Case Report

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Malignancy smell in the air: widespread eroded, hemorrhagic, and lichenified plaques in an older man

Havada malignite kokusu var: yaşlı erkekte yaygın erode, hemorajik ve likenifiye plaklar

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Abstract

Paraneoplastic pemphigus (PNP) is a rare autoimmune mucocutaneous disease associated with underlying neoplasia. It is typically characterized by painful mucosal erosions and dark, patchy skin eruptions. A 66-year-old male was admitted to the internal medicine outpatient clinic with complaints of suddenly started rashes, loss of appetite, dyspepsia, weakness, and unexplained gross weight loss. The patient was cachectic, and physical examination revealed widespread eroded, erythematous, thick-middle, yellow-pitted, lichenified plaques on bilateral arms and legs and the hands and feet dorsum. Scattered seborrheic keratosis lesions on the trunk were also detected. He had microcytic anemia with elevated CA 19-9 measures. Abdomen computed tomography showed a malignant mass in the antrum. An endoscopic biopsy of the gastric mass revealed poorly differentiated adenocarcinoma composed of discohesive signet ring cells, and the skin punch biopsy was compitable compatible with paraneoplastic pemphigus.

In patients with rapidly developing skin lesions with constitutional symptoms, underlying malignancies should be kept in mind.

Keywords: Paraneoplastic pemphigus, Leser Trelat, gastric adenocarcinoma.

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Öz

Paraneoplastik pemfigus (PNP), altta yatan neoplazi ile ilişkili nadir bir otoimmün mukokutanöz hastalıktır. Tipik olarak ağrılı mukozal erozyonlar ve koyu yamasal döküntülerle ile karakterizedir. Altmış altı yaşında erkek hasta ani başlayan kızarıklık, iştahsızlık, hazımsızlık, halsizlik ve açıklanamayan aşırı kilo kaybı şikayetleri ile dahiliye polikliniğine başvurdu. Kaşektik olan hastanın fizik muayenesinde her iki kol ve bacakta, el ve ayak sırtında yaygın aşınmış, eritematöz, ortası kalın, sarı-çekirdekli, likenifiye plaklar saptandı. Gövdede dağınık seboreik keratoz lezyonları da olan hastanın yüksek CA 19-9 düzeyi ile mikrositer anemisi vardı. Karın bilgisayarlı tomografisinde antrumda malign bir kitle görüldü. Mide kitlesinin endoskopik biyopsisinde diskohezif taşlı yüzük hücrelerinden oluşan az diferansiye adenokarsinom saptandı ve cilt biyopsisi paraneoplastik pemfigus ile uyumlu izlendi.

Yapısal semptomlarla birlikte hızla gelişen cilt lezyonları olan hastalarda altta yatan maligniteler akılda tutulmalıdır.

Anahtar kelimeler: Paraneoplastik pemfigus, Leser Trelat, gastrik adenokarsinom.

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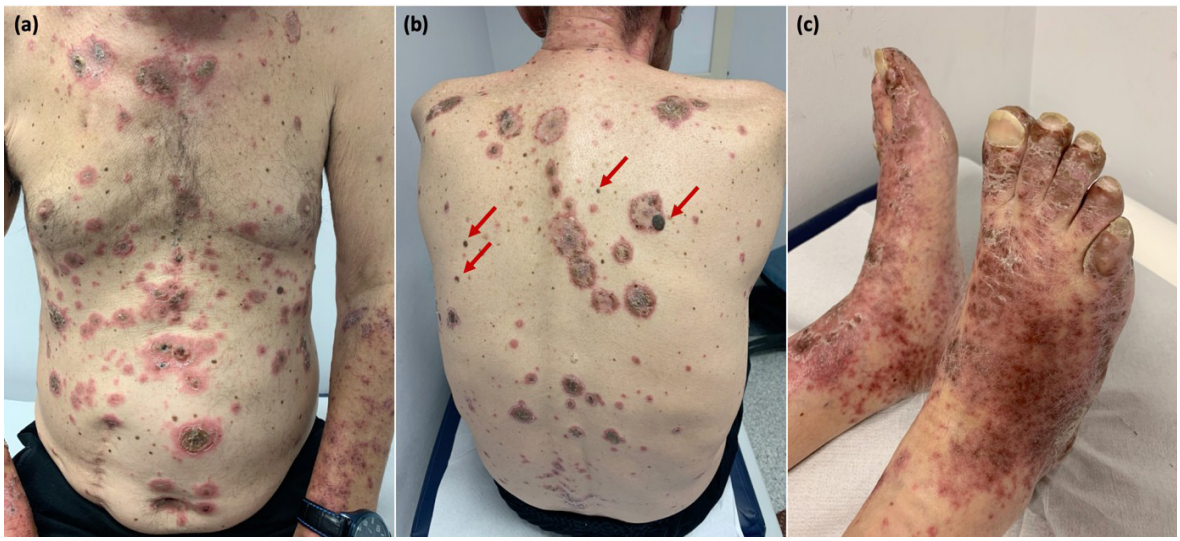
Introduction

Some malignancies may trigger several cutaneous manifestations. Although recognition of some typical paraneoplastic dermatoses may lead to early diagnosis of a neoplasm, some may progress with atypical involvements [1]. Sometimes, patients may apply to the physician for the first time not only with symptoms related to the primary diagnosis but also with cutaneous manifestations that seem unrelated.

Here, we present a rare case of metastatic gastric adenocarcinoma applying to the general internal medicine outpatient clinic with complaints of constitutional symptoms and widespread cutaneous manifestations.

Case report

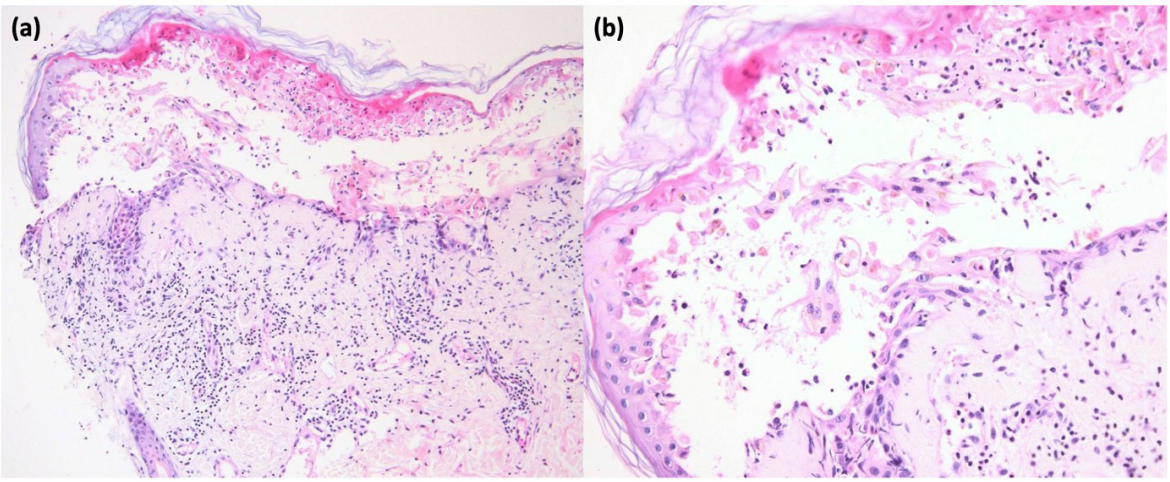
A 66-year-old male patient with type 2 diabetes, hypertension, and coronary artery disease was admitted to the general internal medicine outpatient clinic with complaints of rashes that started on the hands and spread to the trunk and feet 2 months ago. It was accompanied by loss of appetite, dyspepsia, weakness, and unexplained gross weight loss (15 kg per 2 months). The patient was cachectic, and physical examination revealed widespread eroded, erythematous, thick-middle, yellow-pitted, lichenified plaques on bilateral arms and legs and dorsum of the hands and feet. Scattered seborrheic keratosis lesions on the trunk were also detected (Picture 1). He was on metformin, dapagliflozin, telmisartan, metoprolol, and clopidogrel treatment for 4 years and had a 20-pack-year smoking history. Laboratory studies revealed microcytic anemia with a hemoglobin level of 8.3 g/dL, leukocyte count of $5.6x10^3$/ml, platelet count of $321x10^3$/ml, creatinine level of 1.5 mg/dL, serum sodium 126 mEq/L, potassium 3.58 mEq/L, chlorine 94 mEq/L, hemoglobin A1C 6%, normal liver functions, and negative acute phase reactants. He was admitted to the hospital ward, and intravenous fluid resuscitation and topical 0.05% clobetasol propionate were administered. Antinuclear antibody, anti-neutrophil cytoplasmic antibody, rheumatoid factor, indirect immunofluorescence on rat bladder epithelium, prostate-specific antigen, carcinoembryonic antigen, and alpha-fetoprotein were negative, while CA 19-9 was 65.7 U/mL (normal range: 0-35). Subsequently, abdomen computed tomography showed a malignant mass in the antrum, 12 mm thickening in the right adrenal gland, three heterogeneous contrasted lesions, the largest of which is 34x24 mm in the left adrenal gland, and multiple paraaortic, paracaval, and portal hilar lymphadenopathies up to 36x46 mm in size. Upper endoscopy revealed an ulcerovegetative antral mass surrounding the pylorus.



Picture 1. (a) and (b) Widespread eroded, erythematous, thick-middle, yellow-pitted, lichenified plaques on the trunk, (b) arrow signs; scattered seborrheic keratosis lesions on the trunk, (c) dorsum of the feet

An endoscopic biopsy of the gastric mass revealed poorly differentiated adenocarcinoma composed of discohesive signet ring cells. A skin punch biopsy showed intense dyskeratosis, suprabasilar clefting, lymphocyte exocytosis, and acantholysis in the epidermis (Picture 2). Immunofluorescence examination of tissue from the perilesional skin showed weak intraepidermal positivity with C3. IgG, IgA, and IgM were negative. The findings were compatible with Paraneoplastic pemphigus.

Written informed consent for the publication of their details was obtained from the relatives of the patient.



Picture 2. Suprabasal splitting is observed in the lower epidermis, and dense dyskeratosis is observed in the upper epidermis (H&E 40x)

(a) Larger magnification shows acantholytic cells in the splitting cavity with diffuse dyskeratosis and residual basal layer cells due to suprabasal detachment (H&E, 100x) (b)

Discussion

Paraneoplastic pemphigus (PNP) is a rare autoimmune mucocutaneous disease associated with underlying neoplasia. It is typically characterized by painful mucosal erosions and dark, patchy skin eruptions. Widespread epidermal loss can lead to severe dehydration, protein loss, and an increased risk of infection. In at least 80% of patients, the underlying malignancy is lymphoproliferative diseases, primarily non-Hodgkin lymphomas. Despite its association with many solid organ malignancies, only 4 cases of gastric cancer have been published to date [2, 3]

At first glance, the lesions on the trunk suggested Leser-Trelat sign, which is considered to be a fairly rare paraneoplastic cutaneous marker of internal malignancy (gastric adenocarcinoma being the overall most common malignancy, followed by breast cancer and lymphoproliferative disorders/lymphoma), with the hallmark finding being an abrupt eruption of multiple seborrheic keratoses [4]. However, the biopsy was not taken from these seborrheic keratoses lesions; PNP turned out to be a more accurate diagnosis.

Unfortunately, there is no current diagnostic guide or treatment protocol for PNP [5]. The treatment of patients with PNP consists of suppressing of the disease manifestations and managing patient symptoms. Treatment of the underlying malignancy is beneficial in some cases. We consulted our patient with the oncology department for the treatment of gastric adenocarcinoma.

It should be kept in mind that some malignancies may trigger cutaneous manifestations. Underlying malignancies should not be forgotten in patients with rapidly developing skin lesions with constitutional symptoms.

Conflict of interest: No conflict of interest was declared by the authors.

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Authors' contributions to the article

R.I. constructed the main idea and hypothesis of the study. O.A.U. developed the theory and edited the material and method section. B.K. and M.O. have done the evaluation of the data in the results section. Discussion section of the article written by R.I. and O.A.U.

D.A.O. reviewed, corrected and approved. In addition, all authors discussed the entire study and approved the final version.