SELF-HEALING JUVENILE CUTANEOUS MUCINOSIS

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The clinical evolution and histological features of skin lesions in a 14-year-old boy were characteristic of self-healing juvenile cutaneous mucinosis. They were (a) early age of onset with fever, (b) plaque lesion over the nape of the neck and thigh, (c) nodular lesions over the scalp, face and periarticular regions and (d) spontaneous resolution.

Key Word : Cutaneous mucinosis

Introduction

Self-healing juvenile cutaneous mucinosis is a rare entity characterized by early age of onset and selective distribution of the lesions in the form of nodules and plaques, over the scalp, face, neck, abdomen, thighs and periarticular regions.[1-3] The onset is acute with moderate systemic symptoms, and in course of time, all the lesions resolve spontaneously. Though uncommon, with less than 5 cases reported in the world literature, the benign nature of the disease and good prognosis makes the clinical recognition extremely important.

Case Report

A 14-year-old boy presented with fever, arthralgia and skin lesions of 2 months duration. The lesions started in the nape of the neck and subsequently forehead, scalp, nose, back of hands and medial aspect of right thigh were involved over a few weeks time. Two morphologically distinct lesions were noted. The lesions over the neck and thigh were well defined hyperpigmented plaques with furrows and linear papules over it and non-tender. The lesions over the scalp, forehead, nose and periarticular regions (Fig.1) were nodular, multiple, nontender, soft to firm in consistency and 0.5 to 2 cm in size. Skin over the nodules was normal. Systemic examination was within normal limits.

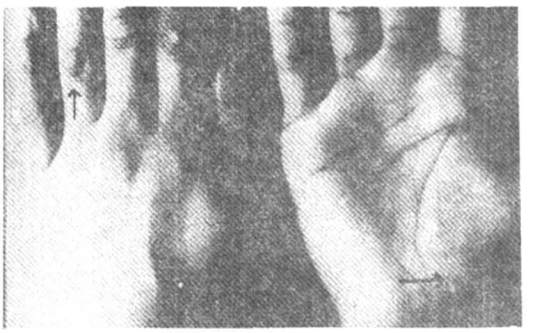

Fig. 1. Nodular lesions over the thenar eminence and proximal interphalangeal joints.

Routine urine and blood examination was within normal limits except for a mild albuminuria, which was transient. Detailed evaluation by the nephrologist did not reveal any renal dysfunction. His biochemical studies including lipid profile were normal. Serological tests for syphilis, streptococcal infections, rheumatoid factor and Australia antigen were negative. Thyroid function studies and radiological examination of skull, hands and chest were normal.

Skin biopsies were taken from the plaque and nodular lesions. Haematoxylin and eosin stained sections from both lesions, showed mild hyperkeratosis, and focal collections of lymphocytes around dermal vessels. The dermal architecture was altered

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due to wide separation of collagen bundles by pale blue homogenous material, consistent with dermal mucinosis (Fig. 2). The same was

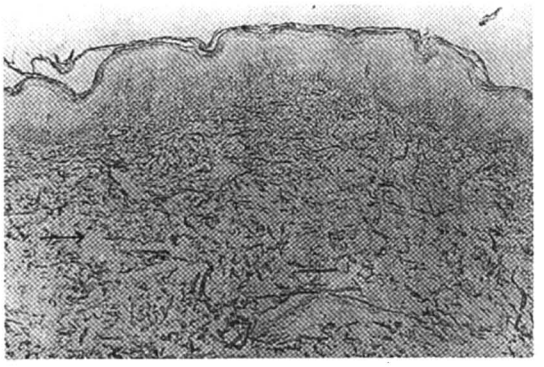

Fig. 2. Microscopic section of the nodular lesion from hand showing wide seperation of collagen bundles by homogenous material (H&E x 30).

Alcian blue postive and PAS negative, a staining pattern typical of acid mucopolysaccharides of dermal origin, predominantly hyaluronic acid. The histological picture was more pronounced in the nodular lesion. Electron microscopic study was done and the dermis showed electron lucent deposit between the collagen fibrils.

A diagnosis of self-healing juvenile cutaneous mucinosis was made with the clinical and histological picture, and the case was followed up without any specific treatment, The plaque lesions resolved in 3 months. Meanwhile the patient developed telogen effluvium. There was complete spontaneous clinical resolution with disappearance of nodular lesions and regrowth of hair by 8 months.

Discussion

Self-healing juvenile cutanneous mucinosis was first identified as a distinct clinical entity by Bonerandi et al in 1980.[1] It is differentiated form other forms of cutaneous mucinosis by its early age of onset, association with systemic symptoms, spontaneous resolution, and absence of dysglobulinemia and thyroid dysfunction. Cutaneous mucinosis is broadly classified into primary and secondary mucinosis.[2] Self-healing juvenile cutaneous mucinosis is a diffuse form of primary mucinosis.

The exact aetiology and nature of self healing juvenile cutaneous mucinosis is unknown. An altered fibroblast function due to a direct action by the virus or by its action through an immune mechanism are suggested as aetiology.[3] The presence of fever and other systemic symptoms in our case supports a viral aetiology.

The systemic symptoms may be recurrent and be associated with systemic hypertension. Both plaque and nodular lesions, as observed in this case, have been described in literature. Both plaque and nodular lesions may be observed in the same patient.[3] As the disease is self-limiting, no specific therapy is recommended. Very few cases of self-healing juvenile cutaneous mucinosis have been reported in world literature and to our knowledge, this is the first reported case in Indian literature.

Acknowledgement

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References

1. Bonerandi J, Andrc L, Follana J, et al . Self-healing juvenile cutaneous mucinosis. Ann Dermatol Venerol 1980; 107:51-7.
2. Black MM. Mucinoses. In : Champion RH, Burton JL, Ebling FJG, editors. Textbook of detmatology. Boston : Blackwell, 1992: 2325-33.
3. Pucevich MV, Latour DL, Bale GF, et al. Self-healing juvenile cutaneous mucinosis. J Am Acad Dermatol 1984; 11 : 327-32.