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EHLERS-DANLOS SYNDROME WITH DIFFUSE ALOPECIA

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A case of Ehlers-Danlos syndrome in a 20-year-old male involving cutaneous and skeletal systems is reported. Generalised alopecia involving scalp was an unusual and interesting feature in this case.

Key Words : Ehlers-Danlos syndrome, Alopecia

Introduction

The Ehlers-Danlos syndrome (EDS) is a collective group of heritable connective tissue dysplasias characterized by abnormal collagen synthesis.[1] It manifests clinically as joint hypermobility, skin hyperextensibility, poor wound healing with abnormal scarring and blood vessel fragility manifesting as easy bruising.[2] If the collagen abnormality is more generalized systemic complications are severe. Depending on clinical, genetic and biochemical defferences ten types of EDS are described.[3] Because of lack of pathognomonic laboratory tests, the diagnosis is mainly clinical.

Here we describe a case of EDS mitis with diffuse scalp alopecia.

Case Report

A 20-year-old male labourer attended the skin OPD for gradual loss of scalp hair of 2 years duration. The hair loss started over vertex area and then gradually involved rest of the scalp to leave scalp hair sparse. There was no history of any drug intake, radiation or any other major illness. Family history for any metabolic disorder was not contributory.

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On examination of the scalp, there was diffuse non-scarring type of alopecia affecting entire scalp. Hair was sparse all over body including scalp. Clinical progression and features ruled out alopecia areata as well as androgenic alopecia. On detailed examination, he had hypermobile joints and hyperextensibility of skin which could be stretched upto 3 cm (Fig. 1). He had 'cigarette paper' scars

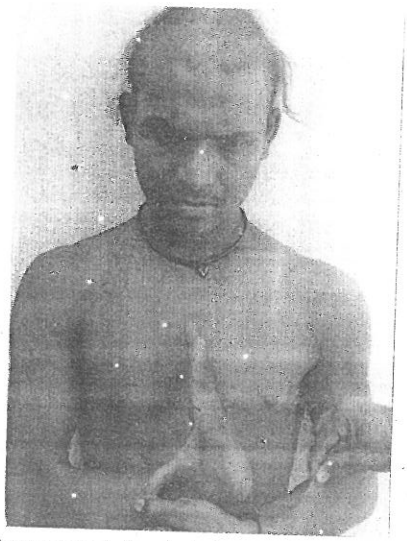

Fig. 1. Hypermobile joints and hyperextensible skin with alopecia

over elbows and knees which he got after trauma and gives history of easy bruising after trauma. According to Beighton`s scoring system,[4] he was a case of EDS-mitis variety; his score being 8.

Heamogram, urine, blood sugar, serum VDRL were within normal range. A punch biopsy was taken from volar skin of left forearm and from scalp. Light

microscopy studies of forearm biopsy showed no collagen changes. Scalp biopsy did not reveal any pathognomonic features indicative of alopecia areata (swarm of bees appearance).[5]

Comments

EDS is an uncommon connective tissue disorder affecting skin, joints, blood vessels leading to a spectrum of clinical features.[2] There has been no report of a hair disorder with EDS in the literature. Progression of alopecia in our patient, its clinical features and histology of the scalp helped to rule out androgenic alpecia and alopecia areata. Other disorders leading to such a diffuse hair loss (e.g., telogen effluvium, drugs, anagen effluvium) were also excluded by appropriate investigations. Perhaps this is the first case report of EDS-mitis with alopecia.

Though the pathogenesis of hair loss remains obscure, it can be postulated to be due to defects in the supporting collagen matrix of the hair papilla, a defect somewhat similar to that which leads to capillary fragility. High resolution E M and immunohistochemical characterization of collagen, that anchors the hair papilla may pinpoint the cause of such alopecia.

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