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POIKILODERMA VASCULARE ATROPHICANS

L Padmavathy, PVS Prasad, K Prasanna, L L Rao

A 65-year-old lady presented with generalised pruritus and discolouration of skin and mucous membranes of 5 years duration. The histopathology from the cutaneous lesions revealed features suggestive of poikiloderma vasculare atrophicans (PVA). Investigations did not reveal any underlying connective tissue disease, lymphoma or systemic disease. A diagnosis of idiopathic poikiloderma vasculare atrophicans was made.

Key Words : Poikiloderma vasculare atrophicans

Introduction

Poikiloderma vasculare atrophicans is freely translatable as a mottled hyper- and hypopigmentation of the skin (poikiloderma) with interspersed telangiectases (vasculare) and areas of atrophy (atrophicans).[1] It may be generalised or localised. It may be an idiopathic disorder or a manifestation of connective tissue diseases (lupus, dermatomyositis, scleroderma), lymphomas (parapsoriasis en plaques and mycosis fungoides) and geno-dermatoses (Rothmund-Thomson syndrome, hereditary sclerosing polkiloderma, dyskeratosis congenita).[2] Other causes include physical trauma (radiodermatitis, burns, freezing) and certain ingested substances like arsenic compounds. In a case of PVA, the spectrum of investigations should include tests for any underlying systemic disease, lymphoma or connective tissue disorder. When PVA exists with no known cause, it is relegated into an idiopathic group.

Case Report

A 65-year-old lady presented with generalised pruritus and discolouration of skin and oral mucous membrane of 5 years duration. She had undergone right radical mastectomy and received chemotherapy elsewhere 6 years ago. She was asymptomatic for a year after surgery. On examination, the skin changes consisted of diffuse areas of hypo-and hyperpigmentation and telangiectases interspersed with atrophy. The lesions were present all over the body including palms and soles (Fig. 1). The left

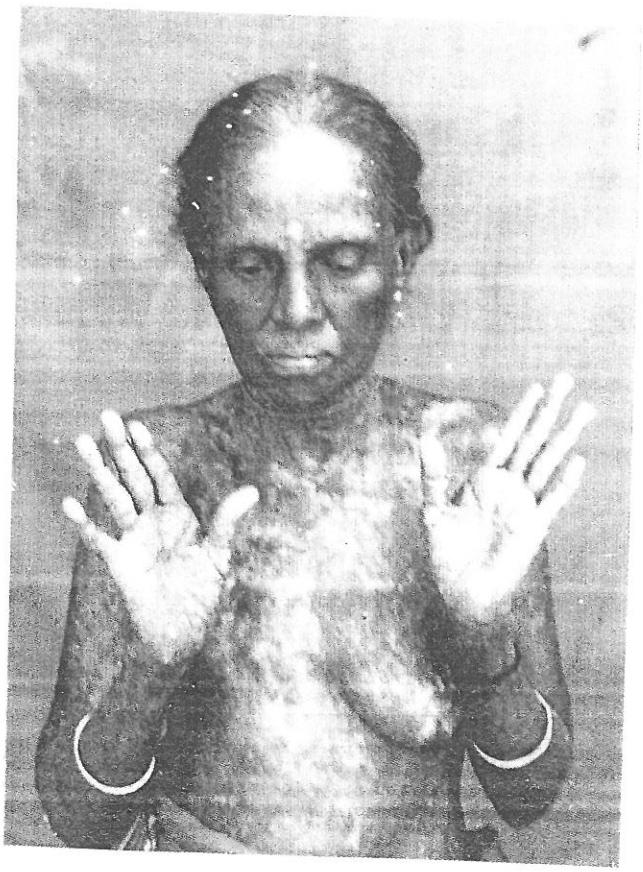

Fig. 1 Widespread poikilodermatous involvement including the palms

From the Departments of Dermatology and Pathology, Rajah Muthiah Medical College and Hospital, Annamalai University, Annamalainagar - 608 002, India.

Address correspondence to : Dr L Padmavathy

axillary and inguinal lymph nodes were mildly enlarged. Investigations: Hb 9 gm %, the peripheral smear showed a macrocytic picture and bone narrow aspiration revealed features of a megaloblastic anaemia. LE preparations were negative. Skin biopsy from the left forearm showed a hyperkeratotic, atrophic epidermis. The superficial dermis contained a band-like inflammatory infiltrate of lymphocytes and histiocytes, as well as showed pigmentary incontinence. Dilated capillary channels were present in the upper and mid dermis. Lymph node biopsy exhibited features of a nonspecific reactive follicular hyperplasia. Muscle biopsy and ultrasonogram of the abdomen were within normal limits. Other laboratory and radiological investigations were non-contributory. The skin biopsy was repeated over a period of 3 years at yearly intervals and consistently showed features of poikiloderma vasculare atrophicans without any evidence of malignancy. The patient was managed conservatively with antihistamines and bland applications. During the 3 year follow up period, she developed traumatic ulcers on the atrophic areas (Fig. 2) which were treated with rest, dressings and appropriate antibiotics.

Comments

Poikiloderma vasculare atrophicans is a condition in which the skin atrophies and may continue to do so throughout the lifespan of the patient.[1] A definite cause could not be found for the atrophying process in our patient. Hence, it is most likely an idiopathic condition. There is no evidence of local recurrence or metastatic spread from the breast lesion or any other evidence of malignancy in our patient even after a 3 year followup. However, the possibility of developing a systemic disease like a lymphoma or mycosis fungoides in the future still remains.

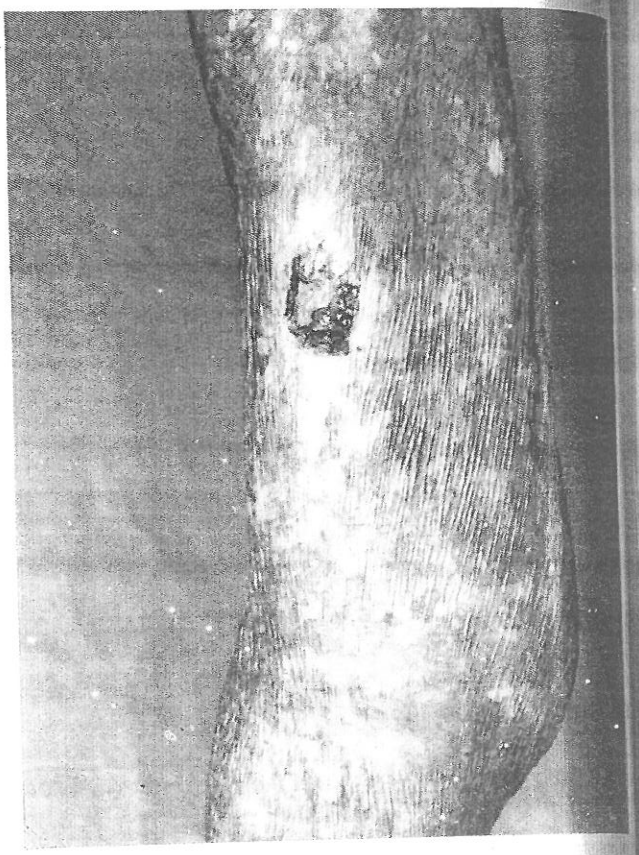

Fig. 2 Traumatic ulcers over the atrophic area

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