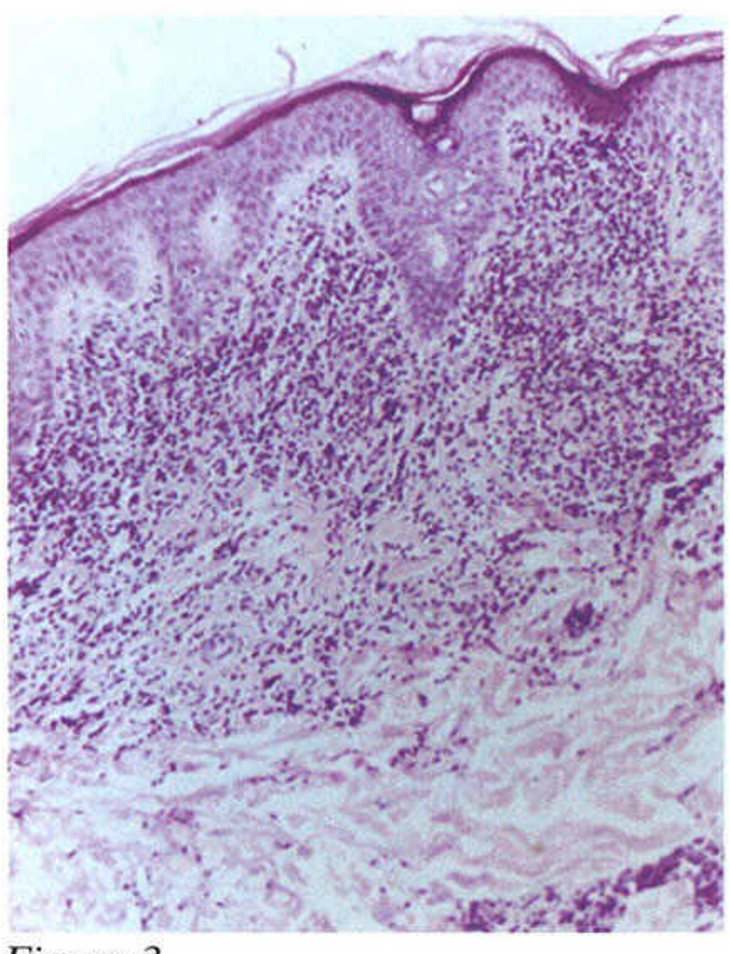

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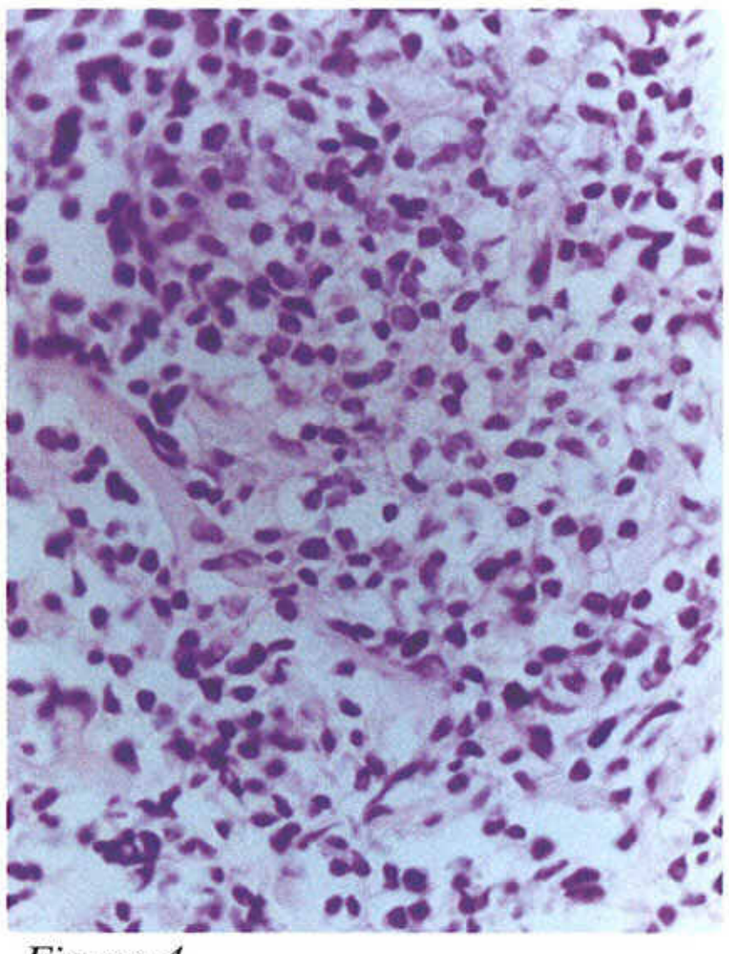


Figure 4

QUIZ - 2 :
Erythroderma in an elderly woman

Case History:

A 69-year-old Bahraini lady presented with itchy, generalized skin rash of two years duration. She had been treated previously with several courses of local and systemic steroids without any benefit. There were no other significant complaints.

Examination revealed a poorly built elderly woman. General examination was with in normal limits. Cutaneous examination showed generalized erythroderma, with scaling and erythema, with multiple excoriation marks (Fig-1,2). There was no significant lymphadenopathy.

Routine investigations such as peripheral blood smear, blood chemistry, autoantibodies, X-rays and ultrasound of chest, abdomen and pelvis were with in normal limits. Skin biopsy was performed (Fig-3, 4).

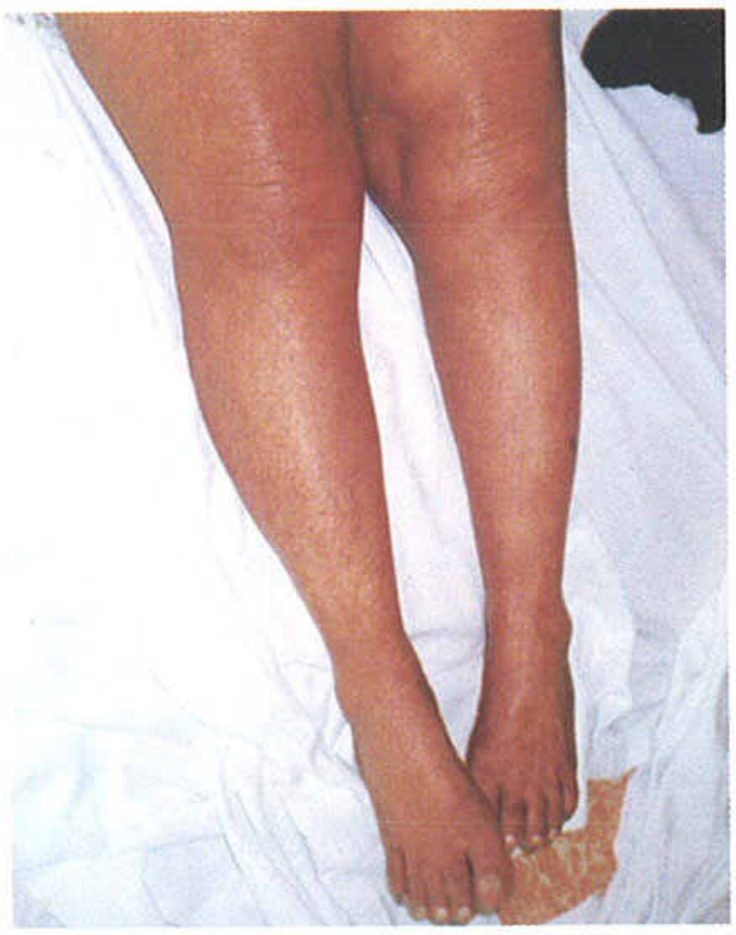
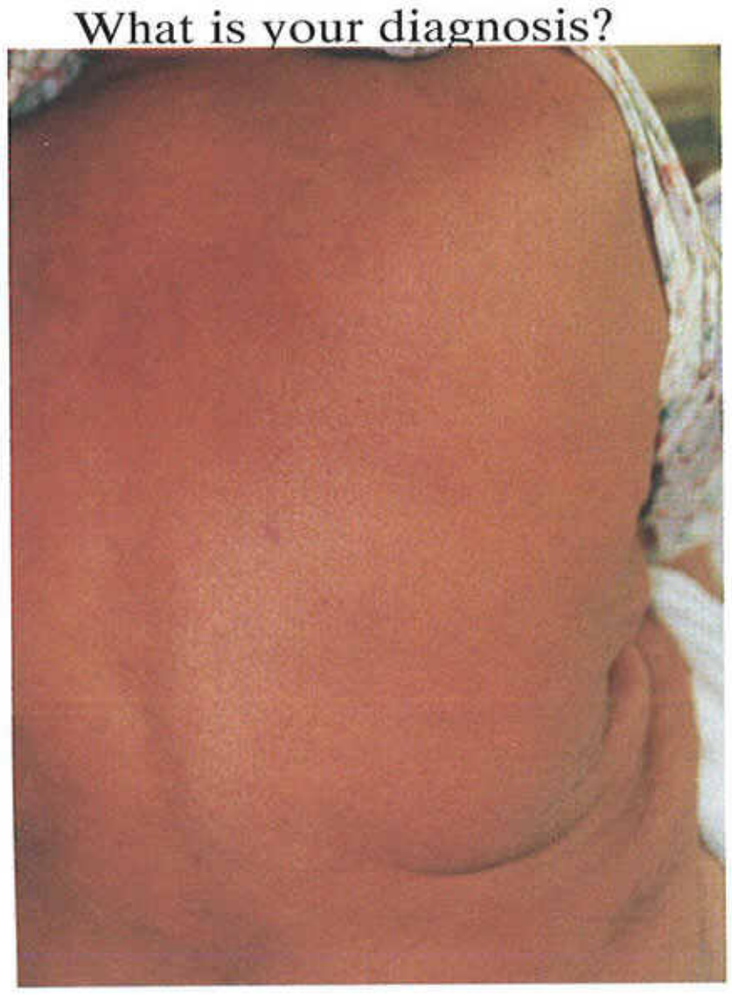


Figure 1

What is your diagnosis?



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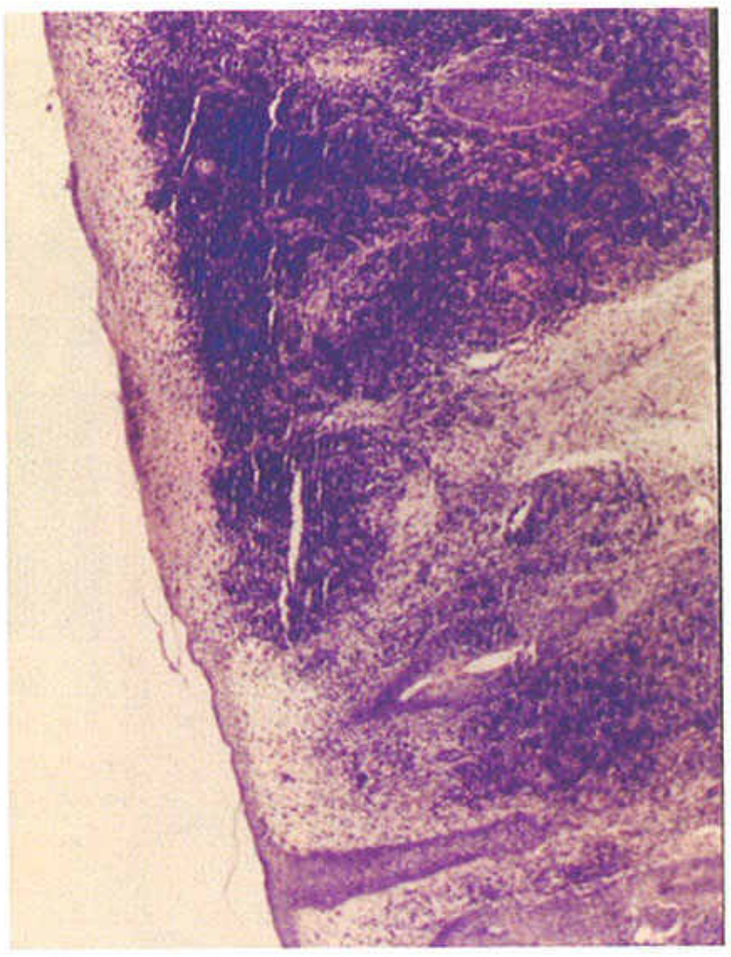

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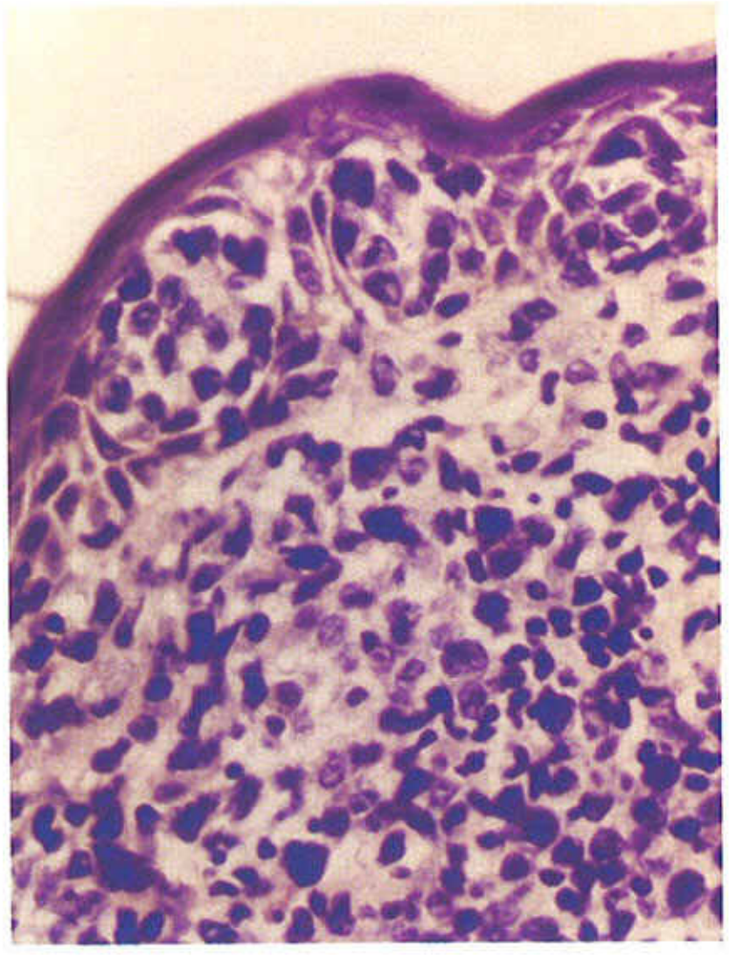

Figure 4

Answer to Quiz 1:
Cutaneous leukemic infiltrate due to chronic lymphocytic leukemia.

Microscopic findings; Skin biopsy showed normal epidermis. Dermis showed a diffuse dense infiltrate extending around blood vessels and appendages, into lower dermis and subcutis. The cells were of variable size and shape, many of them with mitotic figures. The infiltrate extended around blood vessels and appendges, and in between collagen fibres. Immunohistochemical stains showed these to be of B cell lineage. The picture was suggestive of atypical lymphoid infiltrate. Further investigations including lymph node and marrow biopsy were advised to rule out cutaneous lymphoma/leukemic infiltration.

Biopsy of bone marrow and cervical lymph node confirmed the diagnosis of chronic lymphocytic leukemia.

Discussion:

Cutaneous manifestation of leukemia cutis include pruritus, papules, tumours, erythroderma, purpuric lesions, echymoss, and bullae [1]. Of these, erythroderma and bullae are said to occur specifically in Chronic lymphocytic leukemia (CLL)[1].

Dermatomyositis like lesions have been reported in Tcell CLL [2]. Papules often occur in areas previously affected by herpes zoster and herpes simplex[3]. Presence of leukemia cutis in a patient was previously thought to carry poor prognosis. However more recent

reports suggest that this may not always be true [4].

Histological picture [5] shows a diffuse, heavy infiltration of leukemic cells in the dermis. The infiltrate, often extends in between appendages and collagen fibres, and into subcutis. Extensive involvement and disruption of blood vessels and adnexa are some of the characteristic findings in a leukemic infiltrate. There is often a wide range of histological changes in different types of leukemia and among different patients with the same type of leukemia. The cell morphology is not sufficiently diagnostic to differentiate a well differentiated cutaneous lymphoma from a leukemic infiltrate of CLL. Hence, morphologic and histochemical studies of peripheral blood, lymph node and marrow should be carried out [5].

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Answer to Quiz 2:

Erythroderma mycosis fungoides.

Skin biopsy showed epidermis with epidermotropism of mononuclear cells. The cells were large and atypical, often arranged as Pautrier microabscesses. The nuclei were of variable size and shape. Upper dermis showed diffuse band-like infiltrate, consisting of mononuclear cells. Many of these cells were large, atypical, with variable size and shape, and often with mitotic figures. The infiltrate did not extend into lower dermis and subcutis. Immunohistochemical stains confirmed these cells to be of T cell lineage. Bone marrow biopsy showed scattered group of lymphoid cells, suggestive of marrow infiltration.

Discussion:

Erythroderma is an important cutaneous manifestation of mycosis fungoides, common in the elderly and corresponds to T4 stage in TNM classification [1,2,3]. When it occurs in the presence of significant number of atypical lymphoid cells in peripheral blood, it is referred to a Sezary syndrome. It has been recognized that a significant number (up to 75%) of patients with erythrodermic mycosis fungoides have visceral involvement even in the absence of significant clinical findings and hence the presence of erythroderma in such patients warrants detailed investigation. The prognosis in these patients is not satisfactory, even in the absence of visceral disease [50% five-year survival rate] [1,2].

Histological picture on erythrodermic mycosis fungoides may show either the epidermotropic or nonepidermotropic picture [4,5,6]. The latter subgroup has poor prognosis, because of coexistence of lesions in lymph nodes and peripheral blood involvement. The pathognomonic changes of mycosis fungoides, such as epidermotropism and epidermal hyperconvoluted cells, are less pronounced in erythrodermic stage, than in plaque/tumor stage and hence interpretation of such a skin biopsy is often difficult [4]. It has been reported that age at onset, duration of lesion, extent of involvement and absence of epidermotropism on histology are useful parameters to assess prognosis [3,7].

Photopheresis is recommended as the first choice treatment in erythrodermic mycosis fungoides. Other therapeutic alternatives include methotrexate, interferon-2@ and systemic chemotherapy [1,2].

Erythroderma in the elderly presents an important diagnostic challenge to both the clinician and pathologist. It is of paramount importance to differentiate benign erythrodermas from erythrodermic mycosis fungoides [6]. Presence of epidermotropism, pautrier microabscesses in skin biopsy, additional investigations of lymph nodes and peripheral blood, careful follow-up are necessary to arrive at a correct diagnosis.

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