PSORIASIFORM SARCOIDOSIS ASSOCIATED WITH DEPIGMENTATION

R R Mittal, S P Singh, S S Gill

A 65-year-old female had psoriasiform, depigmented, angiolupoid, lupus pernio, papular and verrucous plague type of cutaneous sarcoidosis. Systemic involvement (hepatomegaly, bilateral persistent swelling of the parotid glands, pain in the hands, weakness and atrophic rhinitis of the left nasal cavity) was also present. The histopathological features of a non-caseating epithelioid cell granuloma with giant cells confirmed the diagnosis. The patient is responding favourably to prednisolone 10 mg daily.

Key Words : Epithelioid cell granuloma, Giant cell

Introduction

Sarcoidosis is a widespread, non-caseating epithelioid cell granulomatous, multisystem disease involving the lungs, heart, bones, eyes, kidneys, salivary glands, skin and reticuloendothelial system.[1] Cutaneous sarcoidosis (CS) has been classified into various types.[1] Rarely, it can present as a pruritic maculopapular rash[2] or verrucous lesion.[3] The diagnosis of CS depends on: (i) clinical evidence of multisystemic disease or radiological picture or both, (ii) histologic evidence of non-caseating granulomas, and (iii) negative cultures of sputum and tissues.[2] Hypopigmented sarcoidosis not responding to topical, intralesional or systemic steroids has been reported.[4-5] Psoriasiform sarcoidosis does occur, and sarcoidosis may be associated with psoriasis.[6] CS has been reported from India.[7]

Case Report

A 65-year-old female had a persistent bilateral asymptomatic swelling of both parotid glands, pain and scanty watery discharge from the left nasal cavity, pain in both hands, weakness and multiple asymptomatic skin lesions for 1½ years. The general physical and systemic examinations were normal except for 3 cm hepatomegaly.

Multiple, bilaterally symmetrical, well-defined, scaly erythematous plaques, 1 to 5 cm in diameter, intermingled with depigmented areas, were seen on the extensor aspects of both lower limbs (Fig.1). The Auspitz sign was negative. Brownish, soft-to-firm papules were seen on the back, and on its right upper side was a brownish verrucous, scaly plague , 2x2.5 cm in size, with purplish well-defined margins.

Three dusky violaceous, soft plaques with glistening surfaces and large dilated follicular orifices were seen on the left side of the nose (Fig.2). Ashiny, mauve, oval plaque, 2x3.5 cm in size, doughy in consistency and raised in the centre with telangiectases, was seen along the left nasolabial fold. Just above it was a similar but smaller (1 to 2 cm in diameter) plaque. Discrete and grouped yellowish-orange papules of the same consistency were seen on the left cheek, nose and in an old scar following herpes zoster on the left upper lip. The left septal mucosa was tender, and its surface was yellowish-red and moist,

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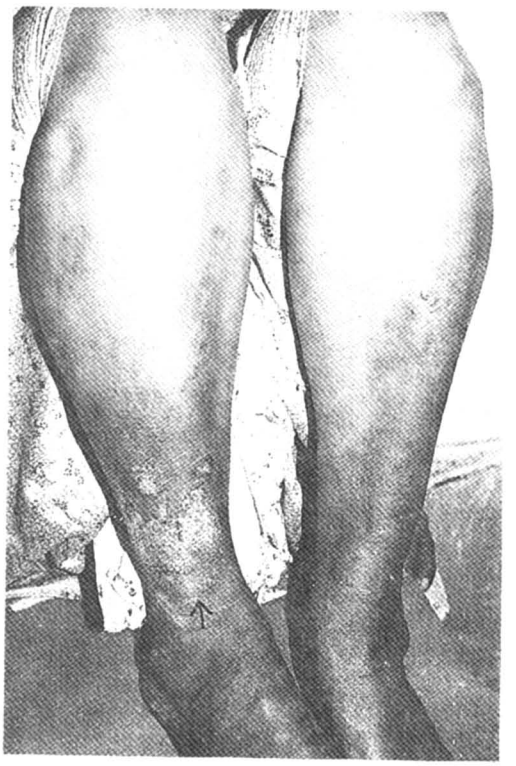

Fig. 1. Psoriasiform sarcoidosis with depigmented areas with contact depigmentation.

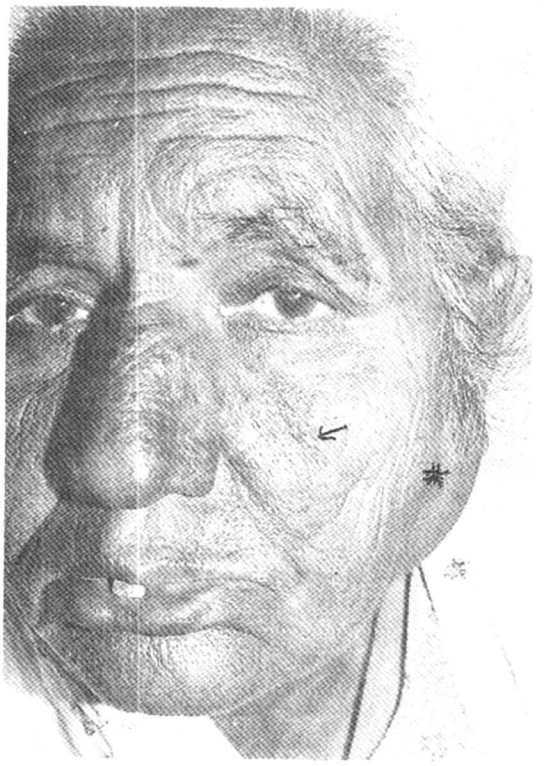

Fig. 2. Angiolupoid CS plaques along the left nasolabial fold, and just above it three lupus pernio plaques on the left side of the nose; yellow papules in the scar of herpes zoster and on the face; hypertrophy of the parotid gland.

suggestive of atrophic rhinitis.

The routine investigations were within normal limits. The ESR was 42 mm. The 24-hours urinary calcium was 700 mg. Radiographs of the hands showed cystic swellings and a lace-like pattern (Fig.3), and of the chest showed bilateral hilar lymphadenopathy, prominent bronchovascular markings and small cavities. On histopathology, a skin lesion from the face showed multiple non-caseating epithelioid cell granulomas with foreign body giant cells (Fig. 4), and from the left leg showed marked acanthosis, uniform elongation of the rete ridges, papillomatosis and mononuclear cell granulomas with giant cells in the papillae (Fig.5). The patient responded favourably to 10 mg prednisolone daily for the last 25 days. The parotid swelling almost disappeared after 2 weeks of therapy.

Discussion

The presence of varied types of CS, i.e. psoriasiform and depigmented (on the legs), angiolupoid (left nasolabial fold and cheek), lupus pernio (left side of the nose), papular CS (face), verrucous papules and plaques (back), were interesting features in the patient. The ultrastructural changes in melanocytes of CS with hypopigmented lesions are similar to, though less extreme than, those seen in other dermatoses

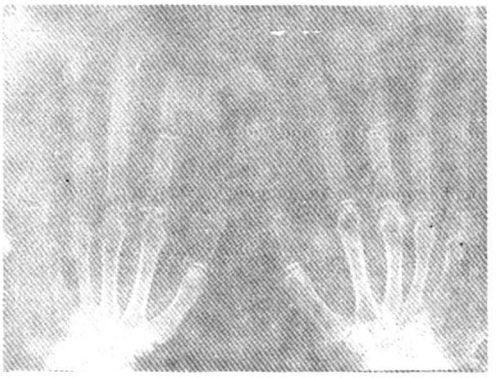

Fig. 3. X-ray of the hands showing cystic cavities with lace like pattern in the distal phalanges.

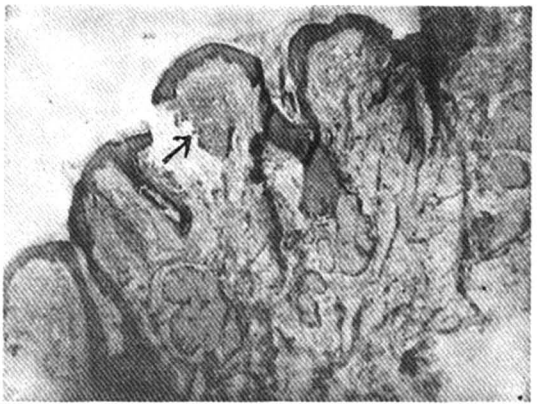

Fig. 4. Multiple non-caseating epitheliod cell granulomas with foreign body giant cells.

characterised by hypopigmentation such as vitiligo, leprosy and pityriasis versicolor. It has been observed that hypopigmentation in CS and other acquired disorders is due to degenerative changes in melanocytes resulting in depressed melanogenesis and not due to failure of transfer of melanosomes to keratinocytes.[4] Early diagnosis of CS is very important, because CS as an initial manifestation in otherwise healthy persons carries a comparatively good prognosis.[8]

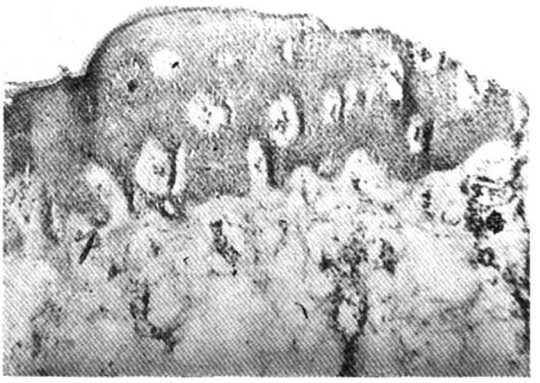

Fig. 5. Acanthosis, uniform elongation of the rete ridges, papillomatosis, and mononuclear cell granulomas with giant cells in the papillae.

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