Original Article

Brief Overview about Port Wine Stain

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Abstract

Background: A group encompassing a wide variety of lesions related to the disorder of vascular development, remain both diagnostic and treatment challenges to treating physicians. The terminology used to describe and classify vascular anomalies is the key for proper diagnosis and treatment. The classification system established by the International Society for the Study of Vascular Anomalies (ISSVA). is now a widely accepted system used to categorize vascular anomalies into two types: (1). vasoproliferative or vascular neoplasms such as hemangioma, and (2). vascular malformations. Vascular malformations, on the other hand, are comprised of abnormally formed channels within a vascular apparatus that are lined by endothelial cells and do not undergo abnormal cellular turnover. They too are congenital in nature, but often go unnoticed at birth, never regress, and grow proportionally with the individual. While most PWSs show a sporadic inheritance pattern, there is a number of associated congenital syndromes in which they manifest greater diffusion in addition to bone hypertrophy such as in Klippel-Trenaunay syndrome (KTS). or neurological involvement, as seen in the Sturge Weber Syndrome (SWS). Initially, PWSs clinically appear as fat pink or bright red hued patches. Although they may lighten during the first few months of life, probably due to the physiological anemia of infancy, this phenomenon is not predictive of a spontaneous regression of the vascular stain. Unlike other types of vascular birthmark, port wine stain do not fade with age. Initially a light pink to red macule at birth, the natural progression of port wine stain is to a deeper reddish-purple lesion that may become hypertrophic or nodular due to progressive vessel ectasia. In addition, the aberrant cosmetic appearance of PWS may significantly impede the patient's psychosocial development and well-being, and because 70% to 80% of these birthmarks occur in the head and neck regions, many patients seek treatment.

Keywords: port wine stain

INTRODUCTION

Port-wine stains are congenital vascular malformations with an incidence of 3–5 per 1000 neonates. There are approximately 25 million patients worldwide **(1).**

Vascular anomalies:

A group encompassing a wide variety of lesions related to the disorder of vascular development, remain both diagnostic and treatment challenges to treating physicians. The terminology used to describe and classify vascular anomalies is the key for proper diagnosis and treatment. The classification system established by the International Society for the Study of Vascular Anomalies (ISSVA). is now a widely accepted system used to categorize vascular anomalies into two types: (1). vasoproliferative or vascular neoplasms such as hemangioma, and (2). vascular malformations **(2).**

PWSs also known as "nevus flammeus," are cutaneous vascular malformations involving the postcapillary venules with potentially devastating physical and psychological complications, which can establish either congenital or acquired in form. Unlike hemangiomas, PWSs do not have a tendency to involute. PWSs are well defined, flat and grow proportionately in surface area with the child**.** Clinically, appears as pink-red to violaceous patches on the skin. Although congenital PWSs are more common, its occurrence in infants is 0.3-1.4% without any sex prediliection**.** While most PWSs show a sporadic inheritance pattern, there is a number of associated congenital syndromes in which they manifest greater diffusion in addition to bone hypertrophy such as in Klippel-Trenaunay syndrome (KTS). or neurological involvement, as seen in the Sturge Weber Syndrome (SWS). **(3).**

Clinical course:

Initially, PWSs clinically appear as fat pink or bright red hued patches. Although they may lighten during the first few months of life, probably due to the physiological anemia of infancy, this phenomenon is not predictive of a spontaneous regression of the vascular stain **(4).**

In contrast to other birthmarks such as hemangioma or the so-called salmon patches, capillary malformations normally persist throughout life and have no tendency toward involution.

Unlike these nosological entities, PWSs persist, untreated, into adulthood becoming thicker and raised, as well as of a darker violaceous color as a result of progressive vascular ectasia **(3).**

This is thought to be a consequence of a neural vascular tone modulation lack resulting from both sympathetic and sensory perivascular innervation faw, as immunohistochemically documented by Smoller**.** and Rydh. Approximately two-thirds of patients with PWSs may develop more severe morphological changes like isolated or multiple aggregated nodules (cobblestone pattern). and progressive soft tissue and/or bone hypertrophy. This risk is particularly true in Sturge-Weber and Klippel Trenaunay syndromes **(5).**

These modifications are conspicuous by the age of 50 but often begin early with overgrowth of soft and bone tissues appearing at an average age of nine and fifteen years, respectively, and during the third decade of life (20 s). for nodules raising **(3).**

Based on clinical and histological evidences, Finley and colleagues**.** differentiated between thickening and nodules occurring in PWSs. Thickening was defined as an exaggeration of the ectasia process whereas nodules were polypoid tumors consisting of thick- and thin-walled vessels of varying caliber and surrounding stroma proliferation, which they categorized as arteriovenous malformations (AVMs). **(6).**

PWS can be diagnosed promptly based on the anatomic location and characteristic appearance of the lesion. However, PWS can exist alone or be associated with many other congenital vascular malformations, such as SWS, Parkes-Weber syndrome, Klippel-Trenaunay syndrome (KTS)., Proteus syndrome and arteriovenous malformations (AVM). Therefore, the co-existence of any other vascular anomalies with PWS, particularly in infants, needs to be examined. Imaging systems, such as Doppler, computed tomography (CT). or magnetic resonance imaging (MRI)., can be helpful in determining any possible vascular malformations located in deep tissues, e.g., cerebral vascular malformations or AVM. PWS needs to be differentially diagnosed from infantile hemangiomas (IHs). which usually involute over time. Molecularly, ECs from IHs are Glut-1 positive, but PWS ECs are not **(7).**

Diagnosis:

Non invasive diagnostic technique :

In recent years, noninvasive diagnostic techniques such as dermoscopy, high-frequency ultrasound (HFUS)., VISIA-CR™ system, cross- polarized diffuse reflectance imaging system (CDR)., reflectance spectrophotometers and tristimulus colorimeter, reflectance confocal microscopy (RCM)., laser Doppler flowmetry (LDF)., optical coherence tomography (OCT)., laser speckle contrast imaging (LSCI)., and spatial frequency domain imaging (SFDI). have been attempted to analyze PWS.

Clinical types of port wine stains:

Port-wine stains were classified into four types: red, purple, hypertrophic and nodular port-wine stains**.** A previous study revealed that port-wine stains may not only be a congenital disease involving vessels, but also involve other tissue components in the skin **(8).**

Complication of port wine stain:

Unlike other types of vascular birthmark, port wine stain do not fade with age. Initially a light pink to red macule at birth, the natural progression of port wine stain is to a deeper reddish-purple lesion that may become hypertrophic or nodular due to progressive vessel ectasia. a review of 415 patient at new York university medical center revealed nearly two third develop nodularity or hypertrophy of their port wine stain by their fifth decade of life. These mature lesions are more prone to spontenous or trauma-induced bleeding. This potentially could serve as a nidus for infection which could be hazardous to patient health. Pyogenic granuloma have a great tendency to occur within port wine stain as they mature a giant proliferative hemangioma arising in aport wine stain have been reported **(9).**

Underling hypertrophy of the underling area can occur even with any extra cutaneous manifestation. Hypertrophy can lead to asymmetric facies or extremities sometimes requires orthopedic intervention. progressive nodularity or hypertrophy of port wine stain may block ear canals, nasal passages, field of vision or closure of the mouth, resulting in interference with vital functions as hearing, breathing, sight and eating or talking. While the nature progression of port wine stain can lead to major complication, the major morbidity of port wine stain is psychosocial. most port wine stain occur on face and neck, the value our society places on facial appearance is high and many studies have shown that physical attractiveness plays a major role in psychological development and social interaction. Attractive persons are often judged to possess more desirable personalities, to be more intelligent, to be more successful in occupation and marriage and to have a higher social status than others **(10).**

Multiple-pass approaches:

The extent of vascular damage may be increased by the use of multiple-pass techniques, although this has been controversial. based on histologic assessment of vascular damage of non-PWS skin, suggested that multiple-pass PDL treatment may be beneficial but stressed the relevance of the interval between pulses. At minimal purpuric doses, they demonstrated an increase in the depth of vascular damage from 0.7 to 1.3 mm with an increasing interpulse interval of up to 60 seconds. Further increase of the interpulse

interval to 5 or 30 minutes resulted in a similar depth of vascular injury as singledose treatments. In contrast, treatment above purpura threshold resulted in increased depth of damage with increasing interpulse intervals up to at least 30 minutes. All interpulse intervals were tolerated well with no adverse effects other than transient purpura. Altogether, multiple passes may be beneficial but further evidence is needed. **(11).**

Epidermal cooling Epidermal melanocytes constitute a major limitation to laser therapy, as they have the capacity to absorb a significant portion of the laser energy and mediate laser-induced scarring and pigmentary damage. Consequently, irradiances must be chosen such that adequate venular heating is achieved without generating excessive temperatures at the basal membrane. This has proven rather challenging, as unequivocal objective parameters to determine the optimal irradiance are currently lacking. With the advent of epidermal cooling technology, however, it has become possible to minimize nonselective epidermal thermal injury and concurrently use higher fluences to treat PWS more effectively **(11).**

Cooling also has the added advantage of significantly reducing the patient's level of pain and discomfort. As a result, the majority of modern clinical laser systems are equipped with integrated cooling technology. Primary cooling methods include contact cooling, cold air cooling, and cryogen spray cooling. Cryogen spray cooling is currently the method of choice of the authors, as we believe it can selectively cool the epidermis and perivascular tissue while minimally affecting the temperature of the underlying blood vessels **(12).**

Experimental treatment modalities:

There are a number of promising experimental therapies for PWS currently being investigated. In this section, several of these modalities are addressed, including photodynamic therapy (PDT)., angiogenesis inhibitors, hemodynamic alterations in PWS vasculature, and site-specific pharmaco-laser therapy (SSPLT).

Photodynamic therapy:

PDT involves the activation of a photosensitizer by visible light. The absorption of light triggers a photochemical-biological reaction that, in the presence of oxygen, leads to reactive oxygen species generation that in turn causes direct endothelial cell damage, thrombosis, and shutdown of vasculature. Oxidative cell damage is limited to those photoilluminated areas containing sufficient photosensitizer concentrations, as the short diffusion distance of reactive oxygen species (# 0.1 microm). limits collateral damage to minimal to none. These aspects impart site specificity to PDT that is similar to selective photothermolysis. The efficacy of PDT depends on factors such as wavelength and light intensity, exposure time, and choice of photosensitizer. It has been suggested that the therapeutic efficacy of PDT is equivalent or even superior to PDL treatment. **(13).**

Although PDT has promise as a treatment approach, there are potential limitations. First, PWS vessels are believed to have a normal endothelial lining, which does not allow for the selective accumulation of photosensitizing agents within the target vessels. The use of antibody-labeled photosensitizers for selective targeting is futile without differential surface antigen expression on the endothelial cells in PWS vasculature. Another major drawback of PDT is the generalized photosensitivity that persists after therapy **(13).**

With the currently used photosensitizers, patients are counseled to avoid direct sunlight for up to 4 weeks after PDT treatment **(14).** This inconvenience can be partially addressed by using photosensitizers with shorter circulation half-lives, such as benzoporphyrin derivative monoacid A, which remains photoactive for no more than 5 days. Other limitations include the long length of treatment and the high costs of photosensitizers. Further study and careful planning of PDT regimens are required to address these issues. The combination of PDL and PDT has been proposed to be synergistic. The suggested mechanism of action entails the administration of an initial, subtherapeutic PDT pulse to create injury to the vascular wall, after which irradiation with the PDL enhances the PDT-induced vascular damage. Because two methods of injury are used, lower radiant exposures can be employed for the PDT and PDL components, minimizing adverse effects such as scarring while inducing more extensive damage to the PWS vasculature. In a chick chorioallantoic membrane model, the combination of PDT and PDL treatment yielded significantly more vascular damage than either modality alone **(15).**

REFERENCES

1. **Lister T, Wright PA, Chappell PH, et al. (2012).** Simulating light transport through skin for color prediction of port wine stain lesions: A review. J Biomed Opt ;17:110901.
2. **Ortiz AE and Nelson JS.(2012).** Port-Wine Stain Laser Treatments and Novel Approaches. Facial Plast Surg ;28:611-20.
3. **Nguyen CM, Yohn JJ, Huff C, et al. (1998).** Facial port wine stains in childhood: prediction of the rate of improvement as a function of the age of the patient, size and location of the port wine stain and the number of treatments with the pulsed dye (585 nm). laser. Br J Dermatol ;138:821-5.
4. **Lee JW, Chung HY (2018).** Capillary malformations (portwine stains). of the head and neck: natural history, investigations, laser, and surgical management. Otolaryngol Clin North Am 51:197– 211.
5. **Tannous Z, Rubeiz N, Kibbi AG et al., (2010).** Vascular anomalies: portwine stains and hemangiomas. J Cutan Pathol 37:88–95.
6. **Klapman MH and Yao JF (2001).** Thickening and nodules in port-wine stains. J Am Acad Dermatol 44:300–302.
7. **Yu W, Ma G, Qiu Y, et al.(2016). 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.;74(3).527–35.
8. **Tan OT, Carney JM, Margolis R, Seki Y, Boll J, Anderson RR, et al. (1986):** Histologic responses of port-wine stains treated by argon, carbon dioxide, and tunable dye lasers: a preliminary report. Arch Dermatol;122:1016-22.
9. **Holloway KB , Ramos-caro FA , Brownlee RE , et al. (1994).** Giant proliferative hemangioma arising in a port wine stain . J Am Acad dematol ;31:675-6.
10. **Clifford MN and Walster E. (1973).** The effect of physical attractiveness on teacher expectations . social Educ ;46:248-58.
11. **Greve B and Raulin C.(2004).** Prospective study of port wine stain treatment with dye laser: comparison of two wavelengths (585 nm vs 595 nm). and two pulse durations (0.5 milliseconds vs 20 milliseconds). Lasers Surg Med 34:168-73.
12. **Nelson JS, Milner TE, Anvari B, et al. (1996).** Dynamic epidermal cooling in conjunction with laser-induced photothermolysis of port wine stain blood vessels. Lasers Surg Med ;19:224-9.
13. **Lu YG, Wu JJ, Yang YD, et al.(2010).** Photodynamic therapy of port-wine stains. J Dermatolog Treat ;21:240-4.
14. **Yuan KH, Li Q, Yu WL, et al., (2009).** Photodynamic therapy in treatment of port wine stain birthmarkserecent progress. Photodiagnosis Photodyn Ther ;6:189-94.
15. **Kelly KM, Kimel S, Smith T, et al. (2004).** Combined photodynamic and photothermal induced injury enhances damage to in vivo model blood vessels. Lasers Surg Med ;34:407-13.