Net Case

Acrokeratosis paraneoplastica (Bazex syndrome)

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ABSTRACT

Acrokeratosis paraneoplastica of Bazex is rare condition, characterized with scaly hyperkeratotic psoriasiform plaques on acral parts of body (helices, nose, and malar and acral surfaces), and in later stages propagation to the limbs and trunk.This syndrome is distinct marker for different neoplastic conditions, predominantly squamous cell carcinoma of the upper aerodigestive tract with possible cervical lymph node metastases.In this paper we present 56 years old male patient, with hyperkeratotic plaques on the skin of his palms, soles, ear lobes and apex of the nose. Detailed examination found tumorous swelling on the left side of his neck. Histopathologic examination revealed solid anaplastic metastatic tumor. Patient died before primary tumor could be found. Bazex syndrome can appear before the diagnosis of internal malignancies, and thus is important for dermatologists to recognize it in favor of early diagnosis of specific malignant process.

Key words: Acrokeratosis, Hyperkeratosis, Paraneoplastic dermatosis, Squamous cell carcinoma

INTRODUCTION

Acrokeratosis paraneoplastica (first described by Gougerot and Rupp in 1922) was named after Bazex.[1] This is an obligatory paraneoplastic dermatosis associated with different malignancies, most often squamous cell carcinoma of upper parts of respiratory and digestive system, but other types of carcinoma are also described. It appears also with cervical metastatic lymphadenopathy.[1-3] Bazex syndrome is characterized with symmetric psoriasiform lesions on acral parts of the body - palms, soles, ear lobes, and nose. Almost all described patients are white males older then 40 years, but a case of Bazex syndrome has been described in young black female patient.[4]

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

CASE REPORT

Our patient was 56 years old construction worker, who was admitted to our Clinic with symmetric hyperkeratotic skin changes on his palms, fingers, soles, and tip of his nose and on the ear lobes [Figures 1-4]. The skin of those parts of his body had a violaceous hue. Some finger and toe nails showed moderate dystrophic changes.

Skin lesions had been present for one month. He didn't have any subjective complaints, except mild itch.

On the left side of his neck, we noticed large tumorous swelling which was investigated with ultrasound and computerized tomography. Those revealed on the left side of the neck, under medial edge of sternocleidal muscle a solid mass, 50 × 30 mm, sharply demarcated, hypoechogenic, with uneven structure. Tumor was not compressible. It infiltrated left sternocleidal muscle and touched large airways, but great blood vessels were spared. Parotid and submandibular salivary gland was normal, as well as thyroid gland.

Laboratory findings were within normal range except for red blood cells (3,58T/L). The chest radiography was normal.

Biopsy of skin changes revealed hyperkeratosis with parakeratosis and marked acanthosis and

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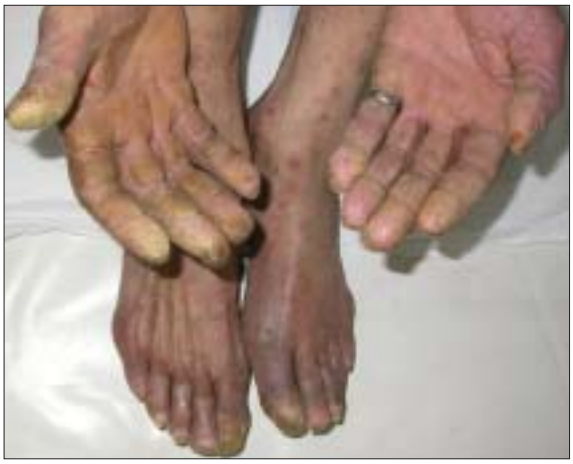

Figure 1: Violaceous scaly lesions involving fingers and toes

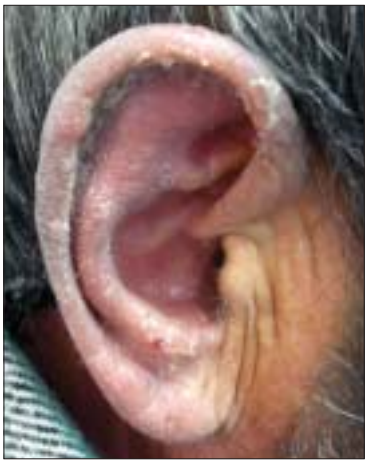

Figure 2: Violaceous scaly lesions over the pinna

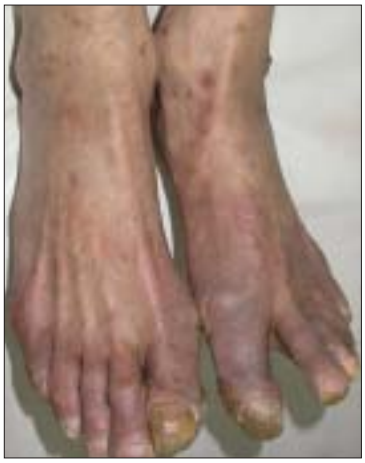

Figure 3: Similar lesions over the toes with nail dystrophy

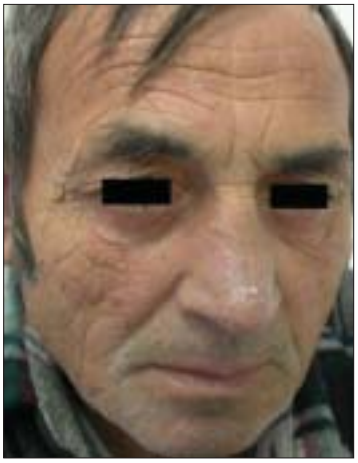

Figure 4: Violaceous scaly tip of the nose

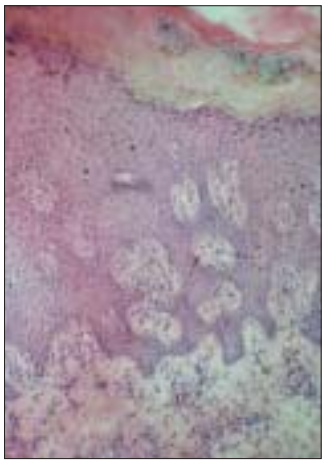

Figure 5: Photomicrograph showing hyperkeratosis with parakeratosis and marked acanthosis and papillomatosis (H & E, x100)

papillomatosis, sparse lymphocytic infiltrate and papillary edema [Figure 5].

Our patient was transferred to Department of maxillofacial surgery for tumor resection. Histopathologic examination showed solid anaplastic metastatic carcinoma with marked desmoplastic reaction. Pathologists mentioned that primary tumor should be searched in thyroid gland, lungs or surrounding tissues. Unfortunately, our patient died before primary tumor was found.

DISCUSSION

Paraneoplastic dermatoses are conditions that may be associated with an internal malignancy. The mucosal or skin lesions may precede, be concurrent with, or follow the discovery of the related cancer. The appearing of those rare skin changes has not satisfactory explanation.

Acrokeratosis paraneoplastica (first described by Gougerot and Rupp in 1922) was named after Bazex.[1] This is obligatory paraneoplastic dermatosis associated with different malignancies, most often squamous cell carcinoma of upper parts of respiratory and digestive system, but other types of carcinoma are also described

(lungs, prostate, colon, breast).[5-10]

According to the initial 1965 description by Bazex and Griffiths, the syndrome consists of 3 stages: The first stage is characterized by a symmetrical scaly erythematous pruriginous rash in the distal parts of the fingers and toes, followed by involvement of the helices and nose. This is followed by hyperkeratosis of the periungual region and dystrophy and onycholysis of the nails. At this stage the neoplastic disease is usually asymptomatic, as skin manifestations usually precede cancer symptoms by 2 to 6 months. In the second and third stages, there is lymph node involvement with progression of the skin lesions, with centripetal spread toward the rest of the extremities and the trunk.[1-3,5]

If we correlate the temporal relationship between onset of the skin lesions and diagnosis of neoplasm, the psoriasiform lesions are known to precede the diagnosis of neoplasm in 65-70 percent of the patients.[3] Cutaneous manifestations follow the diagnosis of neoplasm in 10-15 percent of patients. There is a simultaneous onset of skin lesions and diagnosis of tumor in 15-25 percent of the cases. Skin lesions either improve significantly or resolve completely when the underlying neoplasm is treated in 90-95 percent of the patients whereas they remain unchanged in the setting of persistent disease.[3,5] The reappearance of skin lesions signal a recurrence of the tumor.[3,5,7,8]

Spread to cervical lymph nodes is common at the time of presentation and in many cases (as our patient) the primary site is never discovered.

The histopathological changes in the affected skin are nonspecific. A mild degree of acanthosis is often seen with hyperkeratosis and patchy parakeratosis.[10] There is usually a lymphocytic infiltrate in the upper dermis. Histopathological findings in our case were similar. Less frequently reported changes are eosinophilic hyalinization of prickle cells with scattered vacuolar degeneration. Direct immunofluorescent assay may show local deposits of immunoglobulins, C3 components, or fibrin in the basement membrane.[10]

The pathogenesis of the syndrome is unclear. Several authors suggest an autoimmune mechanism in which cancer cells and epidermal cells may share a common antigen. Other hypotheses implicate stimulated expression of transforming growth factor α by the tumor as the pathogenic mechanism in the skin.[2,6-9]

This skin disease does not respond to steroid or keratolytic therapy, Skin lesions either improve significantly or resolve completely when the underlying neoplasm is treated whereas they remain unchanged in the setting of persistent disease. Some cases show partial improvement with retinoid (etretinate) therapy.[3]

The importance of this paraneoplastic dermatosis lays in the fact that, if the diagnosis has been confirmed an appropriate systematic evaluation for an underlying neoplasm should be performed in a patient without known cancer, and a search for possible recurrent or metastatic disease should be undertaken in a patient with a known history of malignancy.

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