DERMOSCOPY FEATURES OF PORT-WINE STAINS: THE FIRST STUDY OF 148 CASES IN VIET NAM

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SUMMARY

Introduction: Port-wine stains (PWS) are capillary malformation affecting 0.3% to 0.5% of the population. In Vietnam, the dermoscopic features of PWS have not been systematically studied.

Objectives: The aim of this study was to analyze the dermoscopic images of 148 patients with PWS.

Methods: 148 cases with PWS were analyzed basing on dermascopic images.

Results: The most common vascular images were dots or globules vessels (76.4%) and linear vessels (68.9%). Sausage-like vessels and whitish veils were more common in the group of patients aged older than 18 years or with purple type of PWS. For all the dermoscopic features of PWS, excluding linear vessels, there were no significant differences between lesions located in the central facial area and peripheral facial area (p > 0.05). Compared with the peripheral facial lesions, linear vessels were more common in central facial lesions (p < 0.05).

Conclusion: Dermoscopy may provide very useful clinical information before laser treatment of PWS. The vascular manifestations of PWS determined by dermoscopy may correlate with age, colour, location and response to laser treatment.

Keywords: *Dermoscopy, port-wine stains.*

1. INTRODUCTION

Port-wine stain (PWS) is a congenital and progressive capillary malformation reported in 0.3% - 0.5% of newborn infants globally[1]. The appearance of PWS is initially as flat macules, which can range from pale pink to red and then purple in appearance with varying degrees of hypertrophy or nodule formation then occurring with age[2]. Although the pulsed-dye laser (PDL) has been the gold standard for treating PWS, only 10% - 20% of affected patients achieve complete blanching after laser treatments and recurrences are frequent[3-6]. Dermoscopy is a useful diagnostic tool to help visualize blood vessels, which is valuable in predicting the response to treatment of PWS with PDL[7]. We conducted the study to determine the dermoscopic features and the differences among patients with PWS.

2. OBJECTIVES AND METHODS

1.1. Objectives

148 patients with PWS were submitted to the National Hospital of Dermatology and Venereology from August 2020 to September 2021.

1.1.1. Criteria for selecting patients

- Patients diagnosed with port-wine stains.

- Patients agreed to participate in the study (their parents or guardians if they were under 18 years old).

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1.1.2. Exclusion criteria: Patients were treated by laser and other therapies (surgery, cryotherapy, electrocautery…) prior to study.

1.2. Methods

2.2.1. Study design: Analytical cross-sectional study

2.2.2. Study description

- Preparing the medical records for the study.

- Examining and evaluating clinical symptoms.

- Patients underwent dermoscopy (Heine Delta 20 T, Germany, x 20, polarised) and clinical photography (Canon 750D).

- Five dermoscopic images from different fields of the lesion were captured for further analysis.

- Four dermatologists viewed the dermoscopy images and classified the vascular patterns, the pattern that was dominant in the most of the pictures was chosen. Images of the same patient were classified as the dominant vascular type (the vascular pattern accounting for 80% of all images in the same PWS). If more than 80% of the images included two or more vascular patterns, they were defined as mixed.

- The vascular morphology includes dots or globules vessels, sausage-like vessels, linear vessels, reticular vessels, arborizing vessels and mixed vessels. Non-vascular morphology includes whitish veil and fibrosis.

- Data processing.

2.2.3. Statistical analysis

The data were statistically analyzed using SPSS 20.0 for Windows, version 10.0. A chi-square test was used to compare the frequency of the dermoscopic features among the groups. Fisher exact test was used when appropriate. All the p values were two-sided, and p values less than 0.05 were considered statistically significant.

3. RESULTS

3.1. Characteristics of the patients

A total of 148 patients with PWS were enrolled in our study. 100% cases never received the treatment.

Table 1. Distribution of PWS by age

Age (years)	n	%
≤ 5	44	29.7
6 - 17	54	36.5
≥ 18	50	33.8
Total	148	100
Average age (x ± s)	15.4 ± 13.6	

There were 91 (61.5%) females, with the ratio of males to females as 1:1.6. The average age was 15.4 ± 13.6 (range 0 - 69) years. There were 44 (29.7%) cases aged 0 - 5 years, 54 (36.5%) aged 6 - 17 years and 50 cases (33.8%) ≥ 18 years.

3.2. Dermoscopic features

Table 2. The characteristics of PWS on dermoscopy

Characteristic	n	%
Dots or globules vessels	113	76.4
Sausage-like vessels	50	33.8
Venous lakes	13	8.8
Linear vessels	102	68.9
Arborizing vessels	50	33.8
Reticular vessels	8	5.4
Mixed vessels	47	31.8
Whitish veil	43	29.1
Fibrosis	4	2.7

Dots or globules vessels (113 or 76.4%) and linear vessels (102, 68.9%) were the most frequent dermoscopic features of PWS. Other dermoscopic features were sausage-like vessels (50, 33.8%), arborizing vessels (50, 33.8%), mixed vessels (47, 31.8%), venous lakes (13, 8.8%), reticular vessels (8, 5.4%). Nonvascular morphology comprised whitish veil (43, 29.1%) and fibrosis (4, 2.7%).

Table 3. The characteristics of PWS by age

Characteristic		0 - 5 (n = 44)	6 - 17 (n = 54)	≥ 18 (n = 50)	p
Vascular morphology	Dots/globules (n = 113)	26 (59.1%)	45 (83.3%)	42 (84%)	0.006
	Sausage-like vessels (n = 50)	4 (9.1%)	18 (33.3%)	28 (56%)	0.0001
	Venous lakes (n = 13)	0 (0%)	2 (15.4%)	11 (84.6%)	0.001
	Linear vessel (n=102)	38 (86.4%)	33 (61.1%)	31 (62%)	0.012
	Arborizing vessels (n = 50)	21 (47.7%)	14 (25.9%)	15 (30%)	0.06
	Reticular vessels (n = 8)	5 (11.4%)	1 (1.9%)	2 (25%)	0.102
	Mixed vessels (n = 47)	11 (25%)	17 (31.5%)	19 (38%)	0.401
Non-vascular morphology	Whitish veil (n = 43)	1 (2.3%)	8 (14.8%)	34 (68%)	0.0001
	Fibrosis (n = 4)	0 (0%)	0 (0%)	4 (8%)	0.019

In patients of different ages, dots/globules, sausage-like vessels, venous lakes and whitish veil were common in those aged ≥ 18 years, linear vessels were common in < 18 years, and dots/globules vessels, linear vessels were common in those aged between 6 and 17 years. Fibrosis only appeared in patients older than 18 years.

Table 4. The characteristics of PWS by colour

Characteristics		Pink (n = 34)	Reddish (n = 43)	Purple (n = 71)	p
Vascular morphology	Dots/globules (n = 113)	17 (50%)	55 (77.5%)	41 (95.3%)	0.0001
	Sausage-like vessels (n = 50)	3 (8.8%)	14 (19.7%)	33 (76.7%)	0.0001
	Venous lakes (n = 13)	0 (0%)	0 (0%)	13 (30.2%)	0.0001
	Linear vessel (n = 102)	29 (85.3%)	53 (74.6%)	20 (46.5%)	0.0001
	Arborizing vessels (n = 50)	17 (50%)	27 (38%)	6 (14%)	0.002
	Reticular vessels (n = 8)	2 (5.9%)	5 (7%)	1 (2.3%)	0.640
	Mixed vessels (n = 47)	10 (29.4%)	23 (32.4%)	14 (32.6%)	0.945
Non-vascular morphology	Whitish veil (n = 43)	7 (20.6%)	8 (11.3%)	28 (65.1%)	0.0001
	Fibrosis (n = 4)	0	0	4 (2.7%)	0.009

Dots/globules vessels, sausage-like vessels, venous lakes and whitish veil were mainly present in the purple type of PWS (p < 0.05), whereas linear vessels and aborizing vessels were mainly present in pink type of PWS (p < 0.05). Whitish veil was more common in purple type lesions (p < 0.05). Venous lakes and fibrosis were only found in the purple type of PWS.

Table 5. The differences of dermoscopical features between central facial lesions and peripheral facial lesions

Characteristic		Central facial area (n = 38)		Peripheral facial area (n = 62)		p
		n	%	n	%	
Vascular morphology	Dots/globules	27	71.1	50	80.6	0.342
	Saussage-like vessels	14	36.8	21	33.9	0.720
	Venous lakes	6	15.8	6	9.7	0.346
	Linear vessels	31	81.6	37	59.7	0.018
	Arborizing vessels	15	39.5	18	29.0	0.258
	Reticular vessels	4	10.5	3	4.8	0.421
	Mixed vessels	16	42.1	17	27.4	0.117
Non-vascular morphology	Whitish veil	15	39.5	15	24.2	0.095
	Fibrosis	1	2.6	1	1.6	0.613

For all the dermoscopic features of PWS, excluding linear vessels, there were no significant differences between lesions located in the central facial area and peripheral facial area (p > 0.05). Compared with the peripheral facial lesions, linear vessels were more common in central facial lesions (p < 0.05).

4. DISCUSSION

PWSs are vascular malformations composed of postcapillary venules in the dermis. The distribution of vessel size and depth vary among patients and even within the same lesion[8]. The depth of the vessels is critical because the response to laser therapy decreases with deeper vessels, and this has been shown histologically in some studies. The diameter of the vessels is another very important factor that influences the response to laser therapy. As a rule, vessels with a small diameter show less response to PDL than larger vessels, especially if they are located deeply[9,10]. All of these vascular differences are reflected in the color of the lesion, i.e., pink lesions have small deep vessels, purple lesions have larger deep vessels, and red lesions have superficial vessels[9].

Dermoscopy is a helpful tool in distinguishing PWS from other vascular diseases such as telangiectasia, angioma serpiginosum and angiokeratoma; it also helps to predict the therapeutic effect[11]. However, there are no consensus dermoscopic criteria for the diagnosis of PWS. Initially, the dermoscopic features of PWS were divided into two types: blobs and rings. Then, mixed vessel was added as a third type. In recent years, further dermoscopic features of PWS have been added: dots and globules vessels, sausage-like vessels, short clubbed vessels, curved vessels, linear vessels, thin long parallel vessels, network of thin long vessels, arborising vessels, reticular vessels, mixed vessels, white circles, whitish veil,

fibrosis and 'peacock eyes'. Some of these may refer to the same feature, such as curved vessels and thin long parallel vessels, which may be characterised as linear vessels. Arborising vessels have rarely been mentioned and may consist of overlapping linear vessels[12].

The dots/globules vessels are less than 0.3 mm in diameter, also known as type I capillaroscopic pattern of PWS. Histopathology corresponds to dilated capillaries located superficially in the dermis, running perpendicular to the epidermal surface[13]. This is the vascular pattern that shows the best response to laser treatment[14]. In our study, the image of dots/globes vessels was mainly present at 76.4%. All patients in our study had not received any previous treatment. The dots or globules vessels in different subtypes or age of PWS showed significant difference. Dots/globules vessels, sausage-like vessels, venous lakes and whitish veil were mainly present in the purple type of PWS and in those aged ≥ 18 years, whereas linear vessels and aborizing vessels were mainly present in pink type of PWS. The dots or globules vessels in central and peripheral facial face showed no significant difference, but they were more common in peripheral facial lesions than central facial lesions. Kwiek et al[15] investigated the value of dermatoscopical types of vascular pattern for the prediction of PWS efficacy using large spot 532 nm laser, and they concluded that "dots", "globules", and "sausage-like" had the same prognostic value as "superficial vessels" for prediction of the efficacy of 532 nm laser.

Linear vessels, reticular vessels and aborizing vessels also known as type II pattern vessels of PWS, which exhibits deeper, ectatic, horizontal capillaries in the reticular dermis and are considered more difficult to remove than the type I pattern[8,11,14,16]. Combined with the PWS histopathology, arborizing vessels may be horizontal and are likely to be

located at varying and inconsistent depths in the dermis. This pattern suggests that only some malformed blood vessels will be within the scope of the penetration depth of the light source and that others may not be accessible[7]. The sausage-like vessels are thought to be superficial vessels, similar to dotted and globular vessels that also respond well to laser treatment[15]. The mixed vessels, reported as the type III capillaroscopic pattern by Bencini et al., exhibit combined features of both the type I and type II patterns[14]. They have the lowest response rateto PDL treatment when compared with type II and type III patterns[14]. Researchers have found a strong relationship between the type III capillaroscopic pattern and arterial hypertension and hypothesised that long-lasting haemodynamic abnormalities may play a role in the development of this pattern. In our study, linear vessels and reticular vessels were common in < 6 years or pink lesions. For all the dermoscopic features of PWS, excluding linear vessels, there were no significant differences between lesions located at central facial area and peripheral facial area. In previous biopsy studies[9,17], it was conjectured deeper and smaller vessels tend to respond poorly to laser treatment compared with large and superficial vessels. The difference in efficacy of PDL treatment between lateral and central PWS of the face may be a result of the variation in the heterogenous vessels, especially in the depth of the PWS vessels[18]. Linear vessels represent the deep, subpapillary form of PWS, showing horizontally oriented capillaries. Maybe, our results explain why peripheral facial lesion respond better than those located in the central face.

Nonvascular morphology such as whitish veil and fibrosis alse presented in our study. A whitish veil can be seen in many dermatosis including angiokeratoma[19], melanoma[20], and sebaceoma[21]. A whitish veil is associated with connective tissue hypertrophy, focal fibrosis of PWS and previous treatment, especially when it resulted in scarring. The whitish veil is the feature that has been proposed to be associated with the presence of deep vessels, when the whitish veil is present; ectatic vessels of PWS were mostly located in the lower part of the dermis[11]. Procaccini et al[11] proposed previously that a gray-whitish veil reflects malformed dilated blood vessels located in the deeper dermis. Other authors observed in another report on a series of PWS patients that mixed patterns produced a poorer therapy response to laser than superficial patterns. Kwiek et al[15] have consistently identified that the whitish veil is a poor prognostic type for PWS with large spot 532 nm laser.

In this study, the frequency of whitish veil was higher in the purple type, those aged ≥ 18 years. We agreed with Huang et al[12] that the whitish veil may mainly be associated with age, which is consistent with the finding of a statistically significant positive correlation between the presence of 'fibrosis' and a patient's age. The frequency of whitish veil in patients with prior treatment may be due to local scar formation[15]. However, we did not find a significant difference between central facial lesions and peripheral facial lesions.

5. CONCLUSION

In summary, there are a variety of dermoscopic features of PWS. The dermoscopic features of patients with PWS may correlate with age, colour, location and response to laser treatment. The capillaroscopic patterns of PWS should be considered when planning laser treatment of these lesions by using, for example, longer pulse durations and larger spot size, longer wavelengths, which are more penetrating in the presence of a type III pattern or whitish veil.

REFERENCES

1. Jacobs AH, Walton RG. The incidence of birthmarks in the neonate. *Pediatrics*. 1976;58(2):218-222.

2. van Drooge AM, Beek JF, van der Veen JPW, van der Horst CMAM, Wolkerstorfer A. Hypertrophy in port-wine stains: prevalence and patient characteristics in a large patient cohort. *J Am Acad Dermatol*. 2012;67(6):1214-1219. doi:10.1016/j.jaad.2012.05.027.

3. Woo WK, Handley JM. Does fluence matter in the laser treatment of port-wine stains? *Clin Exp Dermatol*. 2003;28(5):556-557. doi:10.1046/j.1365-2230.2003.01327.x

4. Huikeshoven M, Koster PHL, de Borgie CAJM, Beek JF, van Gemert MJC, van der Horst CMAM. Redarkening of port-wine stains 10 years after pulsed-dye-laser treatment. *N Engl J Med*. 2007;356(12):1235-1240. doi:10.1056/NEJMoa064329.

5. Savas JA, Ledon JA, Franca K, Chacon A, Nouri K. Pulsed dye laser-resistant port-wine stains: mechanisms of resistance and implications for treatment. *Br J Dermatol*. 2013;168(5):941-953. doi:10.1111/bjd.12204.

6. Lanigan SW. Port-wine stains unresponsive to pulsed dye laser: explanations and solutions. *Br J Dermatol*. 1998;139(2):173-177. doi:10.1046/j.1365-2133.1998.02351.x

7. Wang X, Suo H, Gao Y, et al. Correlation between the hemoporfin-mediated photodynamic treatment response and the dermoscopy vascular pattern in patients with a port-wine stain: a prospective study. *J Eur Acad Dermatol Venereol JEADV*. 2020;34(12):2795-2801. doi:10.1111/jdv.16596.

8. Eubanks LE, McBurney EI. Videomicroscopy of port-wine stains: correlation of location and depth of lesion. *J Am Acad Dermatol*. 2001;44(6):948-951. doi:10.1067/mjd.2001.113467.

9. Fiskerstrand EJ, Svaasand LO, Kopstad G, Dalaker M, Norvang LT, Volden G. Laser treatment of port wine stains: therapeutic outcome in relation to morphological parameters. *Br J Dermatol*. 1996;134(6):1039-1043.

10. Onizuka K, Tsuneda K, Shibata Y, Ito M, Sekine I. Efficacy of flashlamp-pumped pulsed dye laser therapy for port wine stains: clinical assessment and histopathological characteristics. *Br J Plast Surg*. 1995;48(5):271-279. doi:10.1016/0007-1226(95)90064-0.

11. Procaccini EM, Argenziano G, Staibano S, Ferrara G, Monfrecola G. Epiluminescence microscopy for port-wine stains: pretreatment evaluation. *Dermatol Basel Switz*. 2001;203(4):329-332. doi:10.1159/000051783.

12. Huang YC, Ringold TL, Nelson JS, Choi B. Noninvasive blood flow imaging for real-time feedback during laser therapy of port wine stain birthmarks. *Lasers Surg Med*. 2008;40(3):167-173. doi:10.1002/lsm.20619.

13. Vázquez-López F, Coto-Segura P, Fueyo-Casado A, Pérez-Oliva N. Dermoscopy of port-wine stains. *Arch Dermatol*. 2007;143(7):962. doi:10.1001/archderm.143.7.962.

14. Bencini PL, Cazzaniga S, Galimberti MG, Zane C, Naldi L. Variables affecting clinical response to treatment of facial port-wine stains by flash lamp-pumped pulsed dye laser: the importance of looking beyond the skin. *Lasers Med Sci*. 2014;29(4):1365-1370. doi:10.1007/s10103-014-1525-4.

15. Kwiek B, Rożalski M, Sieczych J, Paluch Ł, Kowalewski C, Ambroziak M. Predictive value of dermoscopy for the treatment of port-wine stains with large spot 532 nm laser. *Lasers Surg Med*. 2019;51(7):569-583. doi:10.1002/lsm.23083.

16. Motley RJ, Lanigan SW, Katugampola GA. Videomicroscopy predicts outcome in treatment of port-wine stains. *Arch Dermatol*. 1997;133(7):921-922. doi:10.1001/archderm.133.7.921.

17. Hohenleutner U, Hilbert M, Wlotzke U, Landthaler M. Epidermal damage and limited coagulation depth with the flashlamp-pumped pulsed dye laser: a histochemical study. *J Invest Dermatol*. 1995;104(5):798-802. doi:10.1111/1523-1747.ep12606996.

18. Yu W, Ma G, Qiu Y, et al. Why do port-wine stains (PWS) on the lateral face respond better to pulsed dye laser (PDL) than those located on the central face? *J Am Acad Dermatol*. 2016;74(3):527-535. doi:10.1016/j.jaad.2015.08.026.

19. Kim TH, Choi EH, Ahn SK, Lee SH. Vascular tumors arising in port-wine stains: two cases of pyogenic granuloma and a case of acquired tufted angioma. *J Dermatol*. 1999;26(12):813-816. doi:10.1111/j.1346-8138.1999.tb02098.x

20. Ronger-Savle S, Julien V, Duru G, Raudrant D, Dalle S, Thomas L. Features of pigmented vulval lesions on dermoscopy. *Br J Dermatol*. 2011;164(1):54-61. doi:10.1111/j.1365-2133.2010.10043.x

21. Nomura M, Tanaka M, Nunomura M, Izumi M, Oryu F. Dermoscopy of rippled pattern sebaceoma. *Dermatol Res Pract*. 2010;2010:140486. doi:10.1155/2010/140486.