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BAZEX SYNDROME - A CASE REPORT

ZESPÓŁ BAZEXA – OPIS PRZYPADKU

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N Dermatol Online. 2011; 2(1): 18-20

Abstract

Acrokeratosis paraneoplastica of Bazex is a rare but distinctive paraneoplastic dermatoses. This condition is a cutaneous marker of supra-diaphragmatic neoplasia. This syndrome is important because the cutaneous findings precede the onset of symptoms referable to the underlying neoplasm by several months in most of the cases.

A 42 year old male who had scaly hyperkeratotic, papulosquamous lesions on face and dorsa of hands and feet with a suspicion of an underlying gastric neoplasm and a diagnosis of Bazex syndrome is being reported in view of the rarity of this condition.

Streszczenie

Zespół Bazexa jest rzadką, ale charakterystyczną dermatozą paraneoplastyczną. Schorzenie jest skórnym markerem nowotworów. Zespół ten jest ważny, ponieważ zmiany na skórze poprzedzają wystąpienie objawów nowotworów narządów wewnętrznych nawet na kilka miesięcy (w większości przypadków).

Przedstawiamy 42-letniego mężczyznę z łuszczącymi, hiperkeratotycznymi, grudkowo-łuszczącymi zmianami na twarzy oraz grzbiecie rąk i stóp z podejrzeniem nowotworu żołądka, u którego rozpoznano zespół Bazexa w związku z rzadkością występowania tej choroby.

Key words: Acrokeratosis, paraneoplastic, dermatoses
Słowa klucze: Acrokeratosis, paraneoplastyczny, dermatozy

Introduction

Bazex syndrome was first described by Gougerot and Rupp in 1922 and was named after Bazex. It is a rare syndrome, much commoner in males than females. It is associated with malignancies of upper aero-digestive tract such as malignancy of pharynx, larynx, esophagus, lung, stomach, but other types of carcinomas have also been described. It appears also with cervical and mediastinal lymphatic metastasis from a distant occult neoplasm.

Bazex syndrome is characterized by symmetrical psoriasiform lesions on acral parts of the body, palms, soles, ear lobes and nose.

Associated features of Acrokeratosis Paraneoplastica of Bazex include pruritus, vesiculation, sterile paronychia, hyperpigmentation, hypopigmentation, bullous lesions and carpel tunnel syndrome.

Identification of this paraneoplastic syndrome may enhance the earlier diagnosis of the associated tumor and thus may be important in curative treatment [1-4].

Case Report

A 42 year old male, mechanic by occupation, normotensive, non-diabetic reported to the outpatient department of Dermatology, SMHS hospital (associated teaching hospital of Govt. Medical College, Srinagar) with a two weeks history of asymptomatic, scaly eruption which started on the dorsal aspect of hands and feet and had subsequently spread to the nose, cheeks, ear lobes and forehead. The patient did not have any subjective complaints. There was no history of blistering over these areas prior to the development of these lesions.

The patient had a significant past history of upper gastrointestinal tract bleeding 2 years back for which he was managed conservatively. At that time eosophagogastroduodenoscopy (EGD) with biopsy was done which revealed a benign Gastric ulcer.

Except for pallor, the patient's general physical examination was normal. Systemic examination was also normal. Cutaneous examination revealed well-defined scaly papules and plaques distributed symmetrically over the dorsa of hands and feet, face, forehead, and earlobes. These lesions had a peculiar violaceous hue at

the periphery. There was a diffuse dark brown hyperpigmentation of malar area of face, forehead, dorsa of hands and feet. Palms and soles were spared (fig1-6). Mucosae and nail examination was also normal.
Skin biopsy for histopathological examination was taken from one of the lesions which revealed hyperkeratotic, acanthotic lining epidermis with prominent basal pigment layer and dermal perivascular inflammatory infiltrate.

Except for microcytic hypochromic anemia, all other blood tests were normal. Stool examination for occult blood was positive. Chest radiographs and electrocardiogram was normal. Ultrasonography of the abdomen revealed a 4.9#3.7 cm peripancreatic mass (suggestive of metastasis). Computed tomography of the abdomen revealed a circumferential Pyloric thickening with Coeliac lymph node enlargement (fig. 7). Esophagogastroduodenoscopy (EGD) revealed an ulceronodular lesion in prepyloric region suggestive of an antral malignancy. Biopsy taken from this lesion revealed an adenocarcinom, Intestinal type with superadded candidiasis (fig. 8).

In view of the constellation of clinical, laboratory, radiological and histopatological findings, a diagnosis of Bazex's syndrome was entertained.

Figure 1. Bazex syndrome

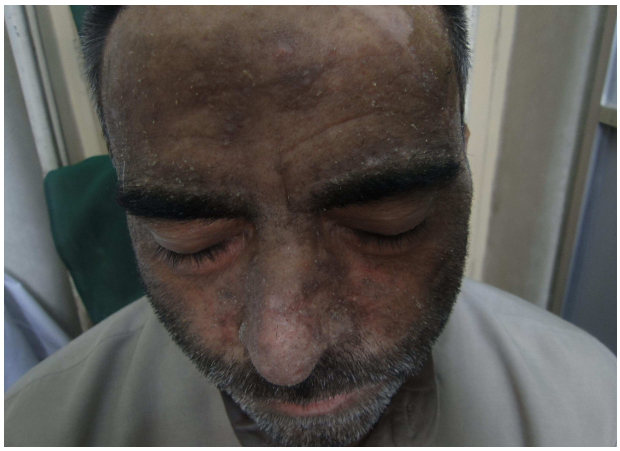

Figure 2. Bazex syndrome

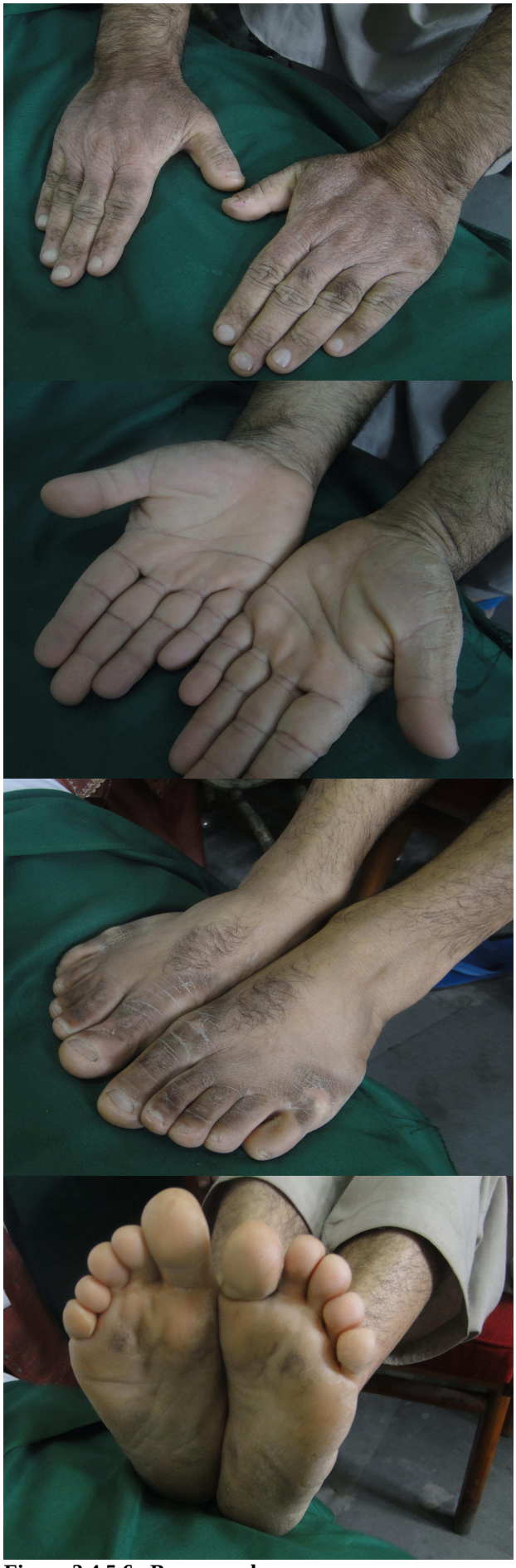

Figure 3,4,5,6. Bazex syndrome

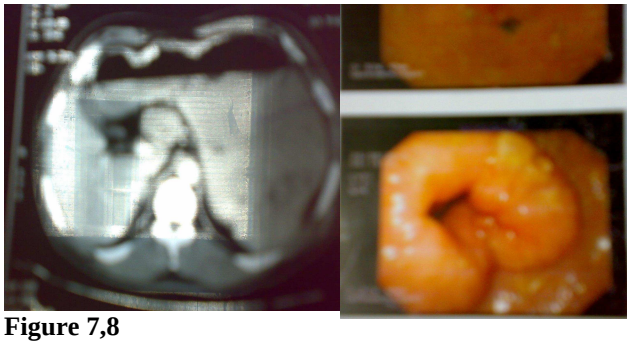

Figure 7,8

Discussion

Acrokeratosis paraneoplastica was first described by Gougerot and Rupp in 1922 and was named after Bazex. This is a rare but distinctive paraneoplastic dermatoses associated with internal malignancies, most often squamous cell carcinoma of aerodigestive tract (pharynx, larynx, esophagus, stomach), but, other types of carcinomas have also been described (lung, colon, prostate)[1-4].

Clinically, the syndrome evolves in three stages. The first stage is characterized by erythema and psoriasiform scaling on the fingers and toes which soon spreads to the bridge of the nose and the helices of the ears. Nail changes are frequent. In the second stage, a violaceous keratoderma of palms and soles develops and the facial lesions spread to the pinnae and cheeks. In the third stage, the eruption extends locally and begins to involve the legs, knees, thighs, arms, trunk and scalp [1,2].

The dermatoses may precede the diagnosis of neoplasm in about 65-70% of the patients. Cutaneous manifestations follow the diagnosis of neoplasm in 10-15% patients. There is a simultaneous onset of skin lesions and diagnosis of neoplasm in 15-25% of the cases.

The histopathological changes in the affected skin are non-specific. A mild degree of acanthosis with hyperkeratosis, parakeratosis and a dermal lymphocytic infiltration is seen.

The pathogenesis of this syndrome is uncertain. Proposed mechanisms include cross reactivity between skin and tumor antigens, the action of tumor-producing growth factors and even zinc deficiency.

The skin lesions either improve significantly or resolve completely when the underlying neoplasm is treated in 90-95% patients, whereas they remain unchanged in the setting of persistent disease. The reappearance of skin lesions signals a recurrence of the tumor. Improvement has been reported with the use of topical and systemic retinoids, topical and oral corticosteroids, salicylic acid, topical vitamin-D analogues, PUVA therapy and zinc [5-9].

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