ACROKERATOSIS PARANEOPLASTICA

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A 60-year-old man with squamous cell carcinoma of hypopharynx and acrokeratosis paraneoplastica (Bazex syndrome) is reported.

Key Words: Squamous cell carcinoma, Acrokeratosis paraneoplastica, Bazex syndrome

Introduction

There are many paraneoplastic diseases of the skin. In 1965 Bazex et al described acrokeratosis paraneoplastica (AP) in a patient having carcinoma of the pyriform fossa. Since then AP has been identified as a reliable marker of internal malignancy especially of upper respiratory tract and upper gastrointestinal tract. Here we report a patient with AP, associated with moderately differentiated squamous cell carcinoma of hypopharyngeal region.

Case Report

A 60-year-old man was seen for progressive thickening and fissuring of palms and soles and pain of finger tips of one month duration. He had no skin disease in the past. He used to smoke 10-12 beedies per day for the last 45 years. On examination, he was pale and had nontender, hard, discrete, enlarged lymph nodes with restricted mobility on right cervical region. Both palms and soles were uniformly thickened and violaceous with scaling and fissures. Thickening and scaling were extending to anterior surface of wrists and dorsum of feet. Dorsal surface of distal half of all the fingers was erythematous. oedematous and tender. Nail folds were tender. There was clubbing of fingers.

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Finger nails had longitudinal bluish bands and subungual hyperkeratosis.

Routine tests of urine and blood, LFT, RFT and blood sugar were normal. X-ray of hands and ultrasound abdomen were normal. Upper GI endoscopy revealed severe antral gastritis and a biopsy showed no evidence of malignancy. Chest X-ray revealed multiple, bilateral patchy opacities suggestive of secondary neoplastic deposits. FNAC from cervical lymph nodes showed cells metastasizing from squamous cell carcinoma. Skin biopsy of palm revealed massive hyperkeratosis, parakeratosis and focal collections of lymphocytes and histiocytes in upper dermis.

Considering the advanced stage of malignancy no specific treatment was initiated by oncologist. A review of the patient after 4 months showed that the erythema and oedema of the fingers had progressed. He had dysphagia to solid foods and loss of weight. Direct laryngoscopy revealed an ulcerated growth on right wall of hypopharynx extending to lateral glossoepiglottic folds, laryngeal surface of epiglottis, aryepiglottic fold, arytenoid and pyriform fossa on the right side. It was staged as T4 N2 M1. Biopsy showed a moderately differentiated squamous cell carcinoma.

Discussion

AP as described by Bazex and Griffiths, 2 begins as erythema and

psoriasiform scaling of the fingers and toes which progress to violaceous palmoplantar keratoderma. Nail folds are prominently affected and are tender. The affected skin appears oedematous. Discolouration, subungual hyperkeratosis and dystrophy of nails may occur. Erythema and scaling may affect the bridge of nose and pinnae and later the arms, elbows, knees, legs, trunk, axillae, face and scalp. Incomplete forms also have been reported.^{2,3}

Sudden onset and rapid progression of palmoplantar keratoderma in our elderly patient aroused surpicion of an internal malignancy. Erythema and oedema of the dorsum of fingers, tender nail folds, bluish hue of the palms and soles, discolouration of nails and subungual hyperkeratosis, with no lesions elsewhere, suggested a diagnosis of incomplete form of AP. Investigations revealed the presence of a squamous cell carcinoma with metastasis, but the primary neoplasm could not be located initially. Subsequently, he was diagnosed to have

moderately differentiated squamous cell carcinoma of the hypopharyngeal region. Bazex and Griffiths² note that the primary neoplasm is frequently undetected in the early stages of AP, even with metastasis to lymph nodes.

To the best of our knowledge, this is the first case of AP reported from India. It is hoped that with greater awareness, more cases may get reported. In patients with acquired palmoplantar keratoderma and internal malignancy, other features of AP have to be looked for.

References

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