Bazex's syndrome with myelodysplasia

K K V N Somaratne[1], B de Silva[2], P Karunasekara[3]

Sri Lanka Journal of Dermatology, 2010, **14**, 26-27

Abstract

Paraneoplastic skin manifestations are skin reactions to internal malignancies. They may appear several years before the symptoms of the malignancy.

Bazex's syndrome is a paraneoplastic syndrome characterized by psoriasiform skin lesions with an underlying solid organ or a haematological malignancy.

Here we report a case of Bazex's syndrome associated with myelodysplasia.

Introduction

Bazex's syndrome (acrokeratosis paraneoplastica) is a rare paraneoplastic syndrome. The typical cutaneous features include symmetrical psoriasiform scaly lesions distributed mainly in the acral areas and the face. Almost always the lesions are noted on the nose and the pinna of ears.

Squamous cell carcinomas of upper respiratory or upper gastro intestinal tract are the commonest associations[1]. Carcinoma of bladder, breast, bronchus uterus and haematological malignancies such as multiple myeloma, lymphoma, leukaemia and myelodysplasia are rare associations.

Cutaneous features develop gradually. Initially there are violaceous eruptions and scaling of acral areas. The eruptions become more hyperkeratotic with keratoderma of palms and soles. Later lesions become generalized. Nail dystrophy and paronychia are often present.

Histology of the skin is not diagnostic[2]. Main features are hyperkeratosis, focal parakeratosis and acan-thosis. Variable epidermal changes such as focal spongiosis with associated exocytosis of lymphocytes, basal cell vacuolar changes and scattered degenerate keratinocytes have also been reported. Pathogenesis is not well understood. Transforming growth factor - alpha is thought to play a role[2].

Skin lesions resolve with the treatment of the associated malignancy. Retinols are useful for the cutaneous lesions when the malignancy is untreatable[1].

Case history

A 48-year old female was admitted with psoriasiform skin lesions which did not respond to usual treatment for psoriasis during the past 5 years.

Skin lesions were symmetrically distributed mainly over the hands, feet, pinna of ears and tip of the nose (Figures 1 & 2). Violaceous plaques were covered with psoriasiform scales. There were fine scales in the scalp and the nails were thickened. The fingers and toes were oedematous (Figure 3).

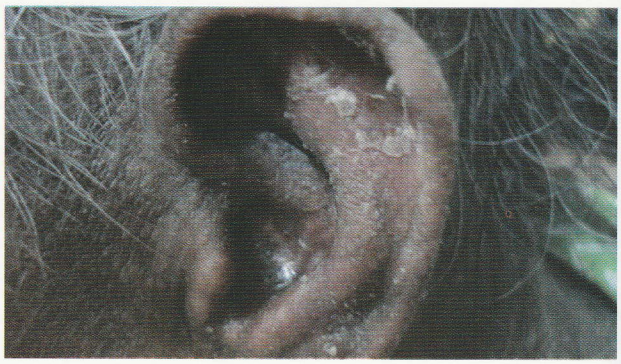

Figure 1. *Scailang of ear lob.*

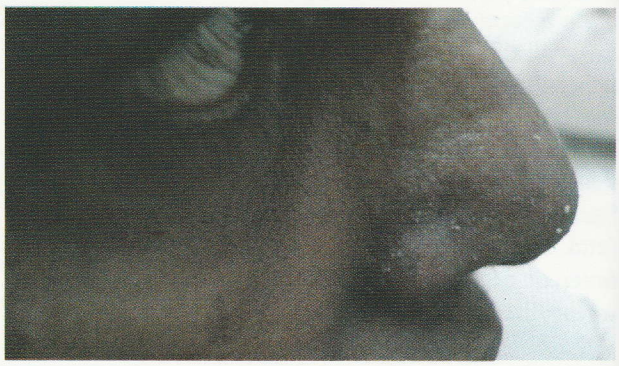

Figure 2. *Involvement of tip of the nose.*

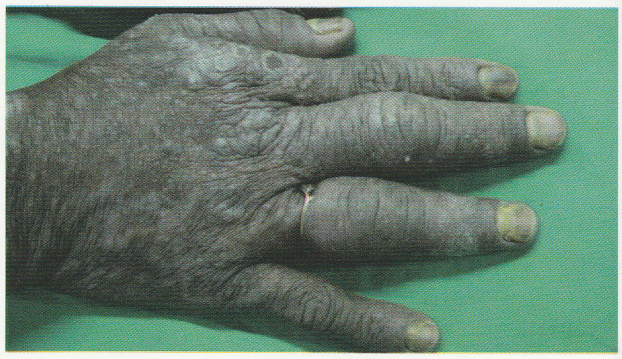

Figure 2. *Swelling and scaling of fingers.*

[1]*Senior Registrar in Dermatology,* [2]*Registrar in Dermatology,* [3]*Consultant Dermatologist, Colombo South Teaching Hospital, Kalubowila, Sri Lanka.*

Laboratory investigations were as follows. Haemoglobin was 9.9 g/dl. There was leucopenia with lymphopenia and thrombocytopenia (she was not on methotrexate or any other cytotoxic drug). Blood picture showed few macrocytes and monocyte predominance. Bone marrow aspiration and trephine biopsy results were compatible with myelodysplasia. Upper gastro intestinal endoscopy and CT scan of abdomen and pelvis were normal. Histology of skin showed hyperkeratosis, focal parakeratosis, focal acanthosis and mixed inflammatory infiltrate.

Discussion

Bazex's syndrome is a diagnosis which is easily missed. Clinical suspicion is important. Since the skin lesions usually preceded the onset of malignancy patients should be followed up for possible underlying malignancy. Our patient had cutaneous features compatible with Bazex's syndrome. Involvement of hands, feet, tip of the nose and pinna of ears was prominent. There were several histological findings which support the diagnosis. Psoriasis was the main differential diagnosis.

Methotrexate can cause pancytopenia. However our patient was not on methotrexate during the past 4 years. Before that she was given a total dose around 200 mg, but the blood counts were normal at that time.

Considering the cutaneous features and haematological findings we diagnosed Bazex's syndrome as the presenting feature of myelodysplasia in our patient.

References

1. Graham RM, Cox NH. Systemic diseases and skin. Rook's Textbook of Dermatology. 7th edition, Blackwell Science Publications 2004; **59**: 21.
2. Weedon D. Skin Pathology. Churchill Livingston Publications 2002; 575-6.
3. Keyn CE, Cheong JEL, Bell HK. Cutaneous manifestations of internal malignancy and management. *Journal of American Academy of Dermatology* 1993; **28**: 147-64.
4. Nguyen VQ. Paraneoplastic diseases. E medicine, 18 sep. 2008.