Eczema within port wine stain: spontaneous and laser-induced Meyerson phenomenon

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Introduction

The first reports of eczematous inflammation within congenital malformations of postcapillary venules appeared in the mid-1990s with descriptions of dermatitis on PWS corresponding to Meyerson phenomenon, usually reported in the setting of melanocytic nevi. There is no universal explanation of the cause or pathogenesis of eczema occurring in PWS, but it may be precipitated by atopic disease or vascular laser treatment of the malformation. Here we described two non-atopic girls with dermatitis developing within their nevi flammei, in one temporally related to KTP laser treatment, and in the other obviously not associated with the treatment. However, in both patients the eczema responded well to a short course of topical corticosteroids.

Case reports

Case 1

The girl was born with a reddish patch on the right side of the face within the innervation areas of the V1–V3 branches of the trigeminal nerve. A detailed ophthalmological exam and brain magnetic resonance imaging excluded associated anomalies. At age 6 she was treated under general anesthesia with a long-pulse Nd:YAG (1,064 nm) laser, but two treatments ended with scar formation and no significant improvement in the lesion. She was then referred to our center at age 12 (Fig. 1). On examination, an erythematous and in places violaceous macular lesion with no infiltrated areas was seen on the right side of her face, including the upper lip, nose, cheek, lower eyelid, and preauricular skin. We commenced a series of partial and later whole-lesion treatments with a 532 nm KTP laser (3 and 4 mm spot sizes, radiant exposures 12 to 28 J/cm², pulse widths 15 to 30 ms) at 6- to 12-week intervals with significant improvement in the appearance of the PWS. Three months following the last laser treatment, at the end of the summer, she noticed pruritic erythematous, oozing papules, and plaques on the lesion above the right cheek and nose (Fig. 2). Family and personal medical history were negative for atopic disorders. We diagnosed Meyerson phenomenon within her PWS and prescribed a mild topical corticosteroid (alclometasone)

Abstract

Port wine stain (PWS, nevus flammeus) is a relatively common vascular malformation of postcapillary venules affecting 0.3 to 0.5% of newborn children. Since the mid-1990s, a case series and several case reports have described dermatitis on PWS corresponding to Meyerson phenomenon, usually reported in the setting of melanocytic nevi. There is no universal explanation of the cause or pathogenesis of eczema occurring in PWS, but it may be precipitated by atopic disease or vascular laser treatment of the malformation. Here we described two non-atopic girls with dermatitis developing within their nevi flammei, in one temporally related to KTP laser treatment, and in the other obviously not associated with the treatment. However, in both patients the eczema responded well to a short course of topical corticosteroids.

Keywords: port wine stain, eczema, Meyerson phenomenon

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Figure 1 | PWS lesions below the right eye, on the cheek, and on the right upper lip prior to laser treatment.

Figure 2 | Eczematous plaques within PWS lesions below the right eye, on the cheek, and on the right upper lip.
ointment for 2 weeks. The inflammation subsided, and 2 months following the cessation of topical treatment she is without eczematous changes within the nevus, but dermatitic patches have appeared behind her right ear.

Case 2

The second patient was a girl born with uncomplicated PWS on the chin and left cheek (V2 and V3) (Fig. 3a). At the age 2 she presented at our center and we referred her to a center abroad specializing in the laser treatment of infants and very young children. Because her insurance would not reimburse the expenses of the treatment abroad, she was referred to the local plastic surgery department. There she underwent a few treatments with a 532 nm KTP laser without any improvement. The treatments were then suspended without further referral. The parents, determined to have their daughter’s birthmark removed, again visited our center when she was 12.

We performed several partial treatments with a 532 nm KTP laser (4 mm spot size, 20 J/cm² radiant exposure, 15 ms pulse width) with excellent clearance (Fig. 3b). However, a week following the fourth treatment, numerous scaling, erythematous, pruritic papules developed on her chin at the site of the remnants of the PWS (Fig. 3c). Personal and family histories for atopic disorders are negative. After a 2-week course of methylprednisolone aceponate cream, the lesions regressed completely and have not recurred for 6 months.

Discussion

Inflammatory reaction in PWS has been described on multiple occasions, first only in the head and neck region of children, later also in adults, on other locations, and also in PWS as part of complex syndromes such as Klippel–Trénaunay (1, 2, 3, 5). Patients have usually presented with scaling, pruritic, and inflammatory lesions that have tended to occur solely or most severely within the borders of the PWS. Commonly these lesions affect the periphery of the PWS most intensely (7). In the largest case series so far, PWS-associated dermatitis has been documented in children both with and without a history of atopic dermatitis (7). Treatment with topical corticosteroids is usually successful in eliminating eczema, but recurrences may ensue over the following years (8). Treatment of PWS with pulsed dye laser has been reported to produce both lightening of the PWS and improvement or resolution of the associated dermatitis (1, 6, 7). However, in rare instances, laser (pulse dye or KTP) therapy may trigger eczematous dermatitis over an otherwise normal-appearing PWS (4, 9, 10). Our two patients differ in possible contribution of laser treatment to the development of Meyerson phenomenon within their PWS: although initial laser treatment lightened the nevi in both, in the first case

Figure 3 | a) PWS on the chin and left cheek at age 2; b) PWS prior to laser treatment; c) and d) 1 week after the third laser treatment: tiny pruritic erythematous papules on the chin and almost complete disappearance of PWS.
laser-induced inflammation completely subsided over a 2-month period prior to the appearance of dermatitis. In the second patient, however, the inflammatory reaction seemed to be triggered by the laser treatment.

The cause of Meyerson phenomenon in PWS is not known despite several hypotheses, including a proinflammatory environment in the setting of ectatic postcapillary vessels (2, 5, 7) or possible genetic mosaicism underlying nevi (5). PWS follows the patterns of embryonic vascular development, although formal proof for a mosaic defect is lacking (11). Clearly, inflammation triggered by laser treatment may be the cause in some patients. Eczematous inflammation starting with PWS may also later spread to other sites even after resolution of the original dermatitis, as illustrated in our first patient.

Although we do not yet know the precise cause of dermatitis within PWS, it seems that it is a benign, self-limiting inflammation easily controlled by topical steroids and, in the long term, nevus clearance by laser light may completely prevent its occurrence.

References