Unilateral Acquired Nevus Flammeus in Women

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Congenital nevus flammeus is a benign vascular tumor characterized by pink to pale red patches that thicken as the patient ages, producing a dull red to reddish blue, cobblestone-textured plaque. We present the cases of 3 women with unilateral acquired nevus flammeus on the cheek whose lesions resolved after minimal treatment with a 585-nm pulsed dye laser. The etiology of acquired nevus flammeus is reviewed and tumor response rates to laser surgery are discussed.

Congenital nevus flammeus is a benign vascular tumor characterized by pink to pale red patches. These patches thicken as the patient ages, producing a dull red to reddish blue, cobblestone-textured plaque. Ectatic blood vessels, which initially lie in the superficial dermis, progress to involve the reticular dermis and subcutaneous layer.[1] Congenital nevus flammeus is classified as either medial or lateral. Medially located nevus flammeus (eg, stork bite, salmon patch, angel kiss) occurs in the occipital region or on the center of the face. Laterally located nevus flammeus (eg, port-wine stain) occurs on the unilateral face or on one or several extremities.[1] Unlike congenital nevus flammeus, acquired nevus flammeus is a vascular tumor that occurs later in life and is sometimes associated with trauma or hormonal changes. We present the cases of 3 women with acquired nevus flammeus who responded rapidly to treatment with a 585-nm pulsed dye laser.

Case Reports

Three patients presented with an asymptomatic, erythematous patch with an irregular margin on one cheek. All denied trauma prior to onset. The average ages at the onset of acquired nevus flammeus and the start of laser surgery were 31 and 40 years, respectively. One patient initially noticed her lesion during the sixth month of pregnancy. A 39-year-old woman presented with an irregular red patch on the right cheek (Figure 1). After 3 treatments with a pulsed dye laser, the acquired nevus flammeus resolved, 14 years after its initial onset (Figure 2). The clinical characteristics of our patients and the results of laser treatment are summarized in Table 1. After 1 to 3 treatments with a pulsed dye laser, all of the lesions cleared. A comparison of the results from our study patients and those from published cases of acquired nevus flammeus is found in Table 2.

Figure 1. A 39-year-old woman has an irregular, 5×4 cm red patch on her right cheek that has been worsening for 14 years.

Comment

Mechanical and thermal traumas have been implicated in the etiology of acquired nevus flammeus.[2-4] Huh et al[5] reported 2 patients with localized facial telangiectasia following frostbite. Genetics, hormonal alterations,[6] solar damage, and unidentifiable sources also have been reported as possible causes.

None of our patients had a history of mechanical trauma or thermal injury, including chronic sun

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Table 1.

Unilateral Acquired Nevus Flammeus: Clinical Summary*

Age, y/Gender	Location	Size, cm	Duration, y	Description	Treatment	Result
36/F	Left cheek	3×3.5	8	Erythematous patch with irregular margin	PDL 7.1 J/cm^2×1	Resolved
39/F	Right cheek	5×4	14	Discontinuous, erythematous macules with irregular margins	PDL 7.0 J/cm^2×3	Resolved
47/F	Left cheek	7×4	10	Erythematous patch with irregular margin	PDL 7.1 J/cm^2×2	Resolved

*F indicates female; PDL, pulsed dye laser.

Figure 2. The acquired nevus flammeus resolved after 3 treatments with a 585-nm pulsed dye laser.

exposure, but one developed acquired nevus flammeus on the left cheek during her sixth month of pregnancy. The other 2 patients denied pregnancy or the use of hormonal medications, including birth control pills. In Brinkman's[11] report of 2 women with acquired nevus flammeus, one patient developed a lesion during puberty and the other developed one during pregnancy. Goldman[7] reported acquired vascular abnormalities in 8 patients, 7 of whom were women, (4 of whom used oral contraceptives for a period of 1 to 3 years).

A number of cases of acquired nevus flammeus without a causative factor have been reported.[4,8] Two of our 3 patients fit into this category. Traub[10] described a 28-year-old man who developed acquired nevus flammeus on the right cheek without an identifiable cause. The largest series of idiopathic acquired nevus flammeus was 8 cases reported by Dinehart et al.[6]

A cause for acquired nevus flammeus has been speculated and proven in some instances. A decreased perivascular nerve innervation in acquired nevus flammeus was reported by Smoller and Rosen[12] who hypothesized a maturation defect in sympathetic innervation,[13] which results in continued unregulated blood flow and predisposes the cutaneous blood vessels to progressive ectasia.[14] Lanigan and Cotteril[15] reported a reduction in vasoactive response in nevus flammeus to vasodilating and vasoconstricting stimuli. Tsuji and Sawabe[3] theorized an abnormal repair of vessels following trauma, which results in dilation and impairs contractility of the vessel wall and atrophy of the dermis. Therefore, poor support for the vessels causes dilation and telangiectasia. Increased estrogen levels causing increased blood vessel proliferation is the hypothesis for pregnancy- and birth control pill–induced acquired nevus flammeus.[10]

Congenital nevus flammeus responds less rapidly and less completely to laser surgery than acquired nevus flammeus.[6] Ashinoff and Geronemus[16] reported that 10 of 12 infants aged between 6 and 30 weeks who had port-wine stains showed more than 50% lightening after 2.9 pulsed dye laser treatment sessions (range, 1–3 treatment sessions). Forty-five percent of their patients revealed a 75% or more lightening of their lesions after a mean of 3.8 treatments with a pulsed dye laser. After an average of 2.5 treatments in 33 patients, Reyes and

Table 2.

A Comparison of the Present Study With Published Cases of Unilateral Acquired Nevus Flammeus*

Source	Characteristics						
	Unilateral	Patients, No.	Women, No. (%)	Average Age, y	Average Age of Onset, y	Cause, No.	Treatment
Present study	Yes	3	3 (100)	41	31	Pregnancy, 1 Unknown, 2	PDL
Colver and Ryan[2]	Yes	1	0	NA	NA	NA	NA
Tsuji and Sawabe[3]	Yes	3	2 (66)	42	40	Trauma, 3	None
Pasyk[4]	Yes	4	3 (75)	37	22	Trauma, 1 Ultraviolet radiation, 2	PDL
Huh et al[5]	Yes	2	1 (50)	49	36	Frostbite, 2	None
Dinehart et al[6]	Yes (8/10)	10	5 (50)	54	16	Birth control pills, 1 Unknown, 4	Pulsed dye copper vapor
Goldman[7]	Yes (5/7)	7	7 (100)	31	31	Birth control pills, 4 Pregnancy, 2 Conjugated estrogen, 1	None
Cobb and Goldman[8]	Yes	1	0	NA	NA	NA	NA
Hogan and Rooney[9]	No	1	1 (100)	44	44	Steroid	None
Traub[10]	Yes	1	0	NA	NA	NA	NA

*PDL indicates pulsed dye laser; NA, not applicable.

Geronemus[17] demonstrated more than 75% lightening of congenital port-wine stains in patients between the ages of 3 months and 14 years. They also reported a 50% to 74% lightening after an average of 1.7 treatments in 31 of 73 patients. Overall, the average lightening after one treatment was 53%. Reyes and Geronemus concluded that the percentage of lightening increased as the number of treatments increased. Dinehart et al[6] also demonstrated excellent lightening results in acquired

port-wine stains after 1 to 3 treatments with pulsed dye laser.

Several lasers that have successfully been used in treating congenital vascular lesions include copper vapor, argon, argon-pumped tunable dye, krypton, Nd:YAG, and 585-nm pulsed dye laser.[18] The pulsed dye laser produces the least scarring because it uses selective photothermolysis. All of the patients in our study responded with resolution of unilateral acquired nevus flammeus after 1 to 4 pulsed dye laser treatments.

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