Hypertrophy in port-wine stains: Prevalence and patient characteristics in a large patient cohort

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Background: Port-wine stains (PWS) may thicken and darken with age. Little is known about the pathogenesis and epidemiology of PWS hypertrophy because of the lack of large studies.

Objective: We sought to assess the prevalence and characteristics of patients with hypertrophic PWS.

Methods: Medical records and clinical photographs of all patients with PWS visiting our clinic between 2005 and 2009 were examined to identify hypertrophy. Patients were sent questionnaires regarding their hypertrophic PWS.

Results: In all, 335 patients (age 0-81 years; 69% female) with PWS were included. Hypertrophy was found in 68 patients (20%; 32 male, 36 female) and classified as thickened (5%), nodular (8%), or both (7%). Color of hypertrophic PWS was mainly red (50%) or purple (44%). Patients with hypertrophy in their PWS were mostly (68%) older than 40 years, and rarely (7%) younger than 20 years. When older than 50 years, 71% of all patients had hypertrophy in their PWS. Median age of onset of PWS hypertrophy was 31 years (12 years for thickened, 39 years for nodular).

Limitations: This was a retrospective study in a selected population.

Conclusion: Hypertrophy is an important feature in the development of PWS and affects a majority of patients older than 50 years. Depth of color of the PWS is associated with hypertrophy, whereas location and size appear not to be related. More attention should be drawn to therapy and prevention of hypertrophic PWS. Diffuse thickening and nodules should be distinguished, as a different age of onset may indicate different pathomechanisms. (J Am Acad Dermatol 2012;67:1214-9.)

Key words: capillary malformation; hypertrophy; nodules; port-wine stain; prevalence; pulsed dye laser; thickening.

Port-wine stains (PWS) are congenital capillary malformations seen in approximately 0.3% of newborns.[1] They are frequently located on the face and neck, but can present anywhere on the body. Although generally flat and pink in infancy, PWS may thicken and darken in color during adult life as a result of progressive vascular ectasia.[2,3]

Abbreviations used:

CI: confidence interval
PDL: pulsed dye laser
PWS: port-wine stains

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Treatment of PWS has improved since the introduction of the pulsed dye laser (PDL) in the 1980s.[4-6] Despite adjustments in technique over the years, the results of laser therapy are unsatisfactory in 20% of patients.[7] PWS often lighten in color with laser therapy, but rarely disappear completely. Blue-purple, hypertrophic PWS in particular do not respond well to treatment with PDL.

Little is known about development of hypertrophy in PWS. Finley et al[8] reported hypertrophy in 2 of 500 patients with PWS, referring to a cobblestone pattern; in 5 additional patients, they observed tumors consisting of proliferating vessels. Geronemus and Ashinoff[9] determined that nearly two thirds of patients with PWS develop nodules and hypertrophy by the fifth decade of life.

CAPSULE SUMMARY

- Port-wine stains (PWS) may thicken and darken with age. Little is still known about the pathogenesis and epidemiology of hypertrophy in PWS.
- We found hypertrophy to be an important feature in the development of PWS, and a burden to patients. Median age at onset of hypertrophy was 31 years. Hypertrophy was observed in 71% of patients older than 50 years.
- Therapy and prevention of hypertrophy in PWS should receive more attention.

Dermatology textbooks cite 2 studies.[10,11] Mills et al[12] reported 31 (10%) cases of PWS with hypertrophy in a demographic study of 283 patients. Klapman and Yao[13] described the prevalence (40%) and characteristics of thickening and nodules in 173 patients with PWS. The peak age of onset of hypertrophy was between 20 and 39 years and the prevalence of nodules increased with age.

PWS are known to have a negative influence on psychologic well-being.[7,14-16] The aim of our study was to assess the prevalence and characteristics of patients with hypertrophic PWS.

METHODS

Patients

This study cohort comprised all patients with PWS who consulted The Netherlands Institute for Pigment Disorders in Amsterdam between September 2005 and March 2009. Patients were eligible for inclusion if they had PWS recorded in their medical file and digital photographs. Patients with Klippel-Trénaunay syndrome were not present in our database. Records were reviewed for gender, age, hypertrophy, color of the PWS, location on the body, and details of previous treatments. Hypertrophy was identified by a physician and classified as either nodular, describing papules and nodules arising in the PWS, or as diffuse thickened, describing both larger plaques and deeper thickening of underlying structures (Fig 1). Characteristics of patients with flat PWS were compared with those of patients with hypertrophic PWS.

A questionnaire was sent to patients who appeared to have a hypertrophic PWS with questions regarding color change, thickening and/or nodules, age at onset of these changes, the main reasons for initiation of laser therapy, and the impact on their daily life. Adult patients were asked to complete and return the questionnaire. Patients younger than 18 years completed the questionnaire with the help of their parents. Similar questionnaires were sent to an age-matched control group with flat PWS.

Statistical analysis

We analyzed all questionnaires and entered them into a Microsoft Office Excel 2003 database (Microsoft Corp, Redmond, WA) together with the characteristics of every patient. Statistical analysis was performed using software (SPSS, Version 16.0, SPSS Inc, Chicago, Ill). Continuous variables were presented as medians with interquartile ranges. The proportions were presented as percentages with a 95% confidence interval (CI). We used multivariate logistic regression models to quantify the risk factors for hypertrophic PWS. Potential confounding factors were evaluated based on a significant outcome (95% CI) in the univariate regression analysis.

RESULTS

Patient characteristics

We identified 342 patients with PWS of whom 7 were excluded because of absence of digital photographs in the file. The remaining 335 patients were included and analyzed. The majority was female (231 patients, 69%). The median age at the time of database screening was 22 years (0-81 years, interquartile range 14-41 years). The most common PWS location was the face (68%) with the majority on the cheeks. The color of the PWS was usually red (49%).

Of 335 patients, 68 (20%; 95% CI 16.0%-24.6%) had hypertrophic PWS (Table I). There was an equal male-female ratio (32 male, 36 female). The median age of this subgroup at the time of database screening was 50 years (12-81 years, interquartile range 32-57 years).

The most common location for hypertrophic PWS was the face (79%) with hypertrophy involving the cheeks in the majority of cases. There were less