BLASCHKOID LICHEN PLANUS IN AN ADULT KASHMIRI MALE: A RARE PRESENTATION

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Introduction

Zosteriform and blaschkoid forms of lichen planus are rare [1]. Both the forms arise either as koebner’s phenomena, wolf’s isotopic phenomena or de novo from normal skin. There is a controversy about the use of terms like zosteriform/dermatomal/blaschkoid LP. We report a case of blaschkoid LP along with scalp LP in a 42 year old adult Kashmiri male. We report the case to add one more case to the list of this rare form of LP, with the peculiarity in our case of late onset of presentation and coexisting scalp LP, and review the literature to address the confusion about the various related terms.

Case Report

A 42 year old male, from urban background, advocate by profession, presented to our out patient department on 28th June 2012, with 6 months duration of violaceous, moderately itchy skin lesions which started on left shoulder and gradually progressed over 6 months to involve upper limb and also appeared on left trunk, relieved only partially by topical steroid application. He also gave history of 2 year duration of asymptomatic violaceous-black eruptions on scalp leaving patches of alopecia on healing and having received topical and oral steroids for the scalp lesions. There was nothing significant in the family and drug history. There was no trauma, pain or blistering prior to or during the evolution of the skin eruption. He had no associated co-morbidity like diabetes, hypertension.

General physical examination was normal and nothing abnormal was found on systemic examination. On cutaneous examination, there were multiple unilaterally distributed violaceous, purple, flat papules and plaques of variable sizes, discrete and coalesced, in continuous and interrupted linear pattern as well as in patterns of whorls and wide bands, confined to the left side of body involving anterior and posterior-lateral aspect of arm and forearm, scapular area, anterior and posterior-lateral aspect of trunk, extending over a length of 15-20 cm, with few lesions healed with post inflammatory brownish hyper pigmentation (Fig. 1a, 1b). Some of the lesions were covered with fine adherent scaling and wickham’s striae (Fig. 2a, 2b). Scalp showed multiple violaceous plaques with scarring alopecia and covered with fine scaling, distributed symmetrically over whole scalp and with normal hair texture (Fig. 3a-3c). The oral mucosa, hair and nails were normal. Complete blood count, liver function tests, kidney function tests, urine examination, chest X-Ray, ECG and ultrasound abdomen were normal. Hepatitis B and C serology was negative. A differential diagnosis of lichen planus, lichen striatus, acquired blaschkoid dermatitis was considered and punch biopsy taken from cutaneous as well as scalp lesions.

Histopathological examination from one of the papules on skin under hematoxylin & eosin staining (H & E) showed hyperkeratosis, irregular acanthosis, basal cell degeneration in the epidermis and band like lymphocytic infiltrate in papillary dermis (Fig. 4a, 4b), and that from the scalp lesion with H & E stain showed atrophic epidermal lining with prominent basal pigmented layer, pigment incontinence and chronic inflammatory infiltrate in dermis (Fig. 5a, 5b), both suggesting lichen planus. Direct immune-fluorescence result was negative.
In view of the typical clinical features and histopathology findings, a diagnosis of LP along blaschko lines (Blaschkoid LP) was made and patient was put on oral mini-pulse of steroids with methyl-prednisolone 32mg on 2 consecutive days and also given potent topical steroid clobetasol and oral antihistamines and is doing well.

Figure 1a, b. Unilateral purple papules and plaques along blaschko lines with brown post inflammatory pigmentation

Figure 2a,b. Close up view of the lesions with wickham’s striae

Figure 3a - c. Cicatricial alopecia with underlying brown-purple pigmentation on scalp
Discussion

Linear form of lichen planus can manifest as zosteriform (dermatomal) or blaschkoid form (along blaschko lines). Dermatome is an area of skin supplied by a spinal cutaneous nerve. Blaschko lines represent a form of “mosaicism”, where two or more genetically distinct cell populations are present in an individual derived from a single zygote [2,3]. The lines of blaschko were first described by a German dermatologist, Alfred Blaschko in 1901 [2]. These are distinct from the other known linear patterns of the skin [3]. Blaschko’s lines do not correspond to any known nervous, vascular or lymphatic structures [2], but represent developmental growth pattern of the skin [4].

The lines of blaschko may be followed by some X-linked, congenital and inflammatory skin disorders [3]. Blaschko lines are V-shaped on the upper spine, S-shaped on the abdomen, inverted U-shaped from chest area to the upper arm, and perpendicular over the front and back of the lower limbs. They never cross the anterior truncal midline, but run along it.

Zosteriform lichen planus is a rare variant of lichen planus, which shows lichenoid papules forming a broader band that follows the dermatomes. The entity of zosteriform/dermatomal LP is controversial. Many argue that the term zosteriform lichen planus has been applied inappropriately in cases that actually arise de novo in the lines of blaschko, rather than in true dermatomes [5,6]. Some believe that true zosteriform LP does not exist except in cases arising on the site of healed herpes zoster [7].

Blashkoid LP [8,9] accounts for less than 0.5% of patients with LP [10]. It has been reported more commonly in children than adults. It is very difficult to explain the occurrence of an acquired disorder like LP along the blaschko’s lines, which are normally followed by the inherited/ genetic disorders. It seems that there exists a genetic pre-disposition to lichen planus and exposure to an appropriate enviromental or endogenous trigger may lead to the development of lichen planus.

Blashkoid LP or the controversial zosteriform/ dermatomal LP may arise as koebner phenomena because of disseminated disease [10] or at site of previous healed zoster as wolf’s isotopic phenomena [6-9,12-15] or appear de novo [15-17] on previously normal non traumatized skin [18].

With the present available literature, it is difficult to differentiate the two terms with confidence. So it remains unexplored if there are two separate forms of unilateral, de novo lichen planus; one type arising in the lines of blaschko (Blaschkoid LP) [8,9] and the other arising within one or more dermatomes (dermatomal LP). Our patient presented with unilateral linear, curled lesions of LP on normal skin along the blaschko lines, along with scalp LP and the peculiarity of adult onset of presentation, which is rare.

Figure 4a, b. Skin biopsy shows hyperkeratosis, irregular acanthosis, basal degeneration and band like lymphocytic infiltrate in dermis

Figure 5a, b. Scalp biopsy shows atrophic epidermas, prominent basal pigment with incontinence and inflammatory infiltrate in dermis
REFERENCES