Five Cases of Acquired Port-Wine Stains

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Acquired port-wine stain is rare. It mimics a congenital port-wine stain morphologically but the pattern of onset is acquired after birth. It represents a progressive ectasia of vessels located in the superficial vascular plexus. The exact mechanism is unknown but some reported cases have occurred after trauma. We herein report five cases of acquired port-wine stains and they were idiopathic in nature.(Ann Dermatol 13(3) 163~166, 2001).

Key Words : Acquired onset, Port-wine stain

Port-wine stain is a well-known congenital lesion consisting of progressive ectatic blood vessels located in the papillary and upper reticular dermis. It is usually present at birth, grows proportionately with aging, and has no tendency for regression. The color gradually deepens with time, becoming dark red during adolescence and violaceous during middle age. In addition, the surface of a port-wine stain becomes raised and nodular as the patient ages.

Rarely, port-wine stains are acquired, but their morphology is identical to a congenital port-wine stain. We report herein five cases of acquired port-wine stains which show somewhat favorable responses to pulsed dye laser therapy.

CASE REPORTS

Case 1

A 23-year-old-woman had a discoloration of the upper cutaneous lip that began as a small red spot approximately 8 years ago. As the patient aged, the lesion became more apparent and slowly enlarged. Physical examination revealed an asymptomatic relatively well-defined 1 × 2 cm-sized deep red patch on the upper cutaneous lip. Precipitating factors, such as history of prior trauma to the area, medication, were unclear and there was no family history of nevus flammeus. Histologic examination was not performed because of its small size. Clinical diagnosis of acquired port-wine stain was made by its morphology and history. She received three treatments with a flashlamp-pumped pulsed dye laser (Candela ScleroPlus, U.S.A.) and showed lightening of the lesions with 50% improvement evaluated both by clinician and patient.

Case 2

A 36-year-old-woman had a fifteen-year history of red patches on the face. She described an occasional pruritus of the lesions. She denied any history of trauma or drug medication. On examination, there were several 2 × 2 cm-sized bright red patches with pinhead sized erythematous papules around the mouth and nose and on the forehead. Histologic examination showed ectatic capillary vessels on the upper dermis with no congestions, consistent with port-wine stain. The diagnosis of acquired port-wine stain was made. She is undergoing laser treatment and shows lightening of about 50% after 3 treatments.

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Case 3

A 15-year-old-woman had a discoloration of her right upper eyelid that appeared approximately 9 months ago. There was no history of trauma to the area. Physical examination showed an asymptomatic 3×4 cm-sized red patch on the right upper eyelid. Histologic examination was not done on patient's refusal but clinical diagnosis of acquired port-wine stain was made by its morphology and history. She has received five treatments with a flashlamp-pumped pulsed dye laser with lightening of about 60% and treatment is in progress.

Case 4

A 30-year-old-woman complained of a longitudinal discoloration on the forehead for 6 months. There was no history of trauma, pregnancy, and medication when it began to appear. No family member had any similar lesions. On examination, there was an asymptomatic longitudinal 1×3 cm-sized red patch with pinhead sized erythematous papules and telangiectasia on the forehead and left inner canthus (Fig. 1). A skin biopsy revealed ectatic capillary vessels in the whole dermis, consistent with port-wine stain (Fig. 2). The diagnosis of acquired port-wine stain was made. The lesion has been treated with a flashlamp-pumped pulsed dye laser and shows lightening of 50% after 3 treatments.

Case 5

A 29-year-old-man presented with a pink-red macule on his right cheek which had been present for about 16 years. He had no history of preceding trauma on the lesion. The lesion gradually became more apparent and enlarged slowly. On examination, there was an asymptomatic 3×3 cm-sized pink-red macule on the right cheek. A skin biopsy showed increased numbers of multiple thin-walled vessels on the upper dermis, consistent with port-wine stain. The diagnosis of acquired port-wine stain was made. He received treatments with a flashlamp-pumped pulsed dye laser and had a complete clearance of the lesion after 4 treatments.

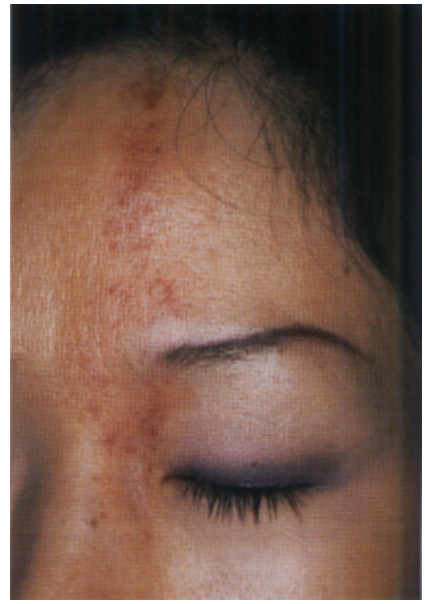

Fig. 1. Longitudinal 1×3 cm sized telangiectatic red patch with pinhead sized erythematous papules and telangiectasia on the forehead and left inner canthus (case 4).

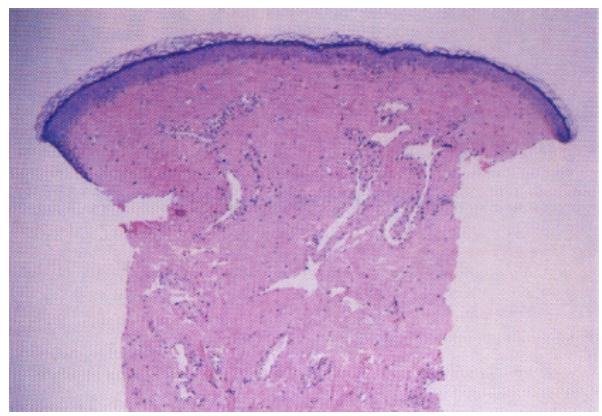

Fig. 2. There are thin walled dilated vessels in the whole dermis (case 4, H&E, ×100).

DISCUSSION

Port-wine stain, a type of capillary malformation, occurs in 0.3% of newborns as a red to purple macular, usually unilateral lesion with predilection for the face[1]. It tends to gradually deepen with time and grow proportionately with the child. In adulthood, it becomes raised and nodular.

But the acquired case of port-wine stain could be, and is unusual. In 1939, Traub[2] reported the first case of an acquired port-wine stain. He presented a 28-year-old man with a port-wine stain of the right cheek present for five years. The ac-

quired port-wine stain, like the congenital port-wine stain, results from a passive ectasia of vessels in the papillary and upper reticular dermis rather than an active proliferation as seen in hemangioma and pyogenic granuloma[3]. Clinically, it shows grouped unilateral telangiectasias, a confluent erythematous patch or group of red macules. In our cases the time of onset was confirmed by family members or by their photographs and the histopathologic examinations of these acquired port-wine stains revealed findings similar to those of the congenital ones.

The exact pathogenesis of port-wine stain remains unclear. Barsky et al.[4] believed that collagen degeneration of the dermal structures supporting the vessels was involved in this progressive ectasia. Recently, it is also thought to be related to a marked decrease of the perivascular nerve elements, which was reported by Rosen and Smoller[5] and an impaired vasoactive response of capillary vessels to both vasodilating and vasoconstricting stimuli demonstrated by Lanigan and Cotterill[6]. Similarly, acquired port-wine stain may be due to trauma to these perivascular neural elements resulting in an ectasia of dermal vessels. Colver and Ryan[7] documented the development of a port-wine stain in an adult after a cricket ball injury to the face. According to Lanigan[3], among nineteen patients with acquired port-wine stain, six were associated with preceding trauma. Hormonal changes are also important. Estrogen has been considered as a causative factor in its development. In 1970, Goldman[8] described four of seven women, who had developed a lesion, had taken oral contraceptive pills for 1-3 years. Brinkmann[9] also reported two cases of acquired port-wine stain in females which occurred during pregnancy and puberty. Dinehart et al.[10] reported one of ten patients with acquired port-wine stain who had a history of trauma and one with taking oral control pills when the stain developed. Besides trauma[3,7,10] and hormone[8,9,10], thermal[11] and solar damage[12,13] may contribute to the development of it. Niemand-Anderssen[11] reported the slow development of a port-wine stain in a 26-year-old man after exposure to intense cold. Pasyk[12] suggested chronic actinic damage as a precipitating factor of lesion development in two patients and Horiuchi[13] reported a similar case as well. In our cases, one case (Case 4) showed no association with preceding trauma, medication, pregnancy and other events before it began to appear. However, in the other four cases of women (Case 1, 2, 3, 5), the lesions developed during their puberty with their association with the hormonal changes such as estrogen.

Although many therapeutic modalities have been introduced to treat port-wine stains, pulsed dye laser therapy has become the treatment of choice. Dinehart et al.[10] described that the patients with acquired port-wine stain responded better to pulsed dye laser therapy than those with the congenital one and Lanigan[3] supposed that it was due to its telangiectatic nature, relatively sparse number and superficial site of blood vessels in the acquired one. We treated five cases of acquired port-wine stains with pulsed dye laser which produces light at 595 nm. They have responded well to laser therapy and the treatment is in progress. The assessment of response was made subjectively both by clinician and patients based on a combination of percentage lightening color and reduction of size compared with pretreatment color photographs.

We report five cases of idiopathic acquired port-wine stains. Closer and wider studies are needed to explain the mechanism of acquired type stains in the future.

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