Is it just a psoriasiform dermatitis?

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Abstract

Bazex syndrome (BS) is a rare paraneoplastic syndrome most frequently associated with squamous cell carcinomas of the upper aerodigestive tract and other tumours. Characteristically, cutaneous lesions precede the diagnosis of malignancy. We report a 72-year-old patient with 1-year history of acral dermatitis. The diagnosis of BS was based on the presence of psoriasiform acral dermatitis and the evidence of two simultaneous tumors (prostate adenocarcinoma and undifferentiated carcinoma of the submandibular gland). It is important to have this syndrome in mind since cutaneous features usually precede an underlying neoplasm.

Keywords: Bazex syndrome, underlying malignancy, paraneoplastic, acral dermatosis

Introduction: Bazex syndrome (BS), also known as acrokeratosis paraneoplastica, is an unusual cutaneous eruption associated with underlying malignancy, most frequently carcinomas of the upper aerodigestive tract [1]. This entity is more common in white males older than 40 years [2]. The pathogenesis still remains unknown, although some theories have been proposed, such as cross-reactivity between tumor and skin antigens, an immunological mechanism or secretion of growth factors by the tumor cells. Zinc and vitamin A deficiency might play a role [1, 2].

Case Synopsis: A 72-year-old male patient presented with asymptomatic lesions involving his nose, fingers, and nails of one year duration. His medical history

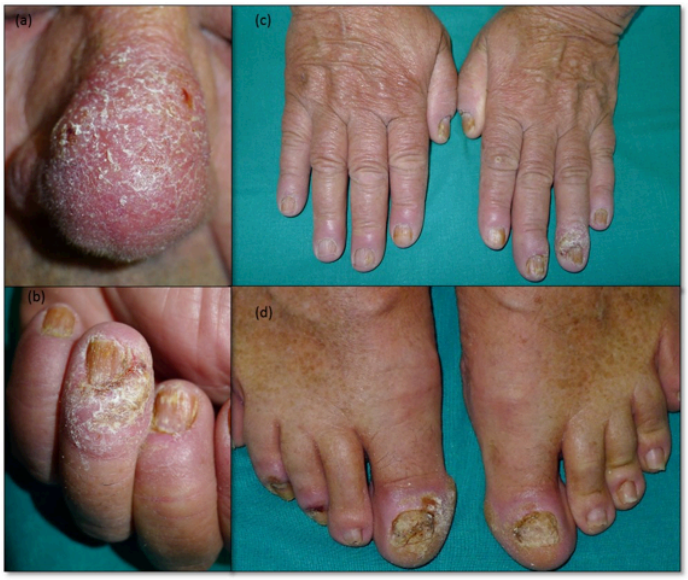

Figure 1. *Clinical images of the patient's eruption, demonstrating A) erythematous scaly plaque and hyperkeratosis on the tip of the nose; B) typical nail and periungual changes; C), D) hyperpigmentation, scaling and erythema on the hands and feet.*

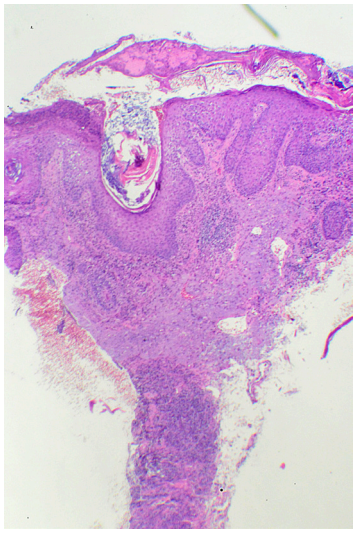

Figure 2. *Parakeratotic hyperkeratosis and acanthosis, associated with lymphohistiocytic inflammatory infiltrate (H&E, 10%).*

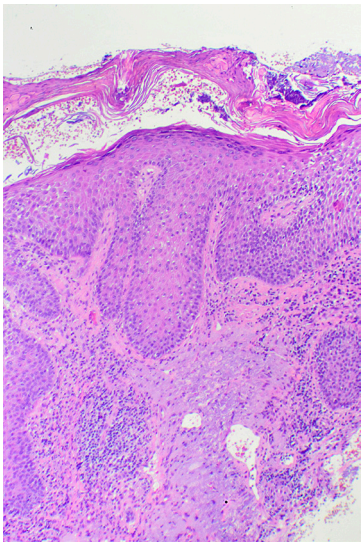

Figure 3. *A detail of skin biopsy showing hyperkeratosis, acanthosis and enlargement of rete ridges with moderate dermal lymphohistiocytic infiltrate (H&E, 40%).*

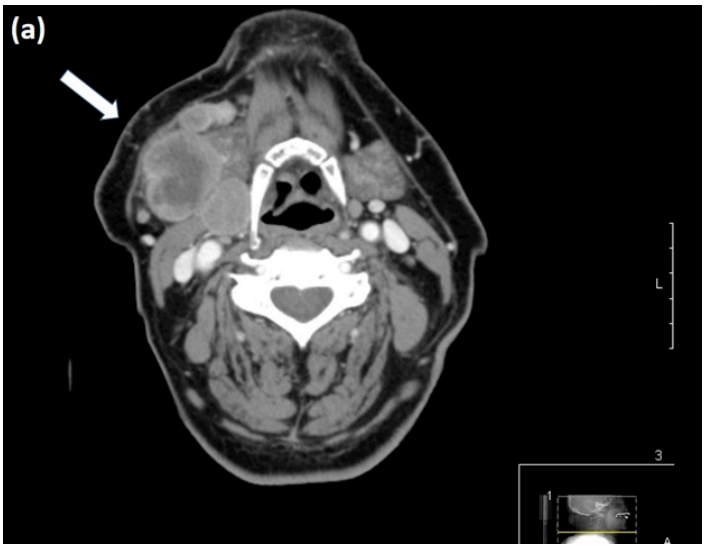

Figure 4. *Computed tomography of the neck demonstrated findings consistent with a submandibular gland tumor. (Arrow highlighting the finding).*

included cigarette smoking and hypertension. Moreover, he had been recently diagnosed with a prostate adenocarcinoma, Gleason score 7, and he was undergoing radiotherapy and hormone therapy. He had no prior history of psoriasis or other skin diseases. The family history was unremarkable. He had been treated with topical antibiotics, antifungals, and topical corticosteroids without improvement.

Dermatological examination showed sharply demarcated psoriasiform plaques on the tip of his nose. His fingernails and some toenails were affected symmetrically with yellow discoloration, ridging, and nail dystrophy. Erythematous hyperkeratotic plaques involving the periungual folds of various fingers and toes were also observed (**Figure 1**). General physical examination showed a mass located in the right submandibular region, which had not been noticed by the patient. A complete blood test only revealed an elevation of the serum prostate-specific antigen (PSA) level. Fungal cultures were negative.

A biopsy specimen was taken from his nose and the histopathological examination displayed parakeratotic hyperkeratosis and acanthosis, associated with a perivascular lymphohistiocytic inflammatory infiltrate (Figures 2, 3). Computed tomography of the neck demonstrated cervical adenopathies and a tumor located in the submandibular gland (**Figure 4**). Histopathological examination of this tumor showed undifferentiated carcinoma of the submandibular gland (classification:T3N2b M0). Morphological and immunohistochemical findings ruled out a metastatic prostate adenocarcinoma (immunohistochemical stains were negative for cytokeratin [CK] 20, CK 7, prostate-specific antigen [PSA], and prostate acid phosphatase [PAP]); stains were positive for vimentin, CK 5/6 and p63).

The diagnosis of acrokeratosis paraneoplastica of Bazex was made based on the psoriasiform acral dermatitis and the evidence of two simultaneous tumors (prostate adenocarcinoma and undifferentiated carcinoma of the submandibular gland). Treatment with radiation therapy and chemotherapy with cisplatin was initiated. The cutaneous tumors minimally improved and the patient died 4 months later owing to metastatic dissemination.

Case Discussion

BS is a rare paraneoplastic syndrome characterized by erythematous to violet scaly lesions, which are typically symmetric, bilateral, and acral. Nails are usually involved with subungual hyperkeratosis, yellow discoloration, onycholysis, and ridging. The most commonly affected sites are the ears (79%), nails (75%), nose (63%), fingers (61%), hands (57%), and feet (50%), [1]. Bullous lesions are infrequently seen [3]. Histopathology is nonspecific, as in the present case, but it is useful to rule out other dermatoses like

psoriasis, eczema, and lupus erythematosus.

In 67% of BS cases, cutaneous findings precede detection of an underlying neoplasm, usually by several months [4]89 were male with a mean age of 60 +/- 8.5 years. Squamous cell carcinomas of the head and neck and squamous cell tumors of unknown primary with cervical lymph node metastases were the most commonly associated neoplasms, suggesting that the factor(s. Therefore, BS is an important marker for internal malignancy. Squamous cell carcinoma of the aerodigestive tract or metastatic cancer to the lymph nodes of the neck are the most commonly associated malignancies [5]. Other neoplasms have been reported include adenocarcinomas of the bladder, colon, prostate gland, lung, and breast and also Hodgkin disease [6–9]. Valdevieso et al. Valdivielso et al. [10] proposed a diagnostic algorithm for BS based on the most frequently associated tumors. Treatment of the underlying malignancy is usually followed by a symptomatic improvement. If the neoplasm is intractable, oral retinoids might control symptoms. Otherwise, the recurrence of the cutaneous eruption is often related to tumor relapse [1, 10].

Conclusion

We report a patient diagnosed with BS related to two different neoplasms (prostate gland cancer and undifferentiated carcinoma of the submandibular gland). Association of BS with prostate gland cancer has rarely been reported [6, 7, 9] and a review of the literature failed to reveal an association between carcinoma of the submandibular gland and any paraneoplastic syndromes. Recognition of the cutaneous features can lead to an earlier diagnosis of unsuspected internal malignancy or the spread or recurrence of an already diagnosed tumor.

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