DERMATOLOGY PRACTICAL & CONCEPTUAL
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Linear atrophoderma of Moulin: a case report and review of the literature

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Key words: linear, atrophoderma, Moulin, linear scleroderma

Citation: Patsatsi A, Kyriakou A, Chaidemenos G, Sotiriadis D. Linear atrophoderma of Moulin: a case report and review of the literature. Dermatol Pract Conc. 2013;3(1):3. http://dx.doi.org/10.5826/dpc.0301a03.

Received: June 14, 2012; Accepted: November 25, 2012; Published: January 31, 2013

Funding: None.

Competing interests: The authors have no conflicts of interest to disclose.

All authors have contributed significantly to this publication.

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ABSTRACT Linear atrophoderma of Moulin is a rare, acquired, linear dermatosis. We present a 17-year-old girl with multiple asymptomatic brownish atrophic plaques in a zosteriform distribution on the left side of the trunk. Clinical presentation and dermatopathology was compatible with the diagnosis of linear atrophoderma. Twenty years after its initial description by Moulin, there are yet a limited number of case reports and unanswered questions regarding this entity.

Case presentation

A 17-year-old girl presented with a six-month history of multiple asymptomatic brownish atrophic plaques in a zosteriform distribution on the left side of her trunk (Figure 1). There was no family history of a similar skin disease. Laboratory studies were unremarkable for any chronic or autoimmune disorder. Biopsy of a lesion showed a normal epidermis with increased pigmentation of the basal layer and a broad dermis with thickened collagen fibers and diminished periadnexal and subcutaneous fat tissue (Figures 2, 3, 4).

Clinical and dermatopathologic findings were compatible with atrophoderma of Moulin. The use of topical steroids and tacrolimus as a second-line regimen did not result in any improvement of lesions.

Discussion

Linear atrophoderma is a rare, acquired, linear dermatosis. It is named after Moulin, who, in 1992, reported on five patients with pigmented and more or less atrophic bands along Blaschko's lines [1].

The age of onset in the first described cases ranged from 6 to 20 years. Lesions were unilateral, forming a recumbent "S" pattern, and the intensity of pigmentation and atrophy was variable. They remained stable throughout an observation period of 2 to 30 years. Of the skin biopsies performed on three patients, there was only irregular and moderate hyperpigmentation of the basal layer. In the dermis, there was no distinct pigment incontinence, no inflammation or alteration of connective tissue texture, and the clinical

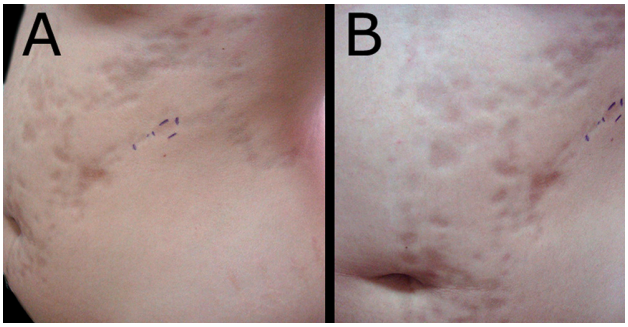

Figure 1. Atrophic plaques on the left side of the trunk. [Copyright: ©2013 Patsatsi et al.]

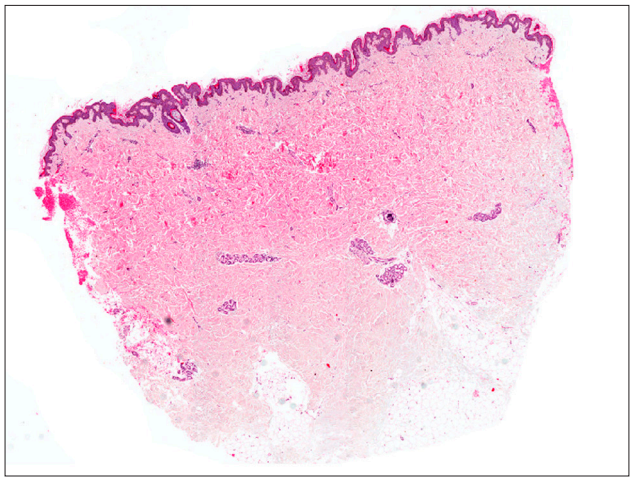

Figure 2. Normal epidermis and broad dermis. [Copyright: ©2013 Patsatsi et al.]

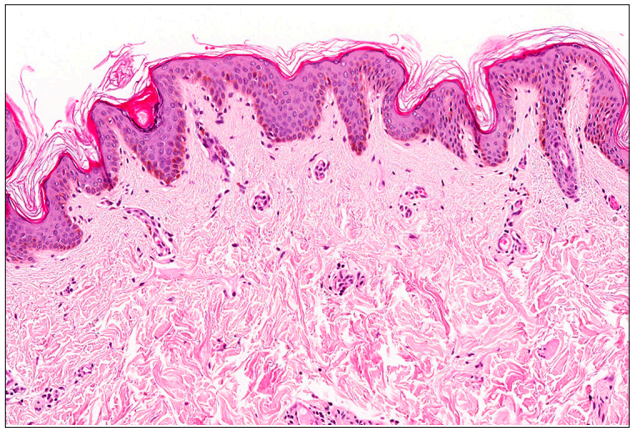

Figure 3. Normal epidermis with hyperpigmented basal layer. [Copyright: ©2013 Patsatsi et al.]

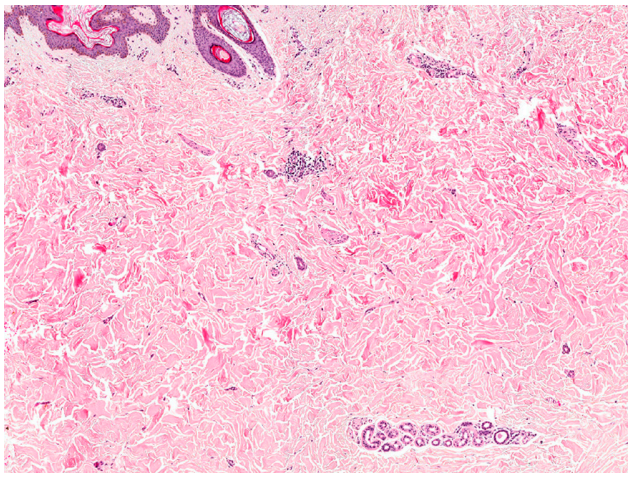

Figure 4. Thickened collagen fibers in the dermis. [Copyright: ©2013 Patsatsi et al.]

impression of skin atrophy was attributed to atrophy of the subcutaneous tissue [1].

Up to now there have been 30 reported cases of linear atrophoderma of Moulin [2-22]. In 2005, Ang et al mentioned that many cases of linear dermatoses were grouped under the umbrella of linear atrophoderma [23]. In Table 1, 30 reported cases that resemble the initial description of linear atrophoderma of Moulin are listed.

Now, twenty years after the initial description of this entity, there are still some unresolved issues. The differential diagnosis of zosteriform or linear scleroderma is not clear clinically nor dermatopathologically. It may well be that linear atrophoderma of Moulin and zosteriform or linear scleroderma belong to the spectrum of a single disease.

With the growing literature it has been demonstrated that age of onset is not limited to childhood or adolescence. The disease may also present later in life. Lesions are not always unilateral. They may be bilateral but in a linear distribution along Blaschko's lines (Table 1). Distribution mainly along the Blaschko lines reflects mosaicism.

Dermatopathologic findings of linear atrophoderma of Moulin vary. The epidermis is normal in the majority of cases. However, in two reports there was vacuolar degeneration of the basement membrane [2,7]. Few reports suggest an inflammatory early stage is suggested. The question of a transient inflammatory early stage that is no longer present by the time of clinical evaluation and biopsy remains.

Although the alteration of the connective tissue is not described in the original paper, most of the reviewed papers describe an increase of collagen. Unaltered, fragmented or decreased collagen fibers have been also reported in isolated cases, as well as edema of the dermis (Table 1). Another issue that has not been addressed is the cause of the clinical presentation of atrophic plaques. Does the loss of subcutaneous fat result in atrophy?

In conclusion, even now, 20 years after the first description of atrophoderma of Moulin this rare disease is puzzling. It seems to occur at any age. It is characterized clinically by the presence of atrophic patches distributed along Blaschko lines and dermatopathologically by a normal epidermis with a hyperpigmented basal layer, a dermis with thickened collagen fibers and loss of subcutaneous fat. Etiology and pathogenesis of this disease remains still unclear.

TABLE 1. Reported cases of linear atrophoderma of Moulin

Cases	Case no.	Age at presentation/ sex	Area of involvement	Disease duration when reported	Histological findings
Moulin 1992, 1st of 5 cases [1]	1	8/M	Left side of trunk		
Moulin 1992, 2nd of 5 cases [1]	2	7/F	Right side of trunk		
Moulin 1992, 3rd of 5 cases [1]	3	15/M	Right side of trunk		
Moulin 1992, 4th of 5 cases [1]	4	20/M	Left side of trunk		
Moulin 1992, 5th of 5 cases [1]	5	6/M	Left arm and trunk		
Baumann et al 1994 [2]	6	22/M	Right arm and trunk		Ballooning in basal epidermis (?), increased collagen in dermis
Braun RP, Saurat JH, 1996 [3]	7	16/M	Left side of back, left side of abdominal area		
Wollenberg A et al, 1996 [4]	8	5/F	Right arm/ trunk	17	Epidermal atrophy, increased collagen in dermis
Artola Igarza JL et al, 1996 [5]	9	16/F	Left part of the trunk		Increased collagen in the dermis
Cecchi and Giomi, 1997 [6]	10	12/F	Right arm and back		Hyperpigmentation in the basal layer of the epidermis
Rompel R et al, 2000 [7]	11	17/F	Right side of the trunk, right buttock		Focal vacuolar degeneration of the basal layer, increased collagen in dermis
Browne C et al, 2000 [8]	12	10/M	Trunk and limbs, bilateral	6	Slightly thinned epidermis with prominent blood vessels, slight increase in the amount of collagen
Martin et al 2002 [9]	13	9/M	Left side of trunk		Increased collagen
Miteva L et al, 2002 [10]	14	20/F	Right arm, buttock and leg	4 years	Increased collagen
Utikal et al, 2003 [11]	15	29/M	Limbs and trunk, bilateral	6 years	Slight edema of the dermis
Utikal et al, 2003 [11]	16	15 /F	Limbs and trunk, bilateral	13 years	Slight edema of the dermis
Danarti et al, 2003 (1st of 4 cases) [12]	17	14/F	Left side of the body	1 year	Perivascular lymphocytic infiltrate
Danarti et al, 2003 (2nd of 4 cases) [12]	18	24/F	Left side of her abdomen and her back, left arm		Biopsy not performed
Danarti et al, (3rd of 4 cases) [12]	19	38 /F	Left medial thigh	1.6 year	Unremarkable epidermis and dermis

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TABLE 1. (continued)

Cases	Case no.	Age at presentation/ sex	Area of involvement	Disease duration when reported	Histological findings
Danarti et al, 2003, (4th of 4 cases) [12]	20	15/F	Left side on the buttock and on her lateral iliac crest along Blaschko's lines	1 year	Biopsy not performed
Miteva L et al 2005 [13]	21	9/M	Left side of the trunk, left upper limb	5 years	Increased collagen
Atasoy M et al, 2006 [14]	22	16/M	Right side of the trunk, right arm	2 years	Epidermal atrophy, fragmented collagen fibers
Zampetti A et al, 2008 [15]	23	37/F	Left arm and trunk	5 years	Hyperpigmentation of epidermal basal cells, slight thickening of the collagen fibers in the mid-deep dermis
Gecchi et al, 2008 [16]	24	9/M	Exclusive involvement of the neck	1 year	A normal epidermis with moderate, diffuse hyperpigmentation of the basal layer A perivascular lymphocytic infiltrate was noted in the dermis, without any other pathologic feature
Lopez N et al, 2008 [17]	25	17 /M	Right upper arm	1 year	Localized hyperpigmentation in the basal layer of the epidermis
Ozkaya E et al, 2010 [18]	26	18/F	Legs, arms and trunk including the axillary regions	1 year	Slight epidermal acanthosis, a slightly hyperpigmented basal layer, and a slight decrease in elastic fibers in papillary dermis
Ripert C et al, 2010 [19]	27	14/F	Left side of trunk	15 months	Dermal atrophy with pigmentation of the basal layer and a perivascular lymphocytic infiltrate
Schepis C et al, 2010 [20]	28	14/M	Left side of trunk	A few months	A hyperpigmented basal layer In the upper dermis, dilated superficial vessels were visible The mid and deep dermis were mildly edematous

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TABLE 1. (continued)

Cases	Case no.	Age at presentation/ sex	Area of involvement	Disease duration when reported	Histological findings
Tukenmez Demirci G et al, 2011 [21]	29	39/F	Left half of the neck	22 years	A normal epidermis outlined by a hyperpigmented-basal layer In the papillary dermis proliferation of superficial vessels with mild lymphocytic infiltrate and melanin-laden macrophages were present Collagen fibers and elastic fibers were normal
Norisugi O et al, 2011 [22]	30	26/M	Right side of trunk, posterior right leg	1 year	Thickness of the subcutaneous tissue was reduced in lesional skin compared to normal skin by ultrasound examination
Our case	31	17/F	Left side of trunk	6 months	Thin epidermis, hyperpigmentation of the basal layer, increased collagen fibers, rather thin subcutis

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