Case Report

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Meyerson Phenomenon within a Nevus flammeus

The Different Eczematous Reactions within Port-Wine Stains

Thomas Hofer

Private Practice, Wettingen, Switzerland

Key Words
Port-wine stain · Eczematous reaction · Meyerson phenomenon · Genetic mosaicism

Abstract
Only few reports about eczematous reactions overlying nevi flammei exist. All of them were observed in children. The description of an eczematous reaction within a congenital nevus flammeus on the left lower leg of a male adult gives reason to discuss this rare phenomenon. Eczema or inflammatory changes within a port-wine stain may mostly be a collision dermatosis with an atopic dermatitis, especially when they arise in children and are localized to the neck and face. When they are observed within a grossly visible vascular malformation, as for example in the Klippel-Trenaunay syndrome, they may have a pathogenesis similar to stasis dermatitis. In rare cases, an eczematous reaction within a nevus flammeus may be the result of genetic mosaicism and is interpreted as a variant of the so-called Meyerson phenomenon.

Introduction

There are only few reports about inflammatory or eczematous skin changes limited to the area of port-wine stains or congenital nevi flammei [1, 2]. These observations are always made in children and are mostly localized to the neck and face. They are rarely found in the context of a Klippel-Trenaunay syndrome [3]. The pathogenesis of this observation is not yet clarified.

In 1971, Meyerson [4] described for the first time a papulosquamous halo dermatitis overlying pigmented nevi. Unlike the more frequently observed halo nevus or nevus Sutton, the nevus remains unchanged after the inflammatory process has disappeared. In 1988, Nicholls and Mason [5] proposed the eponym 'Meyerson's nevus'. Since then, this, also called 'Meyerson phenomenon' [6], has rarely been seen in relation with other skin tumors [7, 8]. In those cases, the pathogenesis again has yet to be clarified.

Here for the first time a male adult is reported who developed an eczematous reaction within a congenital nevus flammeus on his left lower leg. A possible relation between this observation and the Meyerson phenomenon is discussed.

Case Report

A 46-year-old man had noticed a change in the appearance of a congenital nevus flammeus on his left lower leg for 1 year. He hardly complained about itchiness. His personal medical history was uneventful. There was no history of eczema or allergy. Physical examination revealed a brown reddish area, about 12 cm in diameter with a pronounced and scaly margin, within a nevus flammeus on the anteromedial aspect of the left lower leg (fig. 1). Further physical examination revealed no other skin abnormalities. The patient does not have any varicose veins. The investigation with color duplex sonography (Sonoline Versa Pro®, Siemens, Linear Array 5,0L45) did not reveal any insufficient perforating vein under the nevus flammeus. On histological examination focal acanthosis, parakeratosis and spongiosis were found overlying some capillary ectasias. There was no inflammatory infiltrate (fig. 2). Topically applied mometasone (Elocom®) healed the eczema within 2 weeks. Until today no recurrence has been noted.

Discussion

Eczematous skin reactions localized to vascular structures are common. They mostly arise as stasis dermatitis from varicose veins. They can electively be located to dermal varicose venectasias, but are more often less circumscribed. Histological examination shows either a subacute or chronic dermatitis with hemosiderin scattered through the dermis, and the dilated vessels are embedded in a fibrotic dermis [9]. The pathogenesis of stasis dermatitis is not yet completely clarified [10].

All the recently reported cases involving an eczematous reaction, within a nevus flammeus, occurred in children [1–3]. They are mostly localized to the neck (otherwise also called salmon patch of the midline) and

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Dr. Thomas Hofer
Dermatologie und Venerologie FMH
Winkelriedstrasse 10
CH–5430 Wettingen (Switzerland)
Tel. +41 56 427 27 22, Fax +41 56 427 00 12, E-Mail thomas.hofer@active.ch

face. Sidwell et al. [3] suggest that it may be caused by an abnormal production of cytokines, resulting in inflammatory changes within the skin. It is also possible that the endothelial cells could play an important role in the inflammatory process. Since the eczema improves by laser therapy to the port-wine stains [1, 3], Sidwell et al. [3] suppose that the increased vasculature predisposes to the development of eczema. Also Bonifazi and Mazzotta [2] suppose, that '...the vascular malformation, with the relevant vasodilatation, may be the localization factor of the inflammatory changes...'. If this were the explanation for this observation, should it not also be possible to find an eczema more frequently within nevi flammei of adults or within port-wine stains otherwise localized than the head, or even also overlying the 'couperose' of patients with rosacea? Furthermore nuchal nevus flammeus is found in 15.6% of newborns [11]. Atopic eczema in children and adolescents is localized in 10.2% on the nape and in 30.8% on the neck [12]. Therefore the coincidence of an eczema within a nuchal nevus flammeus could also be interpreted as the result of a statistical probability. Eczema taking place within a grossly visible vascular malformation [3], as in the Klippel-Trenaunay syndrome with arteriovenous anastomoses and varicose vein segments, may have a pathogenesis comparable to stasis dermatitis. Unfortunately the reports of Sidwell et al. [3] and Bonifazi and Mazzota [2] lack any information about a histological examination.

In 1971, Meyerson [4] described for the first time a papulosquamous halo dermatitis overlying pigmented nevi. Histological examination shows an irregular acanthosis, parakeratosis and focal spongiosis overlying an otherwise unchanged nevus. There are no inflammatory cells invading the pigmented nevus. A lymphocytic perivascular infiltrate is present just peripheral to the nevus. Unlike the more frequently observed halo nevus or nevus Sutton, the nevus remains unchanged, after the inflammatory process has disappeared. In 1988, Nicholls and Mason [5] proposed the eponym 'Meyerson's nevus'. Since then, the 'Meyerson phenomenon' [6], also called 'nevocentric' halo dermatitis [13], has rarely been seen in relation with other skin tumors [7, 8] or inflammatory skin diseases such as atopic eczema (own observation), pityriasis rosea [14], erythema multiforme [13] and psoriasis [15]. Even if an immunological process is suggested [16–18], the exact pathogenesis of this phenomenon has yet to be clarified.

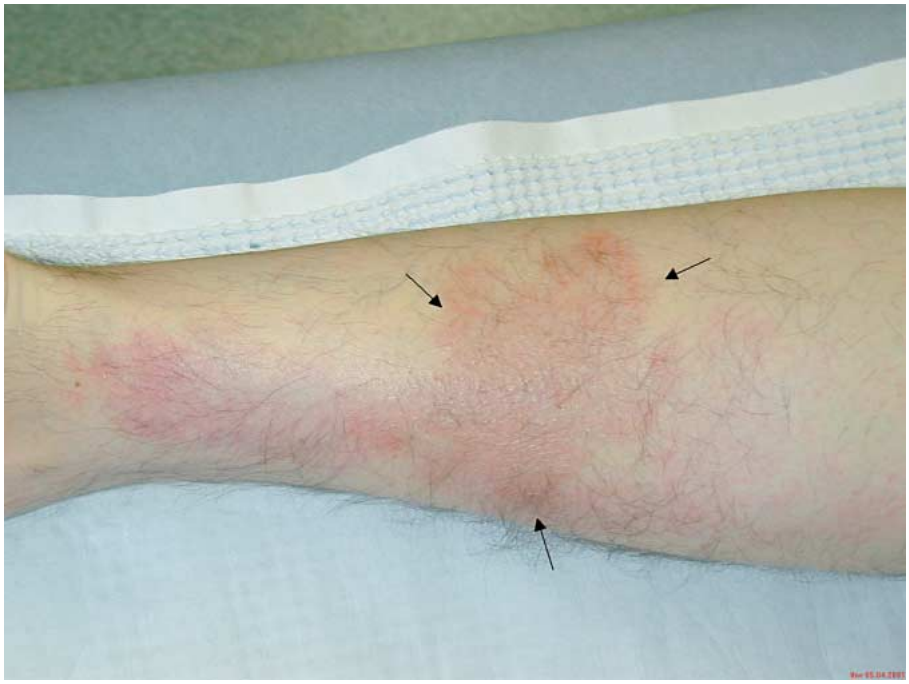

Fig. 1. Eczema (↑) within the port-wine stain.

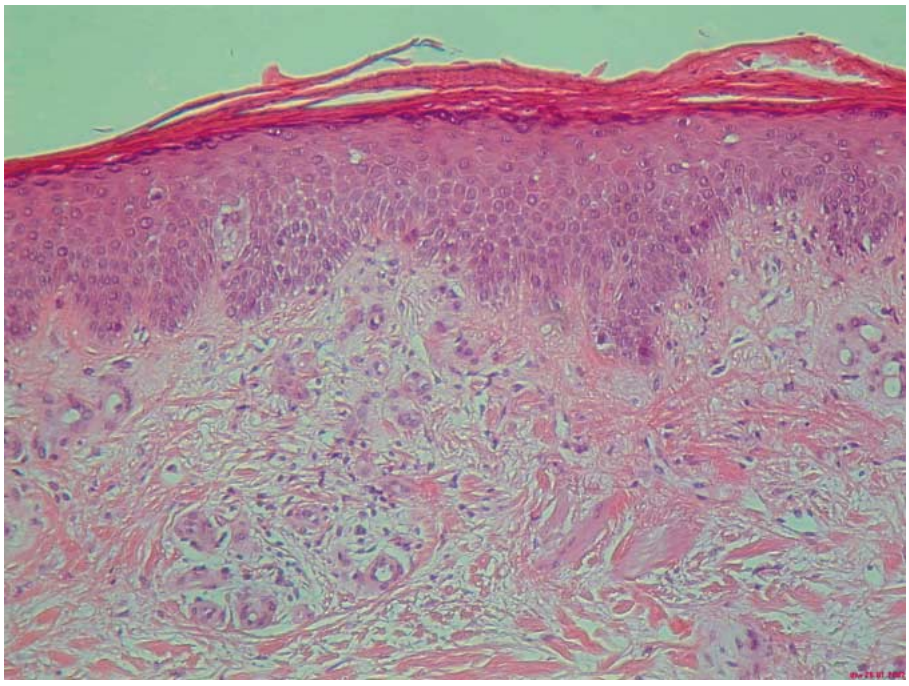

Fig. 2. On histological examination, focal acanthosis, parakeratosis and spongiosis overlying some capillary ectasias are seen. There is no inflammatory infiltrate. HE. × 100.

In 1980, Goudie et al. [19] postulated that endothelial cells, arranged in monoclonal zones, '... may be involved in the pathogenesis of hitherto poorly understood diseases ... for example, vascular clones in the dermis regulate the behavior of epidermal melanocytes'. Their hypothesis tried to explain a harlequin-like pattern of skin pigmentation in a Negress, which we nowadays would describe as a patient with chimerism, her pattern of pigmentation resembling a checkerboard, but also including ovoid lentiginous macules, with their long axes orientated along the lines of Blaschko. Congenital, nevoid and acquired skin diseases following the Blaschko lines are thought to be caused by genetic mosaicism, resulting from lyonization in X-linked disorders, postzygotic somatic mutations in sporadic conditions and gametic half-chromatid mutations [20]. In 2000, Lipsker et al. [21] demonstrated for the first time that also an acquired *inflammatory dermatitis* in Blaschko-linear distribution shows genetic mosaicism. Another genetic concept, loss of heterozygosity in human skin, plays a role not only in cutaneous malignant growth but also in the development of benign skin disorders [22]. It may also be the explanation for the coexistence of a linear and disseminated drug eruption: the loss of heterozygosity in a somatic cell, during early embryogenesis, gives rise to a clonal population of cells being either homozygous or hemizygous for a gene predisposing to drug eruptions. The disseminated rash would reflect a heterozygous state, whereas the more pronounced linear lesions reflect loss of heterozygosity [23].

In 1995, Happle proposed a new definition of what we call a nevus: 'Nevi are visible, circumscribed, long-lasting lesions of the skin or the neighboring mucosa, reflecting genetic mosaicism. With the exception of melanocytic nevi, they do not show neoplastic growth. They never show malignant neoplasia' [24]. He supposes that nevi, as a result of their genetic mosaicism, show a microenvironment which is distinct from that of the surrounding skin. This genetic mosaic may for instance explain the protection of a hairy pigmented nevus from hair loss, whereas the surrounding skin is undergoing an autoimmune reaction giving rise to alopecia areata [25]. In 1972, Burgoyne [26] also described a female patient with alopecia areata: treatment with local injection of triamcinolone diacetate was followed by regrowth of hair at the sites injected, but after a time the hair in these areas fell out, *except* in the area of a *nuchal nevus flammeus.* Port-wine stains fulfil the new definition of a nevus, even if their genetic mosaicism is not yet proved: they are visible, circumscribed, long-lasting lesions. They may develop thickening and nodules over time [27] and enhance vascular proliferation [28, 29] but they do not show neoplastic growth. Therefore one could conclude that nevi flammei, similar to melanocytic nevi, as a result of their genetic mosaicism, show a microenvironment which is distinct from that of the surrounding skin. This presumption would help to explain, why a nevus flammeus does not loose its hair, whereas the surrounding skin is undergoing an autoimmune reaction giving rise to alopecia areata.

It is presumable that – similar to acquired skin diseases, inflammatory dermatitis or drug eruptions following the Blaschko lines – skin reactions like the Meyerson phenomenon occurring in the context of nevi, are also the result of genetic mosaicism. The here presented 46-year-old man has an eczematous reaction, strongly limited to the area of a congenital nevus flammeus on his left lower leg. The surrounding skin does not show any further abnormalities. There is no causative varicose vein detectable. The histological picture resembles the description by Meyerson in 1971. Therefore his skin reaction is interpreted as a new variant of the Meyerson phenomenon.

In conclusion, eczema or inflammatory changes within a port-wine stain may mostly be a collision dermatosis with an atopic dermatitis, especially when they arise in children and are localized to the neck and face. If they are observed within a grossly visible vascular malformation, as for example the Klippel-Trenaunay syndrome, they may have a pathogenesis similar to stasis dermatitis. In both cases, histological examination has yet to be done. Further in rare cases, as in this observation, an eczematous reaction within a nevus flammeus may be the result of genetic mosaicism and is interpreted as a variant of the so-called Meyerson phenomenon.

References

1 Tay YK, Morelli J, Weston WL: Inflammatory nuchal-occipital port-wine stains. J Am Acad Dermatol 1996;35:811–813.
2 Bonifazi E, Mazzotta F: Inflammatory nuchal-occipital port-wine stains (letter). J Am Acad Dermatol 1998;38:130.
3 Sidwell RU, Syed S, Harper JI: Port-wine stains and eczema. Br J Dermatol 2001;144:1269–1270.
4 Meyerson LB: A peculiar papulosquamous eruption involving pigmented nevi. Arch Dermatol 1971;103:510–512.
5 Nicholls DS, Mason GH: Halo dermatitis around a melanocytic naevus: Meyerson's naevus. Br J Dermatol 1988;118:125–129.
6 Worret WI: Halo-Ekzeme um Nävuszellnävi (Meyerson-Nävi). Hautarzt 1990;41:262–264.
7 Tegner E, Björnberg A, Jonsson N: Halo dermatitis around tumours. Acta Derm Venereol 1990;70:31–34.
8 Gallais V, Lacour J-P, Perrin C, Halioua B, Ortonne JP: Halo eczématiforme autour d'un histiocytofibrome: phénomène de Meyerson. Ann Dermatol Vénéréol 1993;120:617–620.
9 Lever WF, Schaumburg-Lever G: Noninfectious vesicular and bullous diseases; in Lever WF, Schaumburg-Lever G (eds): Histopathology of the skin, ed 6. Philadelphia, Lippincott, 1983, p 99.
10 Ramelet AA, Monti M: Cutaneous complications related to venous insufficiency; in Ramelet AA, Monti M (eds): Phlebology: The Guide. Paris, Editions scientifiques et médicales Elsevier SAS, 1999, pp 158–161.
11 Camacho F, Navas J: Nuchal nevus flammeus in alopecia areata. Dermatology 1992;184:158.
12 Kissling S, Wüthrich B: Localizations, manifestations and micromanifestations of atopic dermatitis in young adults: Personal follow-up 20 years after diagnosis in childhood. Hautarzt 1994;45:368–371.
13 Pariser RJ: 'Nevocentric' erythema multiforme. J Am Acad Dermatol 1994;31:491–492.
14 Crovato F, Nazzari G, Gambini C, Massone L: Meyerson's naevi in pityriasis rosea. Br J Dermatol 1989;120:318–319.
15 Botev IN: 'Nevocentric' dermatoses (letter). J Am Acad Dermatol 1995;33:842–843.
16 Fernandez Herrera JM, Aragües Montanes M, Fraga Fernandez J, Diez CA: Halo eczema in melanocytic nevi. Acta Derm Venereol 1988;68:161–163.
17 Feal-Cortizas C, Vargas-Diez E, Buezo GF, Aragües M: Meyerson's nevus immunohistochemical findings in two cases (abstract). Australas J Dermatol 1997;38(suppl 2):222.

18 Krischer J, Pechère M, Salomon D, Harms M, Chavaz P, Saurat JH: Interferon alfa-2b-induced Meyerson's nevi in a patient with dysplastic nevus syndrome. J Am Acad Dermatol 1999;40:105–106.
19 Goudie RB, Spence JC, Scothorne RJ: Do vascular clones determine developmental patterns? Lancet 1980;i:570–572.
20 Bolognia JL, Seth JO, Glick SA: Lines of Blaschko. J Am Acad Dermatol 1994;31:157–190.
21 Lipsker D, Cribier B, Girard-Lemaire F, Flori E, Grosshans E: Genetic mosaicism in an acquired inflammatory dermatosis following the lines of Blaschko. Arch Dermatol 2000;136:805–807.
22 Happle R: Loss of heterozygosity in human skin. J Am Acad Dermatol 1999;41:143–161.
23 Happle R, Effendy I: Coexisting linear and disseminated drug eruption: A clinical clue to the understanding of the genetic basis of drug eruptions. Eur J Dermatol 2001;11:89.
24 Happle R: What is a nevus? A proposed definition of a common medical term. Dermatology 1995;191:1–5.
25 Bon MA, Happle R, Itin PH: Renböck phenomenon in alopecia areata. Dermatology 2000;201:49–50.
26 Burgoyne JS: Regrowth of hair in nevus flammeus. Arch Dermatol 1972;105:758.
27 Klapman MH, Yao JF: Thickening and nodules in port-wine stains. J Am Acad 2001;44:300–302.
28 Burg G: Collision dermatosis: Angiolymphoid hyperplasia with eosinophilia developing within a congenital port-wine nevus. Dermatology 1993;187:293–295.
29 Kim TH, Choi EH, Ahn SK, Lee SH: Vascular tumors arising in port-wine stains: Two cases of pyogenic granuloma and a case of acquired tufted angioma. J Dermatol 1999;26:813–816.