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Case Report

Facial port wine stain with predominant ipsilateral and islands of contralateral oral mucosal involvement: A rare case report

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ABSTRACT

Nevus Flammeus often referred to as PWS (Portwine stain) are benign capillary malformation characterized by multiple dilated vessels in dermis which progressively becomes ectatic as age advances. Portwine stains presents at birth and clinically manifests as flat bright pink, red or violaceous macules that commonly affects the face but can involve other body parts. Over a period of time these lesions become more darker, thicker and nodular often referred to as cobblestoning. Soft tissue hypertrophy of the adjacent areas is also observed especially for facial lesions. The characteristic dermoscopic features are varying morphology of vascular channels like linear, reticular, sausage like, dots or globules and mixed vessels. Mucosal staining is a typical characteristic of facial PWS. This case is reported for its unique variation of facial portwine stain with predominantly ipsilateral oral mucosal involvement with contralateral red macular islands rarely reported in literature with the aforementioned secondary skin changes and typical dermoscopic features.

1. Introduction

Portwine stains are congenital capillary malformations with varying topographic patterns and vivid margins. Recent studies have shown that it results from differentiation-impaired endothelial cells in human skin with a progressive dilatation of immature venule-like vasculatures.[1] A prospective study has proposed an incidence of 0.7% for PWS in new born.[2] PWS initially appears as flat red macules in childhood, as age advances, they become more hyperpigmented with soft tissue hypertrophy and by middle age, often become raised as a result of the development of vascular nodules that are susceptible to spontaneous bleeding or haemorrhage.[3,4] Patients with facial PWS often demonstrates a spectrum of oral manifestation. The most common oral manifestations previously reported were enlargement of the lip (53.3%), stained gums (46.7%), abnormal bite (30%) and spontaneous bleeding of the gums (26.7%).[5] This case report highlights a unique morphological variation of mucosal staining in a patient with facial portwine stain.

2. Case Report

A 60year old male presented with reddish discoloration of skin over right side of the face which has been present since birth and has progressively increased in size over the years. There was no past history of seizures, hemiparesis or vision abnormalities. On dermatological examination there was well demarcated erythematous plaque over right side of face involving the upper lip, maxillary, mandibular, periorbital and temporal region which spanned over all the three divisions (V1,V2 & V3) of trigeminal nerve [Figure 1]. Multiple hyperpigmented discrete and coalescing nodules were noted over maxillary and zygomatic region [Figures 1 and 2]. There was right sided hemihypertrophy of face

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Table 1: Previously documented cases of facial port wine stain with oral mucosal and dermatomal involvement are summarized here with.[6–12]

S.No	Author	Skin involvement with Dermatome	Oral mucosal involvement	Year
1	Namrata C gill et al	Left Facial PWS V1& V2	Ipsilateral staining of palate	2010
2	Tripathi K et al	Right Facial PWS V1 & V2	Ipsilateral hyperplastic lesions on right side of maxilla	2015
3	Shahid M sheikh et al	Right Facial PWS V1,V2 & V3 C1 to C3	Ipsilateral staining of palate	2015
4	Medhini Madie et al	Right Facial PWS V1& V2	Ipsilateral staining of buccal mucosa and soft palate with pyogenic granuloma	2017
5	Medhini Madi et al	B/L Facial PWS V1, V2& V3 C1 to C6	B/L staining of soft palate, labial and buccal mucosa	2017
6	Narender Reddy B et al	B/L Facial PWS , V3	B/L mucosal staining on the floor of the mouth, ventral surface and tip of the tongue and right side of the palate	2018
7	Sasti Priya et al	Right Facial PWS V3	Ipsilateral mucosal staining on ventral aspect of tongue with pyogenic granuloma	2023
8	Present case report	Right Facial PWS , V1,V2 & V3	Predominant ipsilateral staining of palate, buccal mucosa, and tongue, with unique red macular islands of lingual mucosa on the contralateral side	2024

[Figure 1]. Examination of oral mucosa revealed bright red patches over right side of tongue with few areas of sparing over the lateral aspect and red macular islands on left side of tongue [Figures 3 and 4]. Right palatal and buccal mucosa had similar red patches not crossing the midline [Figure 4]. Soft tissue hypertrophy of right side of palate was also noted [Figure 3]. General examination did not reveal any abnormality of limb length, girth and dilated tortuous veins. System examination was normal. Dermoscopy of the lesion revealed dot vessels, globules vessels, linear vessels and sausage vessels [Figures 5 and 6]. Patient was referred for a neurological and ophthalmological evaluation but was eventually lost to follow up.

Figure 1: Unilateral facial portwine stain with soft tissue hypertrophy

Figure 2: Nodularity over port wine stain

3. Discussion

Facial portwine stains are unique birthmarks most commonly involving maxillary and mandibular division of trigeminal nerve. Majority of PWS lesions are unilateral and follow the typical distribution of the trigeminal nerve. A study of 310 PWS patients, 32% had lesions in the area supplied by the maxillary (V2) branch of the trigeminal nerve, 41% had lesions involving both the ophthalmic (V1) and maxillary (V2) branches, 5% had lesions in the combined maxillary (V2) and mandibular (V3) branches, and 10% had lesions across all three branches[13] PWS can be an isolated manifestation or can

Figure 3: Ipsilateral mucosal staining over right buccal mucosa, palate and tongue with red macular islands on contralateralside (Blue arrow)

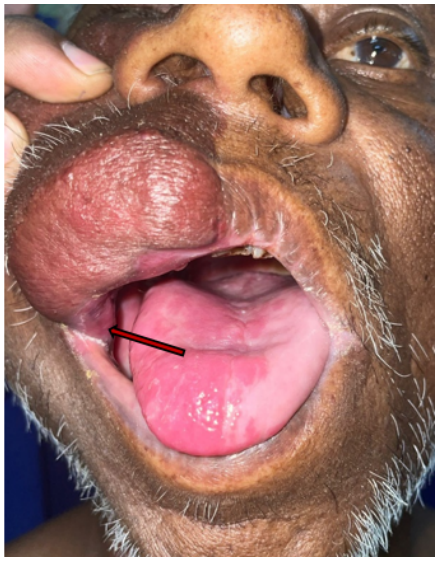

Figure 4: Staining of right buccal mucosa (Red arrow)

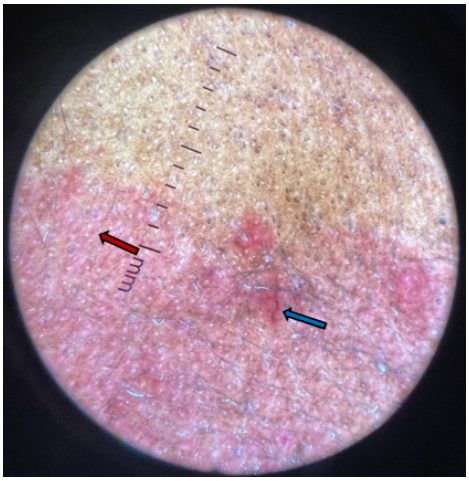

Figure 5: Dermoscopy showing dot vessels (Red arrow) Linear vessels (Blue arrow)

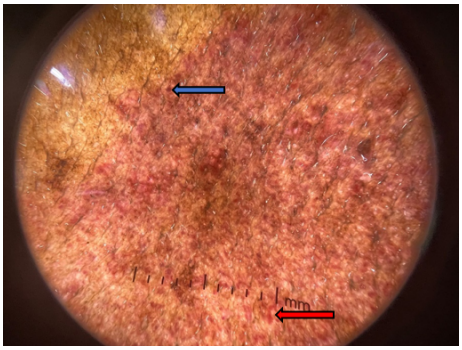

Figure 6: Globule vessels (Blue arrow) Sausage vessels (Red arrow)

be associated with other congenital vascular malformations such as SWS (Sturge weber syndrome), Parkes-Weber syndrome, Klippel -Trenauny syndrome, Proteus syndrome and arteriovenous malformation. When associated with SWS they can present with various neural and ocular manifestations namely leptomenigeal angiomatosis and glaucoma.[14] As age advances many secondary changes are noted over these lesions namely fibrovascular nodules, cutaneous thickening, soft tissue hypertrophy and pyogenic granuloma. Dermoscopy of the PWS in this case showed varying morphology of blood vessels namely dot vessels, globule vessels, linear vessels and sausage vessels. It is helpful to differentiate PWS from other vascular diseases such as telangiectasia, angioma serpiginosum and angiokeratoma.[15] The orodental manifestations were more frequently observed among patients with darker and thicker PWS.[5]

Oral manifestation reported so far in literature are commonly ipsilateral involvement of oral mucosa. Ipsilateral involvement of oral mucosa when traced embryologically is attributed to segmental development of vascular channels during embryogenesis.[16] The red macular islands on opposite side in this case can be due to aberrant vasculogenesis during embryonic development. Predominant ipsilateral mucosal staining of the palate, buccal mucosa, and tongue, interspersed with unique contralateral lingual mucosal red macular islands as noted in our case is seldom documented in literature.

4. Conclusion

The prominent skin manifestation is a key characteristic in patients with port-wine stains (PWS), leading to initial consultations with dermatologists. It is essential for the treating clinician to be aware of this distinct pattern of mucosal staining in PWS to ensure timely identification and to seek appropriate interdisciplinary advice for further management.

5. Source of Funding

None.

6. Conflict of Interest

None.

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