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Case Report

Acquired port-wine stain in an adult female: A rare entity

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ABSTRACT

The port-wine stain in adults, or acquired port-wine stain, is a rare entity. A fifty-year-old female presented with complaints of an asymptomatic erythematous patch affecting the right side of the face for the previous twenty years. The lesion gradually increased in size to involve the area below the eye, right nasolabial fold, cheek adjacent to the nasolabial fold, and area between the right nasolabial fold and the upper lip. On the basis of clinical and dermoscopic examinations, the diagnosis of a port-wine stain was established.

Key words: Acquired port-wine stain, Female sex, Congenital nevi, Dermoscopy

INTRODUCTION

Port-wine stains (PWS) are congenital, telangiectatic nevi consisting of ectatic dermal capillaries, affecting 0.3–1% of newborn infants. In contrast, acquired PWS develop later in life yet are morphologically identical to the congenital port-wine stain. Herein, we describe a case of acquired PWS.

CASE REPORT

A fifty-year-old female presented to the outpatient department of dermatology of a tertiary-care hospital with complaints of an asymptomatic erythematous patch affecting the right side of the face. She noticed it twenty years ago as a patch on the right side of the nose near the medial canthus of the eye (Fig. 1a). The lesion gradually increased in size to involve the area below the eye, right nasolabial fold (Fig. 1b), cheek adjacent to the nasolabial fold, and area between the right nasolabial fold and the upper lip. She denied having a history of a preceding birthmark. There was no history of antecedent mechanical or thermal trauma, drug intake, topical application, or excessive UV exposure.

On examination, the lesions were in the form of macular erythema. A dermoscopic examination revealed red, rounded, globular vessels (Fig. 2).

On the basis of clinical and dermoscopic examinations, the diagnosis of a port-wine stain was established. Laser therapy with pulse dye laser was discussed with the patient, yet she was reluctant to undergo any treatment at this age.

DISCUSSION

Port-wine stains are congenital, vascular malformations present at birth as pinkish-red to purple macules, which may become darker and nodular in adult life [1]. Biopsy specimens reveal an increase in vessel abnormalities with advancing age, which may be due to collagen degeneration and elastosis leading to the weakening of supporting dermal structures [2]. Rosen et al. implicated malformed sympathetic innervation in the pathogenesis of the port-wine stain and failure to regulate blood flow resulting the in progression of vascular injury [3]. Rydh et al. reported a lack of neural innervation in the port-wine stain, along with sympathetic innervation [4]. A study by Breugem et al. revealed the genetic inheritance of the port-wine stain as an autosomal dominant disorder, whose locus is mapped on chromosome 5q [5].

Unlike the congenital port-wine stain, the acquired port-wine stain occurs later in life. Various factors have been proposed for the occurrence of port-wine stains.

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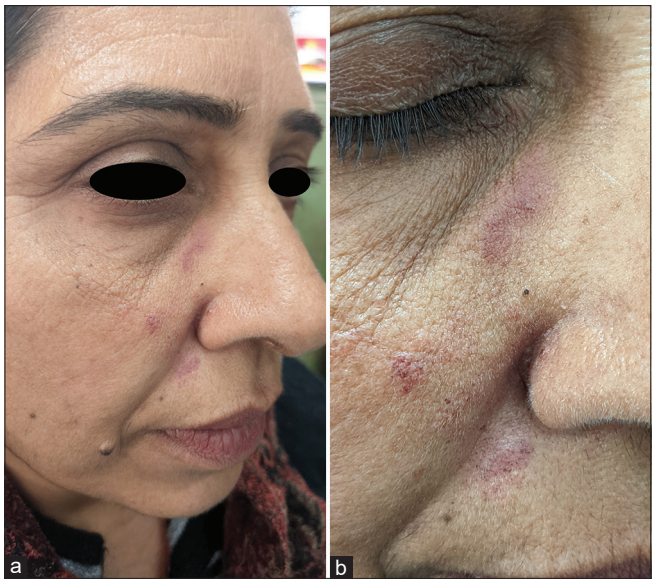

Figure 1: (a) Acquired port-wine stain in the fifty-year-old female involving the right side of the face. (b) Close-up view of the acquired port-wine stain.

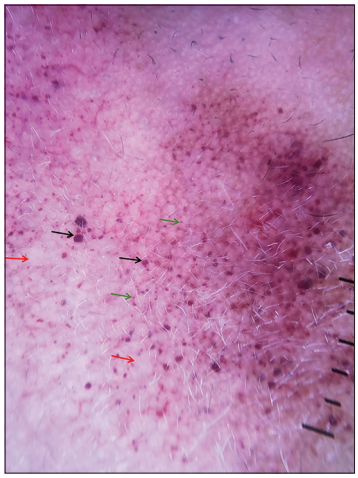

Figure 2: Dermoscopy of the acquired port-wine stain: white veils (*red arrows*), red dots (*green arrows*), globules (*black arrows*).

Post-traumatic capillary malformation may be seen, first described by Fegeler, and hence called Fegeler syndrome [6]. Other proposed etiologies are chronic sun exposure [7] and drugs such as isotretinoin [8] and OCPs [9].

On dermoscopy, findings such as dots, globules, linear vessels, reticular vessels, and whitish veils may be seen [10]. Dermoscopic features help to predict the response to laser therapy. Dots and globules represent the superficial or papillary form of the port-wine stain, which responds better to laser therapy when compared to a pattern showing linear vessels, which is the deep or subpapillary form.

Thus, dermoscopy serves as an important tool for assessing the depth of port-wine stains, as dotted or globular vessels indicate the superficial form and linear vessels indicate the deep form of the port-wine stain. Hence, the response to treatment may be assessed with dermoscopy. The acquired port-wine stain is rare and few cases have been reported in the literature so far.

Consent

The examination of the patient was conducted according to the principles of the Declaration of Helsinki.

The authors certify that they have obtained all appropriate patient consent forms, in which the patients gave their consent for images and other clinical information to be included in the journal. The patients understand that their names and initials will not be published and due effort will be made to conceal their identity, but that anonymity cannot be guaranteed.

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