Erythema Dyschromicum Perstans (EPD)

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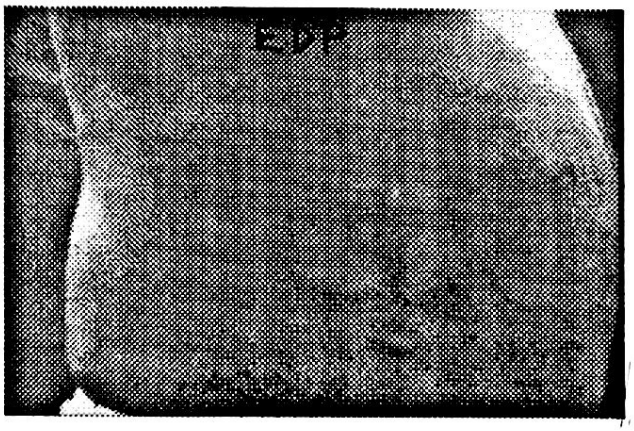
Introduction:

Ramirez first used the term Erythema Dyschromicum Perstans in 1957 in Salvadorians, so that these persons were called los cenicientos (The ash colored ones). Subsequent reports have included cases from Venezuela, Columbia, Mexico, Northern Europe and United Stated. It is an acquired a symptomatic idiopathic macular ashy gray or blue hyper pigmentation that occurs in otherwise healthy individuals. Usually, lesions are flat, there may be a thin raised erythematous border like a "thin piece of string". EDP deserves its colour from melanin complexes in epidermis and dermis. Primary event is vacuolization of basal cell layers. Some authers consider it as a variant of macular lichen planus or lichen pigmentosus.

Case Report



Regular soldier Hav. G.S. 40 years old male was presented with the complaints of asymptomatic gradually enlarging persistent oval macules with well defined slightly raised palpable borders, surface of the lesions were grey to blue coloured, in a referral center. The lesions were numerous varying in size from a few mm to 5 cm in diameter at places they were coalescent and polycyclic. Previous skin biopsy marks were evident on two sites in abdomen. The lesions were present on most of abdomen, a few on chest, back of trunk and proximal parts of all limbs. Initially, the patient noticed a few sking lesions over the abdomen while serving UNIFIL in 1989. He took treatment there as well as back in Kathmandu. Despitd the lesions continued to be progressive and spread to other sites. His friends suspected him as a case of Leprosy and were reluctant to make him as a friend. He became much worried, went from clinician to clinician in vain. Finally he landed to a referral center on June 1997. He was admitted in the ward with a provisional diagnosis of EDP. Later, the skin biopsy histopathological report came which was consistent with EDP. Skin biopsy report revealed basal cell hydroid degeneration, melanin in the epidermis, melanophages over upper dermis, perivascular dermal lympho histiocytic infiltrates.

The patient was put on Daps one 100 mg OD and Vitamin C for 1 year after base line lab investigation. No topical treatment was given.

On June 1998, the treatment was switched over to Clofazimine 100 mg OD as there was no much improvement. After 3 months he came for follow up with a slight improvement.

Discussion :

Andrew described this disease should be differentiated from irritant/allergic reaction to perfume, fragrance by its slightly evaluated well-defined margins. He mentions it common in Central America. Datta A K mentions this disease as reported from many countries including India. Bleehen S S described EDP clinically as numerous macules of varying shades of gray with a red slightly raised and palpably infiltrated margin. Baranda V L assessed the expression of several cell adhesion and lymphocyte activation molecules in EDP lesions, and evaluated the effect of clofazimine therapy on the expression of these molecules. Of 6 patients treated with 100 mg/d for 3 months, only 4 patients completed the dose for 3 months. Before clofazimine therapy, a noticeable expression of intercellular adhesion molecule 1 and HLADR molecule were detected in the kerationocyte basal cell layer. The dermal cell infiltrate expressed the activation molecule CD 69 and the cytotoxic cell marker CD94. After clofazimine therapy, the expression of intercellular adhesion molecule 1 and HLA-DR disappeared, as well as the mononuclear cell infiltrate. Of 4 patients, 2 of them had complete clearing of lesions and the other 2 with a marked improvement. Patients who had had active lesion at the beginning of treatment responded with complete recovery and they remained free of macules. It is well known that clofazimine produces uniform skin coloration that masks the dyschromia. Clofazimine also appeared to exert immunomodulatory and anti-inflammatory effect in patients with EDP. So, this patient was also put on clofazimine 100 mg OD but the response was only partial. This case report is because of rarity of such cases in our country.

Conclusion :

From 1995 till date I have seen only a single case of EDP. Most important thing to stress here is that it is not an alarming disease. Patient remains healthy. Patient needs to be told about the correct diagnosis and prognosis of the disease so that he will be at ease and not panic unnecessarily.

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