ORIGINAL ARTICLE

Orodental manifestations of facial port-wine stains

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Background: Patients with facial port-wine stains (PWS) often demonstrate oral manifestations of their disorder; however, the spectrum and prevalence of such findings among a cohort of patients with PWS has not been established. As a result, dermatologists and oral health specialists may be uncertain how to counsel their patients with PWS regarding oral hypervascularity, bony oral changes, and oral hygiene.

Objectives: We sought to identify physical findings and complications involving the teeth, oral cavity, and perioral structures in individuals with facial PWS.

Methods: This was a cross-sectional study of 30 patients with facial PWS. Descriptive data were collected through anonymous paired surveys completed by patients and their dentists, and analyzed (Fisher exact test) for trends based on physical findings and stage of the PWS.

Results: The most common orodental manifestations according to patients were enlargement of the lip (53.3%), stained gums (46.7%), abnormal bite (30%), and spontaneous bleeding of the gums (26.7%). Staining of the gingiva correlated significantly with gingival hyperplasia (P = .006), maxillary hyperplasia (P = .014), and widened interdental spaces (P = .002), and in all cases gingival staining predated these findings. Lip hyperplasia was reported more frequently by patients than by their dentists (50% vs 18.2%, P = .008). Orodental manifestations were more common among patients with darker and thicker PWS. Hemorrhage after dental procedures was rare (4.5%).

Limitations: Modest sample size and difficulty recruiting control subjects are limitations.

Conclusions: Facial PWS commonly affect the orodental structures, and intraoral staining may predict future complications. (J Am Acad Dermatol 10.1016/j.jaad.2011.11.929.)

Key words: bleeding; dental; face; gingiva; gums; malocclusion; mouth; oral; port-wine stain; teeth.

Port-wine stains (PWS) are congenital vascular birthmarks, occurring in an estimated 0.3% of newborns. Described in the literature as either capillary or venular malformations, these lesions are characterized histologically by ecstatic vessels and a deficiency of nerves in the papillary plexus of the skin in the affected area.[1,2] It is hypothesized that the deficient nerves are of sympathetic origin, and that unchecked parasympathetic influence on blood flow through the postcapillary venules results in progressive vascular ectasia.[1,2] The dilated venules produce a discoloration or "stain" of the skin within the affected region that will be with the patient for life.

Abbreviation used:
PWS: port wine stain

PWS lesions are observed more commonly among Caucasians than African Americans and Asians. There is no gender predilection. They usually begin

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as flat and pink, becoming thicker and darker over time. In advanced lesions, nodules or "cobblestones" may be present.[3] Adjacent bone and soft tissue often becomes hyperplastic; this process is not well understood but is thought to result from the release of growth factors from skin affected by the PWS.

Head and neck PWS usually follow the distribution of the branches of the trigeminal nerve, but more than one branch may be simultaneously involved. When the maxillary and/or mandibular divisions are involved, the bone and soft tissue of the oral cavity are often affected.[4-12] As a result, dentists and dental specialists often anticipate complications such as bleeding caused by the hypervascularity of the gingival and oral soft tissues, along with functional and cosmetic deformities from bony overgrowth involving the jaws and teeth. Although there are a number of case reports describing orodental manifestations of facial PWS,[4-12] to our knowledge, no previous study has established the spectrum and prevalence of such findings among a larger population. The goal of this study was to identify and quantify the common orodental complications linked to PWS of the face.

CAPSULE SUMMARY

- Patients with facial port-wine stains (PWS) often demonstrate oral manifestations of their disorder.
- This article identifies physical findings involving the oral and perioral structures in individuals with facial PWS and identifies patients at risk for future complications.
- Familiarity with the orodental manifestations of PWS may result in improved dental care for patients with PWS.

METHODS

Approval for the study was obtained from the Institutional Review Board of the Eastern Virginia Medical School. Study design was a cross-sectional survey of patients with PWS recruited from the practices of collaborating physicians who treat PWS, and from volunteers who frequent www.birthmarks.com, a World Wide Web site devoted to individuals with PWS. A medical database format was used, collecting experimental (rather than observational) data for variables established prospectively. Patients were eligible for enrollment if they were between 1 and 89 years of age and had a PWS involving the face. Patients were specifically excluded if their PWS did not involve the face, or if the PWS was an infantile "salmon patch" (capillary vascular malformation) involving the forehead ("angel kiss") or nape of the neck ("stork bite").

Enrolled participants were provided a questionnaire to be completed by the participant with accompanying instructions, and a paired questionnaire to be completed by the participant's dentist based on a clinical examination. Data were collected anonymously, although surveys were numbered for purposes of pairing participant questionnaires with those of their dentists.

Collected data were analyzed using Fisher exact test for concordance between patient and dentist, and for trends based on physical findings, facial distribution,[13] and stage of progression[2,3] of the PWS as determined by color, thickness, and/or cobblestone formation. Patients with PWS involving the forehead and scalp were intended to serve as a control group because only those with mid-facial and lower-facial lesions were deemed likely to have oral cavity involvement.

RESULTS

In all, 31 patients with facial PWS were enrolled in the study. Only one patient with solitary involvement of the forehead (V1 [first division of trigeminal nerve] dermatomal distribution) responded, effectively eliminating the control group; this patient was not included in the final study group of 30. Mean age of the participants was 28.8 years (SD ± 19.6; range 1-62). Eleven patients had their PWS in the labio-infraorbito-angular or V2 (second division of trigeminal nerve) dermatomal distribution, 4 had a jaw-neck or V3 (third division of trigeminal nerve) dermatomal distribution, and 9 had a labio-infraorbito-angulo-(forehead)-temporal or combined V1/V2 distribution. Six patients had some combination of these distributions (1 bilateral perioral [V2 and V3], 1 forehead and jaw [V1 and V3], and 4 covering the entire half of their face [V1, V2, and V3]. Four patients reported having Sturge-Weber syndrome (facial PWS, glaucoma, seizures, mental retardation, and ipsilateral leptomeningeal angioma). Fifteen participants described their PWS as confluent (consistent discoloration of skin throughout the affected area), whereas 9 patients reported geographic distribution (patches of normal-colored skin within the PWS); 6 patients did not comment on the consistency of their PWS.

Patients' attention to dental hygiene was assessed to illuminate any confounding impact on orodental complications. Of the 30 participants, one patient reported seeing a dentist less than once a year, 4 went to the dentist yearly, and 25 made two or more

Table I. Orodental manifestations as reported by patients (N = 30)

Orodental manifestation	Affected individuals (%)
Lip hyperplasia	16 (53.3)
Stained gingiva	14 (46.7)
Bleeding gingiva	8 (26.7)
Malocclusion	9 (30)
Spacing between teeth	7 (23.3)
Gingival hyperplasia	6 (20)
Maxillary hyperplasia	5 (16.7)
Prolonged inflammation after dental procedures	3 (10)
Hyperplasia of tongue	3 (10)
Gingivitis	2 (6.7)
Ulcers	2 (6.7)
Numbness	1 (3.3)
Mandibular hyperplasia	2 (6.7)
Hemorrhage from dental procedures	1 (3.3)
Bleeding tongue	0 (0)
Increased mobility of teeth	0 (0)

dental visits per year. Similarly, one patient reported brushing less than once a day, 4 patients brushed once a day, and 25 brushed two or more times daily. Twenty participants flossed twice a week or less; 10 patients flossed daily.

Trauma and hormonal fluctuations can also act as a stimulus for PWS progression; as a result, several parameters were assessed in relation to the appearance of dental symptoms. Three patients admitted to the appearance of dental problems within 6 months of pregnancy or menopause, and two patients reported worsening of dental problems shortly after undergoing periodontal surgery. Of interest, 4 patients reported the development of orodental complications within 6 months of undergoing laser surgery for their PWS, although they did not specify whether any of the treatment was intraoral.

Orodental manifestations of PWS as reported by patients are listed in Table I. The most common dental complication reported by patients was lip enlargement (53.3%), followed by red stained gums (46.7%), abnormal bite (30.0%), spontaneous bleeding of the gums (26.7%), and widening of spaces between teeth (23.3%). The problems most frequently reported by dentists were red stained gingiva (41%), malocclusion (31.8%), gingival hyperplasia (27%), and gingivitis (22.7%) (Figs 1 to 3). In all, 22 patients had a paired dentist response, among which there was good correlation between manifestations reported by patients and their dentists (Table II); only the prevalence of lip enlargement reported by patients was statistically different from that reported by their dentists (50% vs 18.2%, respectively; $P = .008$).

Fig 1. Oral cavity of patient with extensive right facial port-wine stain. Staining is noted in mucosa of gingiva, vestibule, palate, and tongue on right side. Hyperplasia of right side of tongue is also seen.

Fourteen of the 30 patients (46.7%) reported staining of the gingiva. All patients who reported hyperplasia of the jaws, hyperplasia of the gingiva or tongue, or abnormal spacing between the teeth were also among this group (Table III). Gingival staining correlated significantly with gingival hyperplasia ($P = .006$), maxillary hyperplasia ($P = .014$), and widened interdental spaces ($P = .002$). Gingival staining predated these findings in all cases, noted at birth in 6 individuals, at age 4 years in two patients, and at age 14 years in one patient; no age of onset was reported by the remaining 5 patients. Among those patients in whom stained gingiva developed at a later age, this finding was noted either before or at the same age as the appearance of other dental complications.

Hyperplasia of the lips did not correlate with gingival involvement ($P > .6$). Of the two patients who reported enlargement of the mandible, only one had a PWS in the jaw-neck/V3 distribution.

Orodental manifestations were more common among patients with darker and thicker PWS, although most of the correlations did not reach statistical significance (Tables IV and V). The exception was the correlation between darker PWS and gingival bleeding, with 7 of 18 patients with red or purple PWS reporting bleeding gingiva, compared with only one of 11 of those with a pink PWS ($P = .048$). The discrepancy between flat and thickened PWS was most pronounced for gingival hyperplasia, bleeding of the gingiva, and malocclusion. Darker and thicker PWS were also noted with increasing age. The average age was 21.8 years for those with a pink PWS, 25.2 years for red PWS, and 41.3 years for purple PWS. Mean age for individuals with a flat PWS was 19.2 years, whereas for those with a thickened or cobblestoned PWS mean ages were 46.6 years and 48.0 years, respectively.

Although bleeding gums were reported frequently by patients (26.7% total, 22.7% in paired

Fig 2. Maxillary alveoli and dentition in patient with right facial port-wine stain. Bony hyperplasia, abnormal dental eruption, and staining of palate and vestibular mucosa are noted on right side compared with left. Inflammation of marginal gingiva and accumulation of plaque are also more significant on affected side.

Fig 3. Subtle left facial port-wine is best appreciated in upper lip. Underlying maxillary alveolus demonstrates mucosal staining, widened interdental spaces, and alteration of occlusal plane.

survey), significant hemorrhage after dental procedures was not commonly reported by their dentists (4.5%). Four patients reported experiencing complications after oral surgery procedures, one with prolonged bleeding and the other 3 with extensive swelling/inflammation after the procedure.

DISCUSSION

Orodental manifestations of PWS, such as staining of the oral soft tissues, hyperplasia of the gingiva, oral bleeding, overgrowth of the bony alveoli, and possible interruption in dental eruption sequence, have been reported to date only in case reports and case series.[11] As a result, opinions in the literature vary widely regarding the dental, periodontal, and surgical treatment of patients with PWS. Although one author claims, "Port wine stains or capillary malformations rarely present major problems for the oral and maxillofacial surgeon,"[9] another suggests, "Periodontal probing should not be done as even gentle probing can result in uncontrolled bleeding."[14] To our knowledge, our study is the first to attempt to describe the spectrum of orodental findings in PWS in a larger cohort to better establish their long-term dental management.

In this study of patients with mid-facial and lower-facial PWS, enlargement of the lips was the most common finding, reported by about half of the cohort. However, results from the paired survey revealed a reporting discrepancy between patients and their dentists. We speculate that in mildly affected patients, lip enlargement is more apparent and more cosmetically critical to patients and may be underreported by their dentists. Alternatively, the results may suggest a focus by the dentist on abnormalities of the intraoral rather than the perioral structures. Another explanation would be overreporting of lip enlargement by patients; however, given the magnitude of the statistical difference for this single finding we believe this explanation is less likely. Regardless, dental professionals should assess patients with PWS for lip enlargement that may impact their psychosocial well-being, and in severe cases may interfere with the performance of intraoral procedures.

Staining of the gingiva was reported with high frequency in the patient and dentist surveys and

Table II. Orodental findings in paired patient-dentist responses (N = 22)

Orodental manifestation	Reported by patients (%)	Reported by dentists (%)	Kappa coefficient (95% CI)	*P* value
Stained gingiva	10 (45.5)	9 (40.9)	0.35 (−0.04 to 0.75)	.706
Bleeding gingiva	5 (22.7)	4 (18.2)	0.58 (0.16 to 1.00)	.564
Hemorrhage	1 (4.5)	1 (4.5)	1.00 (1.00 to 1.00)	NA
Inflammation	1 (4.5)	0 (4.5)	NA	NA
Gingival hyperplasia	5 (22.7)	6 (27.3)	−0.09 (−0.47 to 0.29)	.739
Lip hyperplasia	11 (50.0)	4 (18.2)	0.36 (0.06 to 0.66)	.008*
Maxillary hyperplasia	4 (18.2)	3 (13.6)	0.49 (0.001 to 0.98)	.564
Mandibular hyperplasia	0 (0.0)	0 (0.0)	1.00	NA
Abnormal spacing	4 (18.2)	3 (13.6)	0.15 (−0.33 to 0.64)	.655
Malocclusion	6 (27.3)	7 (31.8)	0.46 (0.05 to 0.86)	.655
Mobile teeth	0 (0.0)	1 (0.0)	NA	NA
Tongue hyperplasia	1 (4.5)	1 (4.5)	−0.05 (−0.11 to 0.02)	1.000
Tongue bleeding	0 (0.0)	0 (0.0)	NA	NA
Gingivitis	1 (4.5)	5 (22.7)	−0.08 (−0.22 to 0.06)	.103
Oral ulcers	0 (0.0)	0 (0.0)	NA	NA

Kappa coefficients or *P* values cannot be calculated because of low response rate.
CI, Confidence interval; *NA*, not applicable.
*Statistical significance at $P < .05$.

Table III. Selected orodental manifestations stratified by gingival staining

	Unstained gingiva, n = 16 (%)	Stained gingiva, n = 14 (%)	*P* value
Gingival hyperplasia	0 (0.0)	6 (42.9)	.006*
Abnormal spacing	0 (0.0)	7 (50.0)	.002*
Maxillary hyperplasia	0 (0.0)	5 (35.7)	.014*
Mandibular hyperplasia	0 (0.0)	2 (14.3)	.209
Tongue hyperplasia	0 (0.0)	3 (21.4)	.089
Bleeding gingiva	3 (18.8)	5 (35.7)	.417
Lip hyperplasia	8 (50.0)	8 (57.1)	.695
Prolonged inflammation	0 (0.0)	3 (21.4)	.089

*Statistical significance at $P < .05$.

Table IV. Selected orodental manifestations stratified by port-wine stain color

	PWS color			
Complication	Pink, n = 11 (%)	Red, n = 10 (%)	Purple, n = 8 (%)	*P* value
Stained gingiva	4 (36.4)	4 (40.0)	5 (62.5)	.638
Bleeding gingiva	1 (9.1)	2 (20.0)	5 (62.5)	.048*
Maxillary hyperplasia	2 (18.2)	0 (0.0)	3 (37.5)	.121
Abnormal spacing	2 (18.2)	1 (10.0)	4 (50.0)	.190
Lip hyperplasia	4 (36.4)	5 (50.0)	6 (75.0)	.299
Malocclusion	2 (18.2)	2 (20.0)	4 (50.0)	.334
Gingival hyperplasia	2 (18.2)	2 (20.0)	2 (25.0)	1.000

PWS, Port-wine stain.
*Statistical significance at $P < .05$.

predated nearly all of the orodental problems related to PWS. Of the 9 patients who reported the age at which stained gingiva were observed, two thirds reported this trait had been present since birth. Even those who first noted stained gingiva at a later age did so before or at the same age as the appearance of other orodental manifestations. As a result, gingival staining may be considered a predictor of progressive orodental involvement, and dentists should follow up individuals so affected with increased vigilance.

Gingival staining demonstrated a strong correlation with other orodental complications, although it did not predict enlargement of the adjacent lip (Table III). All patients who reported abnormal spacing between teeth and/or hyperplasia of bony structures also experienced staining of the gingiva. This is consistent with literature indicating that lesions extending into the gingiva are those most likely to exhibit overgrowth of adjacent bone.[2] Soft tissues also seem to be affected, because all the participants who reported tongue enlargement or gingival hyperplasia also had stained gingiva. Those with gingival staining were also the only individuals to report prolonged inflammation after tooth extraction or surgery; however, although bleeding was also more common among these patients, the difference was not statistically significant.

Bony alveolar changes may include erosion caused by periosteal pressure from a growing mass of soft tissue or to periodontal disease, or expansion

Table V. Selected orodental manifestations stratified by port-wine stain thickness

Complication	Flat, n = 19 (%)	Thickened, n = 9 (%)	*P* value
Stained gingiva	6 (31.6)	7 (77.8)	.042*
Gingival hyperplasia	2 (10.5)	4 (44.4)	.063
Bleeding gingiva	3 (15.8)	4 (44.4)	.165
Malocclusion	3 (15.8)	4 (44.4)	.165
Maxillary hyperplasia	2 (10.5)	3 (33.3)	.290
Lip hyperplasia	8 (42.1)	6 (66.7)	.419
Abnormal spacing	4 (21.1)	3 (33.3)	.646

*Statistical significance at $P < .05$.

as a result of the same growth factors affecting the oral soft tissues.[11] It has been theorized that such changes may result in increased interdental spacing, malocclusion, and mobility of the teeth. Our study confirmed that patients with stained gingiva were more likely to have bony expansion with increased interdental spacing; however, mobility of the teeth was not observed in our cohort, and in our experience this occurs only in the most advanced intraoral lesions. One may conclude from these data that procedures to surgically correct hyperplasia of the alveolar bone and soft tissue may eventually be required among patients with stained gingiva and can be performed safely in most patients with facial PWS, including those with gingival staining.[4]

Color and texture changes in facial PWS lesions occur in affected patients as they mature, but these processes can begin at any age. Darkening of the PWS is suggestive of increased vascular ectasia[2]; in our study darkening predicted a statistically higher risk of gingival bleeding although texture changes did not (Tables IV and V). Gingival staining was more common among individuals with darker skin lesions, but the difference was not statistically significant. To demonstrate whether darker PWS in patients of the same age have a higher risk of gingival involvement, a larger cohort would likely be necessary. Patients in our study with thickening or nodularity of their PWS demonstrated a statistically increased likelihood of gingival staining, and tended to have a higher risk of orodental manifestations, although the correlations did not reach statistical significance (Table V). Nevertheless, because gingival staining is a predictor of additional orodental manifestations, one might infer that texture changes in skin lesions may be a harbinger of additional orodental changes.

The vascular ectasia associated with oral PWS may predispose patients so affected to minor gingival bleeding. Our study demonstrates an increased likelihood of gingival hyperplasia in these patients, which theoretically further increases the risk of local inflammation and bleeding caused by food impaction and accumulation of plaque within periodontal pockets. Patients with oral PWS should therefore be instructed to perform meticulous oral hygiene to reduce the risk of gingival bleeding, particularly as they age and hyperplasia increases. Our data failed to demonstrate a statistically elevated rate of bleeding among patients with PWS and gingival staining; we attribute this to the small number of patients enrolled. Patients also did not report an unusually high rate of more serious bleeding after oral surgical procedures. This is not surprising because PWS affects small vessels, bleeding from which may be a nuisance but is not usually severe or life-threatening. Bleeding from the tongue was also not a problem for any of the 30 patients participating in this study. We caution, however, that only a small number of patients in this study underwent such procedures, and a much larger cohort will be required to determine whether there is in fact any statistical increase in surgical bleeding among such patients with PWS. In the meantime, we suggest that dentists treating or manipulating the oral soft tissues in patients with oral PWS do so with vasoconstrictors and cautery or electrosurgery wherever possible, and prepare in advance with absorbable hemostatic dressings and extra time for adequate hemostasis.

Results of our study must be interpreted with caution because of a small sample size and lack of a control group. The association between PWS and manifestations with a high background prevalence such as gingival bleeding can be demonstrated only in a larger controlled study. In addition, the data from a survey such as ours reflect only those patients who respond, and therefore involve some degree of selection bias. Nevertheless, one may conservatively conclude that PWS involving the oral soft tissues is a predictor of additional orodental complications including tissue hyperplasia and shifting of teeth, and that patients with darker facial PWS may be prone to gingival bleeding that is safely treated with appropriate preparation.

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