Acta Derm Venereol 2014; 94: 361–362

SHORT COMMUNICATION

Linear Collagen Naevus: An Unusual Connective Tissue Disorder

Walter Dubiel[1], Ingomar Kiehlmann[2] and Rudolf Happle[3]*
*[1]Dermatologist, Heubuck 3, Horben, [2]Laboratory of Dermatohistopathology, Offenbach, and [3]Department of Dermatology, Freiburg University Medical Center, Hauptstr. 7, DE-79104 Freiburg, Germany. *E-mail: rudolf.happle@uniklinik-freiburg.de*
Accepted Aug 26, 2013; Epub ahead of print Oct 24, 2013

Connective tissue naevi are circumscribed skin lesions characterised by an increase or structural alteration of either collagen or elastin fibres (1). Collagen naevi occur in a patchy form as a feature of tuberous sclerosis, whereas elastin-rich naevi are noted in Buschke-Ollendorff syndrome. In both of these autosomal dominant traits, rather large lesions probably represent a type 2 segmental manifestation, being superimposed on the nonsegmental phenotype (2). By contrast, the linear collagen naevus tends to occur as a sporadic and isolated disorder (3, 4). Here we describe a further case of this unusual naevus.

CASE REPORT

A 20-year-old woman presented with a linear skin lesion involving her right leg. At the age of 7 years she had first noted some ivory-coloured papules in her right popliteal area. Within one year these lesions increased and formed plaques arranged in a band starting from the dorsal surface of her thigh and running down to the calf. During the past few years, the originally whitish lesions took a somewhat pale-pink hue. The patient reported that none of her family members had ever shown similar skin lesions.

On physical examination, multiple firm, pale-pink papules and plaques were noted in a Blaschko-linear arrangement extending from beneath the right buttock to the calf (Fig. 1). Microscopic examination of a lesional biopsy obtained from the right thigh showed dermal accumulation of dense, coarse collagen fibres, whereas the elastic fibres appeared to be rarified and fragmented (Fig. 2). No signs of inflammation were noted.

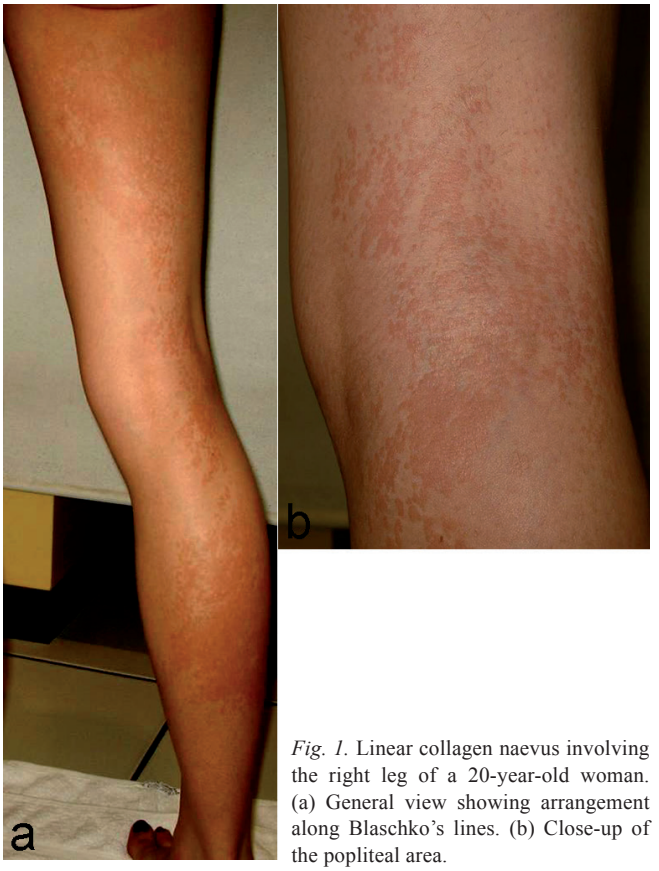

Fig. 1. Linear collagen naevus involving the right leg of a 20-year-old woman. (a) General view showing arrangement along Blaschko's lines. (b) Close-up of the popliteal area.

DISCUSSION

This patient had a typical collagen naevus of the linear type. Similar cases have previously been reported under

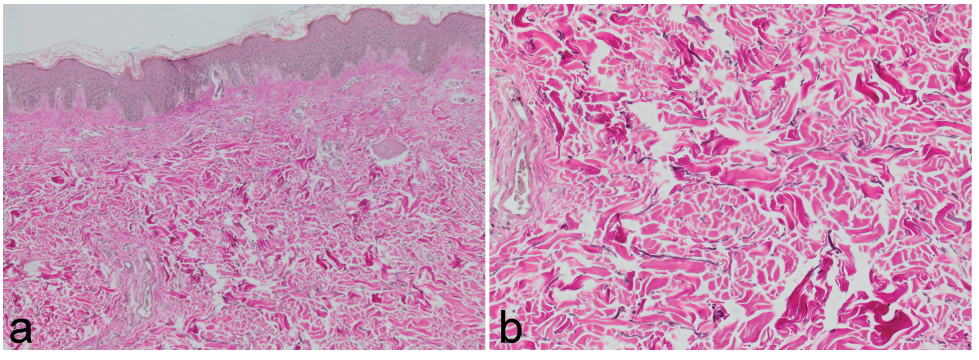

Fig. 2. The rather broad dermis shows abundance of coarse collagen fibres, whereas the elastic tissue fibres are diminished and partly fragmented. (a) Elastica-van Gieson, ×20. (b) Elastica-van Gieson, ×40).

the term "zosteriform connective tissue naevus" (5–7), but for obvious reasons the arrangement of this disorder is not "zosteriform" (8). Other authors preferred the name "papulolinear collagenoma" and thought that the disorder followed Blaschko's lines but they were not quite sure about this assumption (4, 9, 10). The present case lends further underpinning to their view. Remarkably, a linear collagen naevus that was noted in a patient with phacomatosis pigmentokeratotica showed likewise an arrangement along Blaschko's lines (11).

Several other authors have documented a Blaschko-linear arrangement of connective tissue naevi (12–15). It seems difficult, however, to determine whether these naevi were of a collagen or elastin type.

In conclusion, the present case lends additional support to the notion that the linear collagen naevus tends to occur in a nonsyndromic form and is arranged along Blaschko's lines.

ACKNOWLEDGEMENT

We thank Ms. Ludmilla Jost, Bad Homburg, for taking the clinical photographs.

The authors declare no conflicts of interest.

REFERENCES

1. Uitto J, Santa Cruz DJ, Eisen AZ. Connective tissue nevi of the skin. J Am Acad Dermatol 1980; 3: 441–461.
2. Happle R. Superimposed segmental manifestation of both rare and common cutaneous disorders: a new paradigm. Actas Dermosifiliogr 2009; 100 (Suppl 1): 77–85.
3. Herbst VP, Kauh YC, Luscombe HA. Connective tissue nevus masquerading as a localized linear epidermal nevus J Am Acad Dermatol 1987; 16: 264–266.
4. Romiti R, Romiti N. Papulolinear collagenoma. J Am Acad Dermatol 2004; 50: 797–798.
5. Amjadi M, Khorrami-Arani N, Mashman G, Allen PW. Zosteriform connective tissue nevus: a case report. Am J Dermatopathol 2007; 29: 303–305.
6. Choi YJ, Lee SJ, Choi CW, Kim WS, Lee GY. Multiple unilateral zosteriform connective tissue nevi on the trunk. Ann Dermatol 2011; 23 (Suppl 2): S243–246.
7. Brazzelli V, Muzio F, Barbagallo T, Fornara L, Donadini F, Guerci B, et al. Zosteriform connective tissue nevus in a pediatric patient. Pediatr Dermatol 2007; 24: 557–558.
8. Happle R. "Zosteriform" lichen planus: the bizarre consequences of a misnomer. Acta Derm Venereol 1998; 78: 300.
9. Girard C, Bessis D. Papulolinear collagenoma. J Am Acad Dermatol 2006; 54 (Suppl. 5): S240.
10. Lo LK, Tsai TF, Chen YF, Hung CM, Ko WC. Papulolinear collagenoma with arborizing arrangement: report of a case. Pediatr Dermatol 2009; 26: 111–112.
11. Boente MC, Asial RA, Happle R. Phacomatosis pigmentokeratotica: a follow-up report documenting additional cutaneous and extracutaneous anomalies. Pediatr Dermatol 2008; 25: 76–80.
12. Steiner K. Connective tissue nevus. Arch Dermatol Syphilol 1944; 50: 183–190.
13. Kozminsky ME, Bronson DM, Barsky S. Zosteriform connective-tissue nevus. Cutis 1985; 36: 77–78.
14. Yeh SW, Magalhaes AM, Vasconcellos MRA, Michalany NS, Yamashita JT. Zosteriform connective tissue nevus: a case report. Int J Dermatol 2003; 42: 720–722.
15. Asano Y, Ihn H, Tamaki K. Linear connective tissue nevus. Pediatr Dermatol 2007; 24: 439–441.