

SHORT COMMUNICATION

Acquired Port-wine Stain with Local Heat

Shiang-Yu Yang¹ and Cheng-Che E. Lan^{1-3*}

Departments of Dermatology, ¹Kaohsiung Medical University Hospital, No. 100, Tzyou 1st Road, Kaohsiung 807, ²College of Medicine, Kaohsiung Medical University, and ³Kaohsiung Municipal Ta-Tung Hospital, Kaohsiung Medical University, Kaohsiung, Taiwan. *E-mail: laneric@cc.kmu.edu.tw
Accepted Jun 25, 2015; Epub ahead of print Jun 30, 2015

Port-wine stains (PWSs) are usually considered to be congenital vascular malformations characterized by dilatation of capillaries in the papillary and upper reticular dermis with normal endothelial cells. They often present as well-defined, pinkish erythematous to purple macules at birth, which become darker and elevated over time. Most lesions are located on the face or neck and may follow the distribution of the trigeminal nerve. “Never warm on palpation” is generally considered an important diagnostic clinical feature of PWSs (1). Acquired PWSs are rare skin lesions that are morphologically and histopathologically identical to congenital PWSs (2). We report here a patient with acquired PWS on the left side of her face and neck with local heat, and propose that skin temperature is not a reliable diagnostic feature for identifying PWSs.

CASE REPORT

A 46-year-old woman presented with erythematous to violaceous, slowly enlarging macules with telangiectasia over her left cheek and neck for one year (Fig. 1a). The lesions had slowly enlarged and became more obvious. Mild warmth and swelling were noted during physical examination with no other associated symptoms. No obvious induration or elevation of the lesions was noted. The woman had previously been diagnosed with rosacea and had received topical treatment. However, her symptoms progressed and she was referred to our hospital for further evaluation.

The woman reported no systemic disease and took no medications before the onset of her skin lesions. In addition, she denied antecedent trauma to the left side of her face and reported no preceding birthmark during the first 45 years of her life. There was no family history of PWSs. Skin biopsy showed ectatic blood vessels lined by single flat endothelial cells in

the upper to middle dermis. Scattered perivascular infiltration of lymphocytes was also observed (Fig. 2a). The pathological features are consistent with PWS.

To document the perfusion profile of the lesion, Lisca PIM II Laser Doppler Perfusion Imager (Lisca Development AB, Linköping, Sweden) was used to evaluate blood flow in the woman's face. Laser Doppler perfusion imaging (LDPI) is a non-invasive method of obtaining a two-dimensional map of skin perfusion. Photography of the user-specified region of interest (ROI) identified increased blood flow around the lesion on our patient's left cheek. Blood flow was increased even in the region with no obvious clinical manifestation (Fig. 1b).

Although PWS has classically been described to be “never warm on palpation”, warmth was detected on our patient's lesions during physical examination. Skin temperature and total local microcirculatory blood perfusion were documented using laser Doppler perfusion measurement (LDPM) (PeriFlux System 5000; Perimed, Stockholm, Sweden). The patient in this case had a higher temperature and elevated blood perfusion on the left side of her face and neck compared with the right side. More specifically, the skin temperatures on the patient's right cheek, lower face, and neck were 32.1°C, 31.4°C and 31.9°C, respectively, while the skin temperatures on the corresponding left side were 33.5°C, 34.4°C and 34.5°C, respectively. Similarly, the blood perfusion (arbitrary unit) on the patient's right cheek, lower face, and neck were 36.1, 30.9, and 12.4, respectively, while the blood perfusion on her corresponding left side were 393.4, 138.5, and 101.8, respectively.

Haemangioma simplex should also be considered in middle-aged patients with elevated erythematous lesions. Although our patient had mild swelling of the left side of her face, she did not present with obvious induration or elevation of the lesions, and therefore, clinically, haemangioma simplex was unlikely to be the diagnosis. In addition, the histopathology of haemangioma simplex is characterized by an intradermal or subcutaneous multi-lobular proliferation of numerous small vascular spaces lined by plump endothelial cells. In our patient, only dilatation of capillaries in the papillary and upper reticular dermis with normal endothelial cells was noted microscopically in the biopsy specimen. Therefore, neither the clinical nor the histopathological findings favoured a diagnosis of haemangioma. Based on the increased temperature of the patient's lesion, a diagnosis of arteriovenous malformation (AVM) was considered. However, orcein immunohistochemical staining did not reveal any elastic fibres around the ectatic vessels in the biopsy specimen (Fig. 2b), thus the diagnosis of AVM was excluded.

Pulsed dye laser was suggested as a treatment option, but the patient decided to defer therapy.

DISCUSSION

The aetiology of PWS is unknown. Smoller & Rosen (3) hypothesized that a decrease in perivascular nerve function in PWS may cause a lack of neural modulation of vascular tone, leading to progres-

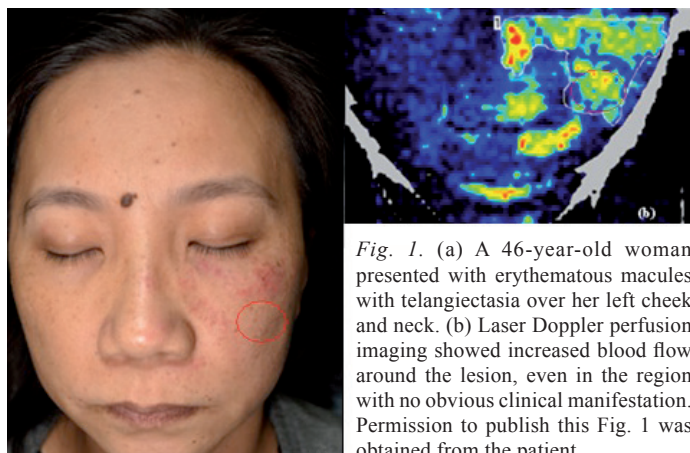


Fig. 1. (a) A 46-year-old woman presented with erythematous macules with telangiectasia over her left cheek and neck. (b) Laser Doppler perfusion imaging showed increased blood flow around the lesion, even in the region with no obvious clinical manifestation. Permission to publish this Fig. 1 was obtained from the patient.

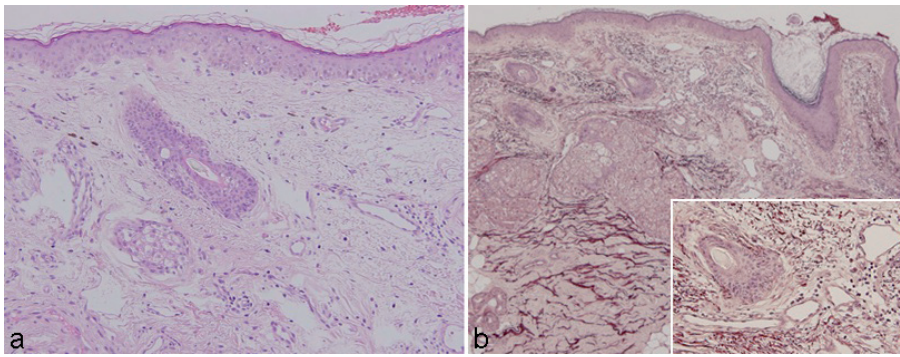


Fig. 2. (a) Histopathology showed ectatic blood vessels in the upper-to-middle dermis and scattered perivascular infiltration of lymphocytes (haematoxylin and eosin, original magnification 100 \times). (b) Immunohistochemical staining showed no elastic fibres around the ectatic vessels in the biopsy specimen (orcein, original magnification 40 \times). Inset (200 \times).

sive ectasia of vessels, which could play a role in the development of PWS. A significant reduction in the vasoactive response in PWS has been reported, which further supports this hypothesis (4).

Acquired PWSs are quite rare. The first report was published by Traub (5) in 1939. Adams & Lucky (6) reviewed 59 reported cases of acquired PWSs and found that 17 (27%) were related to trauma. Many reports suggest that acquired PWS is attributed to other causes, including oral contraceptive pills (7), chronic actinic exposure (8), oral isotretinoin (9), and cluster headache (10). However, as in the case reported here, most cases of acquired PWS have not been associated with a definite cause.

LDPI is a non-invasive technique that can provide a two-dimensional map of skin perfusion based on dynamic light scattering in tissue. While the laser beam scans the tissue sequentially, moving blood cells generate Doppler components in the back-scattered light (11). Troilius & Ljunggren (12) reported that 15 of 19 patients had an increased blood flow within the PWS, in comparison with normal contralateral skin as measured with LDPM, and the blood flow did not correlate well with the photometrically registered erythema. In parallel, an increased blood flow was detected within the PWS, even in regions with no clinical visible erythema. This situation will certainly increase the difficulty of treatment, and indicates that using blood flow imaging for pre-laser therapy planning and subsequent follow-up is necessary to ensure treatment success.

Because local heat was found on the PWS area in our patient during physical examination, we performed LDPM on her bilateral face and neck. This examination showed a higher temperature on her left side, which supported our clinical finding. To exclude a diagnosis of AVM, we investigated the expression of orcein by immunohistochemistry that showed no elastic fibres around the dilated vessels of the upper dermis. This finding demonstrates that the local temperature of PWSs could be elevated, and that the classical description of “never warm on palpation” regarding PWS is not accurate and should be modified. However, the documentation of this patient’s course of illness is relatively short, and longer follow-up with additional angiography may completely eliminate the possibility of AVM.

In summary, we describe here a case of a middle-aged woman with telangiectasia and local heat on her left cheek and neck. She was diagnosed with acquired PWS after clinical, histopathological and immunohistochemical evaluation. LDPI showed an increased blood flow within the affected skin, but the blood flow did not correlate well with the clinically observed erythema. In addition, although PWS is generally thought to be “never warm on palpation”, skin physiology examination of this patient clearly demonstrates that skin temperature is not a reliable diagnostic tool for identifying PWS.

The authors declare no conflicts of interest.

REFERENCES

- Boon LM, Vikkula M. Vascular malformations. In: Goldsmith LA, Katz SI, Gilchrist BA, Paller AS, Leffell DJ, Wolff K, editors. Fitzpatrick’s dermatology in general medicine. 8th edn. New York City: McGraw-Hill Education, 2012: p. 2076–2094.
- Dinehart SM, Parker RK, Herzberg AJ, Pappas AJ. Acquired port-wine stains. *Int J Dermatol* 1995; 34: 48–52.
- Smoller BR, Rosen S. Port-wine stains. A disease of altered neural modulation of blood vessels? *Arch Dermatol* 1986; 122: 177–179.
- Lanigan SW, Cotterill JA. Reduced vasoactive responses in port wine stains. *Br J Dermatol* 1990; 122: 615–622.
- Traub EF. Naevus flammeus appearing at the age of twenty three. *Arch Dermatol* 1939; 39: 752.
- Adams BB, Lucky AW. Acquired port-wine stains and antecedent trauma: case report and review of the literature. *Arch Dermatol* 2000; 136: 897–899.
- Goldman L. Oral contraceptives and vascular anomalies. *Lancet* 1970; 1: 108–109.
- Pasyk KA. Acquired lateral telangiectatic nevus: port-wine stain or nevus flammeus. *Cutis* 1993; 51: 281–283.
- Hoque S, Holden C. Acquired port wine stain following oral isotretinoin. *Clin Exp Dermatol* 2005; 30: 587–588.
- Vagefi MR, Florell SR, Lin CC, McCann JD, Anderson RL. Acquired port wine stain of the upper eyelid after cluster headache. *Ophthal Plast Reconstr Surg* 2008; 24: 490–492.
- Wårdell K, Jakobsson A, Nilsson GE. Laser Doppler perfusion imaging by dynamic light scattering. *IEEE Trans Biomed Eng* 1993; 40: 309–316.
- Troilius AM, Ljunggren B. Evaluation of port wine stains by laser Doppler perfusion imaging and reflectance photometry before and after pulsed dye laser treatment. *Acta Derm Venereol* 1996; 76: 291–294.