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Caso Clínico

Síndrome de Bazex: descrição de um caso clínico associado a carcinoma epidermoide de hipofaringe

Bazex Syndrome: report of a case associated with a primary squamous cell carcinoma of the hypopharynx

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Resumo

Síndromes paraneoplásicas associadas ao cancro da cabeça e pescoço são raras. A síndrome de Bazex (BS), também conhecida como acroqueratose paraneoplástica, é uma manifestação dermatológica rara, caracterizada por placas psoriasiformes hiperceratósicas escamosas em partes acrais do corpo (hélices, nariz, mãos e pés) e, em fases posteriores, propagação para os membros e tronco. Esta síndrome é um marcador distinto para diferentes neoplásias, predominantemente para o carcinoma de células escamosas do trato aerodigestivo superior. Descrevemos um caso clínico de um homem de 60 anos de idade, com carcinoma epidermóide da hipofaringe e metastização à distância, com manifestações cutâneas exuberantes, evidenciando as lesões cutâneas típicas dessa síndrome rara. Perante a suspeita de SB, deve ser realizada uma biópsia cutânea seguida de uma avaliação completa do trato aerodigestivo superior. A identificação de uma síndrome paraneoplásica pode melhorar o diagnóstico precoce do tumor associado e, assim, possibilitar o tratamento curativo.

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Palavras chave: Síndrome de Bazex, Acroqueratosis paraneoplasica, Síndrome paraneoplásica, Carcinoma de células escamosas, Dermatose psoriasiforme

Abstract

Paraneoplastic syndromes associated with head and neck cancer are rare. Bazex syndrome (BS), also known as acrokeratosis paraneoplastica, is a rare dermatologic manifestation, characterized by scaly hyperkeratotic psoriasiform plaques on acral parts of the body (helices, nose, hands and feet) and, in later stages, propagation to the limbs and trunk. This syndrome is a distinct marker for different neoplastic conditions, predominantly squamous cell carcinoma of the upper aerodigestive tract. We report the case of a 60-year-old male patient with a primary squamous cell carcinoma of the hypopharynx with distant metastization and exuberant clinical manifestations, showing the typical cutaneous lesions of this rare syndrome. Therefore, when suspecting BS, a skin biopsy followed by a complete evaluation of the upper aerodigestive tract should be performed. Identification of a paraneoplastic syndrome may enhance the earlier diagnosis of the associated tumour and may thus enable curative treatment.

Keywords: Bazex syndrome, Acrokeratosis paraneoplastica, Paraneoplastic syndrome, Squamous cell carcinoma, Psoriasiform dermatosis

Introduction

Paraneoplastic syndromes are defined as clinical, hormonal, haematological, neurological or dermatological disorders related to a neoplasm, but not directly associated with invasion by the primary tumour or metastases[1]. They occur in up to 8% of cancer patients[1], and their association with tumours of the head and neck is rare.

Bazex syndrome (BS), also known paraneoplastic acrokeratosis, is a rare example of paraneoplastic cutaneous disease[2] that has been associated with either a primary malignant neoplasm of the upper aerodigestive tract or metastatic cancer to the lymph nodes of the neck[3,4].

BS was first reported by Gougerot and Rupp[5], in 1922, however in 1965 the association was firmly established by André Bazex, who was the first to associate metastatic pyriform sinus cancer[6-9] with an unusual distribution of psoriasis-like cutaneous lesions involving not only the palms and feet but also other body parts such as cheeks, nose and ears[8]. Since then, atypical psoriasiform skin lesions serve as cutaneous markers for an extracutaneous malignant tumour[8].

Case Report

A 60-year-old man was referred to our hospital with a history of anorexia, weight loss (8 kg in three months) and progressive dysphagia (3 months). He had no significant past medical history, except a history of alcoholism and a 60-pack-a-year history of cigarette smoking. Six months before presentation, he noted the acute onset of scaling on the soles of his feet, fingers and toes as well as dystrophy of the finger-

nails and toenails. In addition, areas of hyperpigmentation had begun to develop on the nose and ears, but without associated pruritus.

On physical examination, skin lesions were symmetrical, hyperkeratotic and hyperchromatic, involving the distal part of the hands and feet (figure 1 and 2), as well as the nasal dorsum and the helices of both ears (figure 3 and 4). The distal phalanges were thickened in combination with thick yellowish nails and subungual longitudinal striae, demonstrating hyperkeratosis and onychodystrophy (figure 1). This patient did not have a history of palmoplantar keratosis and the cultural and direct examination for fungi was negative. No drug was suspected of causing adverse skin reactions.

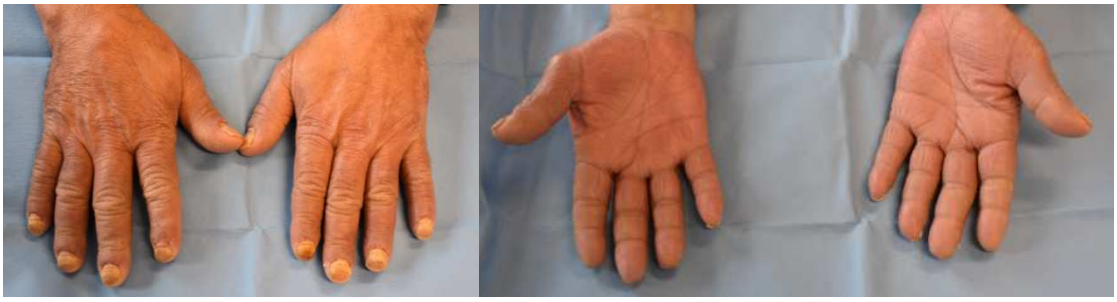


Figure 1: Hyperpigmentation, scaling, erythema, and nail changes on dorsum of hands. Note the yellowish coloration, hyperkeratosis and distal onycholysis that affects all the fingernails (left); Pronounced hyperpigmentation, scaling, and erythema on palmar hands (right).

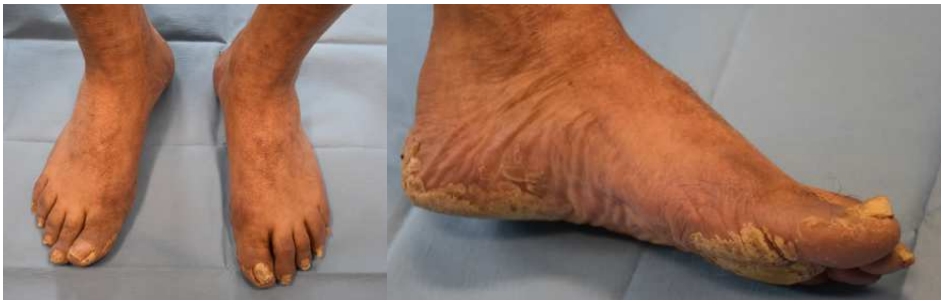


Figure 2: Symmetric hyperkeratotic plaques on the distal part of the feet (left); Scaling and hyperpigmentation of the plantar skin with relative sparing of the central portion (right).

Figure 3: Hyperpigmentation and hyperkeratosis on the tip of the nose.

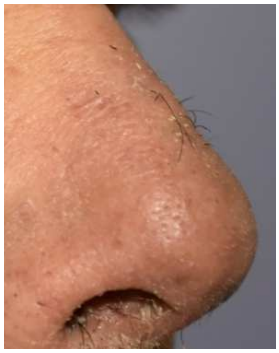
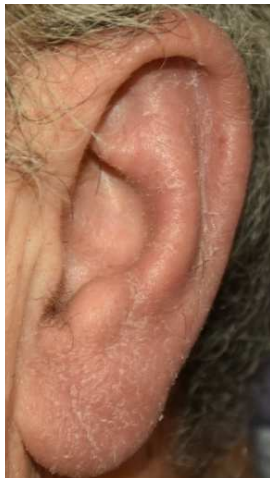


Figure 4. Violaceous desquamation of the rigth ear.



Due to the patient's history of alcohol and tabaco abuse associated with his recent weight loss and dysphagia, a Laryngoscopy was performed which revealed an ulcerated lesion in the postcricoid region, with extension to arytenoid and arytenoepiglottic folds and bilateral adenopathic conglomerate in level II and III.
The Computed Tomography (neck, thorax, abdomen and pelvic) revealed a neoformation originating from the proximal hypopharynx (postcricoid region with supraglottic and cervical oesophagus invasion). Bilateral tumoral lymphatic spread, level IIB and IIA on the left side, and level IIA and III on the right side. Bulky liver, with numerous hypodense lesions, with many small areas of parenchyma spared by secondary lesions. Secondary lesion at the upper polar end of the spleen. Adrenal metastases. Peritoneal implant near the diaphragm and para-esophagogastric adenopathy's.
Biopsy was performed on the skin lesions and hypopharynx, revealing discrete irregular acanthosis and focal parakeratosis of the epidermis and SCC of the Hypopharynx.
Routine serum chemical analyses and urinalysis were irrelevant.
The SCC of the hypopharynx was staged as IVc (T4aN2cM1) and his skin condition was diagnosed as a paraneoplastic condition known as Bazex syndrome, based on the new onset of typical skin findings associated with the tumour. Given that the tumour was not amenable to surgical treatment the patient was proposed for Radiotherapy associated with chemotherapy, unfortunately the patient passed away before starting treatment with septic shock-superinfection of hepatic/peritoneal metastasis.

Discussion

BS is a rare paraneoplastic cutaneous disorder with a peak incidence in the 60-year old age group and a male predominance of about three-to-one[8]. Bolognia *et al* found that in 67% of patients, cutaneous lesions preceded the diagnosis of an underlying malignancy, while in 18% of the cases, the lesions appear simultaneously and in 15% the lesions developed after the neoplasm was discovered[9,10]. In 113 patients diagnosed with Bazex syndrome, neoplasms of the oropharynx and larynx (48.6%) were predominant, followed by lungs (17.7%), unknown location (16%), oesophagus (10.6%), prostate (1.7%), and isolated cases in the uterus, vulva, stomach, liver, thymus and bone marrow [9].

The skin changes can be divided into three stages[4,11]. The first stage is characterized by poorly defined erythematous, violaceous and scaly lesions on the acral regions, with tenderness but no pruritus in most cases[7]. At this stage, there is no evidence of the underling tumour[4,12]. In the second stage, the lesions worsen with hyperkeratosis of the palms and soles of the feet, and an underlying malignancy is frequently identified due to primary tumour symptoms [4]. In the last stage, if the tumour is not treated, the skin lesions progress to the trunk, legs and arms[4,13].

The pathogenesis of BS is unknown. However immunological mechanisms are hypothesized [7,10]. The diagnosis is clinical, given that the histological findings are unspecific [9,13]. A differential diagnosis should be made with other erythematous-squamous dermatoses[13,14]. BS should be suspected upon involvement of the tip of the nose or helices associated with considerable nail changes, like dystrophy, discolouration and onycholysis and when an acral dermatosis has failed to respond to correct treatment[14].

Upon suspicion of a patient with BS and no previous underlying neoplasm, a complete examination must be performed, including an otolaryngologic examination[9]. Like other paraneoplastic cutaneous, the only effective treatment is to treat the primary tumour[9]. The skin lesions significantly improve when the underlying neoplasm is treated, although[9,10] nail changes and residual hyperpigmentation may persist. The recurrence of the skin lesions suggests recurrence or oncologic progression[,9,10,14].

Identification of a paraneoplastic syndrome may enhance the earlier diagnosis of the associated tumour and may thus enable curative treatment. So, recognition of these hallmark signs is of the outmost importance to perform a prompt diagnosis of an underlying malignancy.

Conflicts of interest

All authors report no conflict of interest.

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