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A CASE OF PARANEOPLASTIC ELASTOSIS PERFORANS SERPIGINOSA ASSOCIATED WITH T-CELL NON-HODGKIN LYMPHOMA

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Abstract

Perforating dermatoses are a group of diseases characterized by transepidermal elimination of dermal material. This group is divided into four subgroups: Kyrle disease, reactive perforating collagenosis, elastosis perforans serpiginosa and perforating folliculitis. They can be primary or secondary. Secondary perforating dermatoses may develop due to chronic renal failure, diabetes mellitus, autoimmune diseases and malignancies. With elastosis perforans serpiginosa, the rarest of this group, transepidermal elimination of dermal elastin tissue is present. It may be idiopathic but may also develop Down syndrome, Ehlers-Danlos syndrome, Marfan's syndrome, osteogenesis imperfecta, acrogeria, Rothound-Thomson syndrome, cutislaxa and due to using D-penicillamine medication. Here in is presented, as the second case in the literature, a 60-year-old patient with paraneoplastic EPS associated with T-cell non-Hodgkin lymphoma.

Keywords: elastosis perforans serpiginosa, non-Hodgkin lymphoma, paraneoplastic Syndromes.

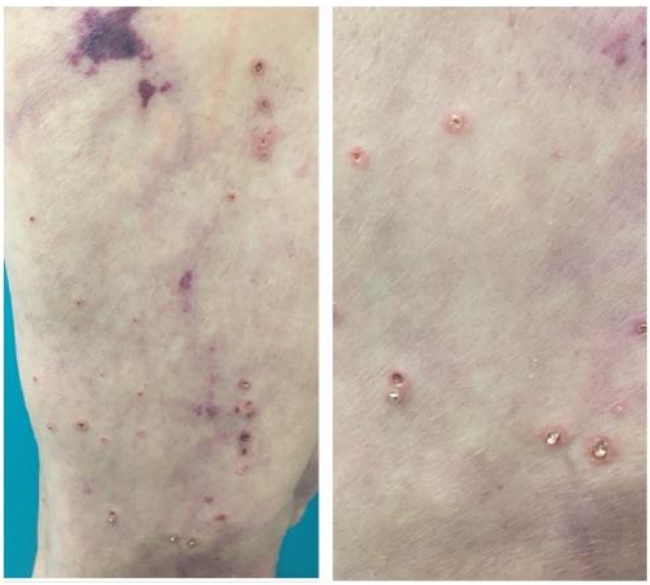
Introductıon:

Perforating dermatoses are a group of diseases characterized by transepidermal elimination of dermal material. This group is divided into four subgroups: Kyrle disease (KD), reactive perforating collagenosis, elastosis perforans serpiginosa (EPS) and perforating folliculitis.They can be primaryor secondary. Secondary perforating dermatoses may develop due to chronic renal failure, diabetes mellitus, autoimmune diseases and malignancies. In the transepidermal elimination of dermal materialin perforating dermatoses the material eliminated comprises: dermal material without collagen-elastin tissue in KD; elastin tissue in EPS; collagen in reactive perforating collagenosis; and degenerated hair follicles in perforating folliculitis. Elastosis perforans serpiginosa, the rarest of this group [1,2].It may be idiopathic but may also develop Down syndrome, Ehlers-Danlos syndrome, Marfan's syndrome, osteogenesis imperfecta, acrogeria, Rothound-Thomson syndrome, cutislaxa and due to using D-

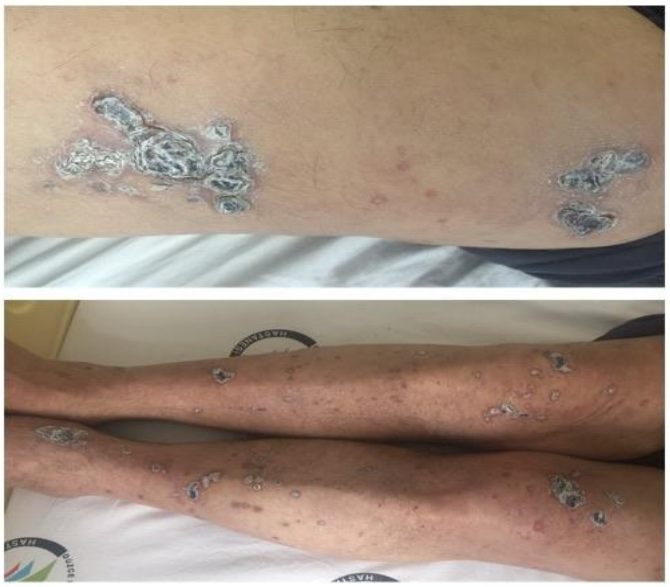
penicillamine medication [3].Herein is presented the case of a 60-year-old patient with T-cell non-Hodgkin lymphoma-associated paraneoplastic EPS with presentation of atypical lesions.

Case:

A 60-year-old male patient was admitted to the hospital due to swelling in the neck region lasting for two months.His history showedno significant of disease.Physical examination revealed lymphadenopathy in the left cervical region. The presence of cervical, mediastinal, axillary and inguinal lymphadenopathy and splenomegaly were detected in the examinations carried out. Excisional biopsy of the lymphadenopathy in the patient's neck region identified T-cell non-Hodgkin lymphoma(angioimmunoblastic lymphomas subtypes). During this period, our clinic was consulted by the patient, who had developed eruptions on both lower extremities. From the history it was learned that the lesions had started three weeks ago. The existing lesions expanded and had new lesions and that these lesions were accompanied by itching.The dermatological examination detectedthe presence of dome-shaped papules of 10-15 mm in size withkeratotic plugs in the middle and erythematous borders (Photograph 1).The lesions were on both legs and there were no trunk and arm involvement.A punch biopsy specimen was obtained from these lesions. The skin biopsy revealed transepidermal elimination of basophilic inflammatory debris containing coarse elastic fibersshowing positivity with van Gieson's elastic stain (Figs. 1 and 2).



Photograph1. Lateral region of the right leg showingdome-shaped papules 10-15 mm in sizewithkeratotic plugs in the middle and erythematous borders



Photograph 2. On both lower extremities the surface is covered with dry plaque lesions with erythematous borders.

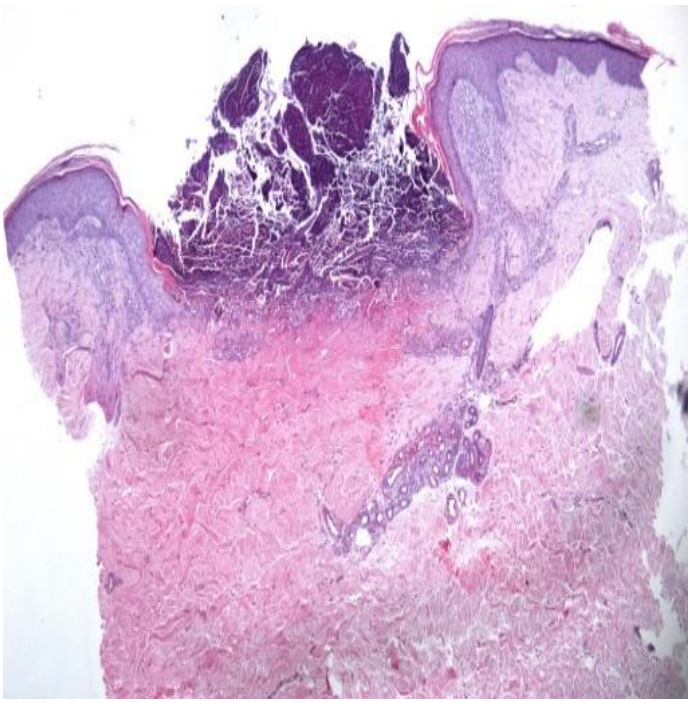


Figure 1.Low-magnification view ofelastosis perforans serpiginosa:

Transepidermal elimination of basophilic material (HE, 40×)

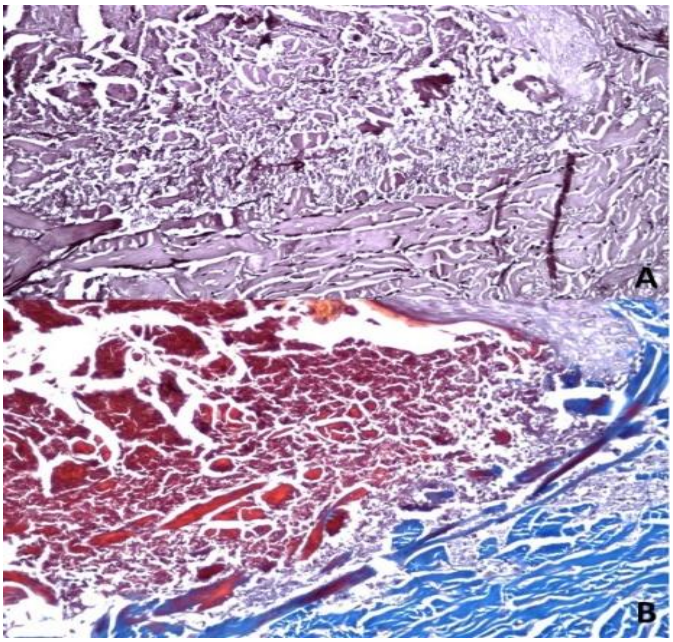


Figure 2.A: Transepidermal elimination of abnormal elastic tissue (van Gieson elastic stain, 400×); B: Masson's trichrome stain showing negativity for collagen (Masson's trichrome, 400×).

Figure1. Low-magnification viewof elastosis perforans serpiginosa:Transepidermal elimination of basophilic material (HE, 40×) **Figure 2.**A: Transepidermal elimination of abnormal elastic tissue (van Gieson elastic stain, 400×); B: Masson's trichrome stain showing negativity for collagen (Masson's trichrome, 400×).

Within two weeks, it was observed that the patient's lesions had gradually enlarged and upon merging with each other, formed a surface covered with dry plaque lesions with erythematous borders(Photograph 2).

The patient was offered moisturizer treatment for skin lesions. Considering that it could be paraneoplastic syndrome , the patient was followed up. CHOP (Cyclophosphamide, Doxorubicin, Vincristine and Prednisone) treatment protocol for T-cell non-Hodgkin lymphoma was started by hematology clinic. Second round was continued with the addition of etoposide to the CHOEP. No new lesion was removed after the first cure.The lesions were regressed one month after chemotherapy. The onset of symptoms shortly before diagnosis of underlying malignancy, absence of a new lesion, and the rapid response of the skin following chemotherapy are consistent with a paraneoplastic dermatosis.

After obtaining the written permission of the patient, the photographs were taken.

Dıscussıon:

In the transepidermal elimination of dermal materialin perforating dermatoses the material eliminated comprises:dermal material without collagen-elastin tissue in KD; elastin tissue in EPS; collagen in reactive perforating collagenosis; and degenerated hair follicles in perforating folliculitis [1,2].EPS is a rare skin condition characterized by hyperkeratotic papules, transepidermal elimination of abnormal elastic fibers, and focal dermal elastosis[3]. EPS, which was identified by Lutz in 1953. It may be idiopathic but may also develop Down syndrome, Ehlers-Danlos syndrome, Marfan's syndrome, osteogenesis imperfecta, acrogeria, Rothound-Thomson syndrome, cutislaxa and due to using D-penicillamine medication[3,4].However, it remains unclear whether the relationship between these conditions and EPS is causal or coincidental, as EPS also commonly occurs in individuals without anyunderlying disease. EPS associated with malignancy has been reported only in a stage-4 ovarian cancer patient [5].In our case, the lesions were diagnosed together with the malignancy, the may be stop of new lesions with chemotherapy, the regression of old lesions, and the support of paraneoplastic syndrome associated with malignancy of lesions. Despite the fact that EPS was reported as paraneoplastic in two patients along with our case, reactive perforating collagenosishas been reported as paraneoplastic in 15 patients in the literature, including five patients with lymphoproliferative diseases and 10 patients with solid cancers (prostate cancer: 3, hepatocellular cancer: 2, breast cancer: 1, colon cancer: 1, thyroid cancer: 1, renal cancer: 1, periampullary cancer: 1) [6,7].EPS lesions are characterized by serpiginous, arcuate, annular or clustered 2-4 mm keratotic papules. Areas of predilection include the neck, face, nape, arm and flexural regions [4,8]. The exact etiology and pathogenesis of EPS is unknown. It is believed that the local skin inflammation induces formation of perifollicular or intraepithelial tunnels through which abnormal elastic fibers are secreted [9]. It has been shown thata 67-kD elastin receptor is

present in the keratinocytes associated with the elimination of elastin tissue in EPS and that elastin-keratinocyte interaction plays an important role in the transepidermal elimination of elastin [5].In the biopsies an increased amount of elastic fibers can be found, both just underneath the epidermis and in the reticular layer of the dermis. The fibers are compacted, twisted and fragmented, what can be clearly seen in the staining for elastic fibers such as in van Gieson staining [9].

The location and clinical presentation of the lesions in our case did not resemble those typical of EPS. This, together with the fact that the van Gieson staining showed positive and the Masson's trichrome staining showed negative for transepidermal elimination material in the tissue sections, led to the diagnosis of the case as atypical clinical EPS.

Although lesions may spontaneously resolve, they often persist for longer periods.There is no effective standard treatment of EPS, and so far there have been only single reports of the use of different drugs for systemic and topical therapy.Suggested treatment options include cryotherapy, intralesional and topical corticosteroid therapy, topical calcipotriol, topical tretinoin, oral isotretinoin. However, no 'gold standard' therapy exists among these treatment modalities [3].

In conclusion,In our case, the onset of symptoms shortly before diagnosis of underlying malignancy and the rapid response of the skin following chemotherapy are consistent with a paraneoplastic dermatosis.We present a memorable case of paraneoplastic elastosis perforans serpiginosa in association with malignancy T-cell non-Hodgkin lymphoma.

There is no conflict of interest between authors.

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