PACIFIC JOURNAL OF MEDICAL SCIENCES

{Formerly: Medical Sciences Bulletin}

ISSN: 2072 – 1625

Pac. J. Med. Sci. (PJMS)

www.pacjmedsci.com. Emails: pacjmedsci@gmail.com; pacjmedsci1625@gmail.com

CASE REPORT:

NEVUS FLAMMEUS: A RARE PRESENTATION WITH ORAL MANIFESTATION

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ABSTRACT:
Port-wine stain (PWS) is one of the vascular birthmarks. Port-wine stains are capillary vascular malformations, characterized by flat lesion with pink or red stain and may involve skin, soft tissue or bone. Port-wine stains on the face can be a cosmetic problem. Port-wine stain (PWS) is one of the vascular birthmarks. Port-wine stains are capillary vascular malformations, characterized by flat lesion with pink or red stain and may involve skin, soft tissue or bone. Port-wine stains on the face can be a cosmetic problem. Oral manifestations of PWS are common. Dentists often anticipate complications such as bleeding caused by the hyper-vascularity of the gingival and oral soft tissues, along with functional and cosmetic deformities from bony overgrowth involving the jaws and teeth. This article reports a case involving the oral and perioral structures in individuals with facial PWS.

Keywords: Port wine stain; Nevus flammeus; Sturge-Weber-Dimitri syndrome.
Submitted May 2018, Accepted June 2018

INTRODUCTION:

Port-wine stain (PWS) is also known as nevus flammeus. These are congenital vascular lesions that have psychological and physiological implications for the patient [1]. PWS is a hamartomous capillary malformations; it is named because of the deep red hue that appears on the skin and mucus membrane[2].These birth marks occur in 0.3 percent of the live newborns and affecting females and males equally [3]. PWS are often located on the face usually unilateral and follows the course of the trigeminal nerve. It appears at birth as a diffuse light pink macula and tends to become darker and thicker with age. Unlike hemangiomas, PWS generally do not fade over time, and do not disappear; but may expand while the child is growing [4].

The port-wine nevus, localized especially over the right side on the face, is detected in 87 to 90% of the cases. Bilateral involvement can be detected in about 33% of the cases and extension of the lesion over the middle line is observed in 50% of the patients [5].

PWS lesions are characterized histologically by ecstatic vessels and a deficiency of nerves in the papillary plexus of the skin in the affected area. These deficient nerves are of sympathetic origin, and that unchecked parasympathetic influence on blood flow through the post capillary venules results in progressive vascular ectasia [6]. Here we report the case of a male patient showing gingival changes with port-wine stain.

CASE REPORT:

A 22- year-old male reported to the dental clinic with complaints of decay in the lower teeth since 4 years. On examination diffused purple colour patch on the left side of the face was seen, extending about 3 cm below the hairline superiorly to the angle of the mouth inferiorly laterally from the left tragus, medially to the midline. Intraorally purple discolouration was observed on the mucosal aspect of upper lip, upper vestibule and gingival on the left side (Figure 1). The gingival tissue of the left upper jaw was reddish and darker than the right side with alveolar hypertrophy. The caries index was high involving almost all posterior teeth. No history of CNS disorders was reported by the patient. Patient did not have any complain about the non aesthetic facial view, and did not accept the treatment of his gingival pigmentation.

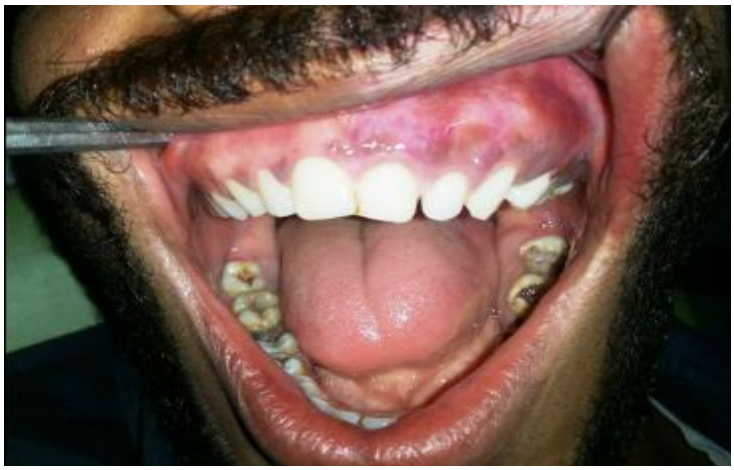

Figure 1. Intraoral view with gingival involvement on the left upper gingival region.

DISCUSSION:

Port wine stains are congenitally acquired; resulting from abnormal regulation of blood flow that leads to a progressive vascular dilation and the characteristic discolouration. These lesions are more commonly seen among Caucasians than African Americans and Asians. They usually begin as flat and pink, becoming thicker and darker over time. In advanced lesions, nodules or ‘‘cobblestones’’ may be present [7].

.In the head and neck region port wine stain usually follow the distribution of the branches of the trigeminal nerve. When the maxillary and/or mandibular divisions are involved, the bone and soft tissue of the oral cavity are often affected. This may result in complications such as bleeding caused by the hypervascularity of the gingival and oral soft tissues during dental procedures, along with functional and cosmetic deformities from bony overgrowth involving the jaws and teeth [8].Orodental manifestations includes staining of the oral soft tissues, hyperplasia of the gingiva, oral bleeding, overgrowth of the bony alveoli, and possible interruption in dental eruption sequence and lip enlargement [9].

Not all patients with facial PWS will have struge weber syndrome. Only the patients with involvement along the distribution of ocular branch of trigeminal nerve are at the risk for development of this condition [10].

Port-wine stains are associated with the following syndromes, Sturge-Weber-Dimitri syndrome characterized by noninherited and nonfamilial, port wine stain, leptomeningeal angiomas and Klippel-Trenaunay syndrome characterized by port wine stain, angiomatosis of the extremities [11].

Differential diagnosis for PWS includes nevus flammeus neonaturum pyogenic granuloma and venous varicosities [2].

Presence of port-wine stains on the face can be a cosmetic problem and usually cause deep psychological trauma to the patient and the development of personality is affected in almost all patients [12].Patients with port wine stains may experience feelings of stigmatization, embarrassments, anxiety and depression [1].

Treatment modalities for PWS include curettage, cryotherapy, dermabrasion, tattooing, chemical cauterization, electric cauterization, photodynamic therapy, intense pulsed light, spectrophotometric devices [3]. Pulsed tuneable dye laser (PDL) has become the treatment of choice. Laser therapy has been the most successful at eliminating port wine stains. It is the only method that can destroy the tiny blood vessels in the skin without significantly damaging the skin [13]. Port-wine stains can also be treated by a high dose of hydrocortisone given orally in infancy results in regression of well-localised small lesions [12].

A simple and effective technique for removal of gingival pigmentation is surgical excision of the epithelium and pigmented gingiva. Other therapeutic approaches for treatment of

gingival pigmentation such as cryosurgery, chemical abrasion, free gingival grafts, gingivectomy and laser. PWS involving gingiva does not require treatment, except for the aesthetics demands [3].

CONCLUSION:

Oral manifestations of PWS demands dentist's knowledge about this condition, associated syndrome, clinical features and treatment modalities. The consultations from various medical specialists are needed for proper treatment along with professional counselling for psychological benefit for the patient. Periodic systemic and oral examinations are recommended to prevent any cranial and oral complications.

Source of support: Nil, **Conflict of Interest**: none declared

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