Congenital Port-Wine Stains affecting Face and Neck region: A rare entity with review of literature

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Abstract
Port-Wine Stains (PWSs) are more common vascular lesions comprised of progressive ectasia of blood vessels in the dermis and it can establish either congenital or acquired in form. The exact etiology is unknown, but various factors have been proposed for its pathogenesis like trauma, drugs, tumors and herpes zoster infections in acquired PWSs. Here we report a congenital PWS in a 5-year old girl affecting face and neck region.

Keywords: Port-wine stains, Sturge-weber syndrome, Vascular malformation.

Introduction

Vascular malformation are very common in newborn infants, accounting 44%.[1] Port-Wine Stains (PWSs) are more common vascular lesions comprised of progressive ectasia of blood vessels in the dermis.[2] PWSs also known as "nevus flammeus," are cutaneous vascular malformations involving the postcapillary venules with potentially devastating physical and psychological complications,[3] 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.[2,3] 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 predilection.[4] Here we report such a rare case of congenital port-wine stains affecting face and neck region in a 5-year old girl.

Case Report

A 5-year old girl reported to the department of oral medicine and radiology with a chief complaint of red patches on the lower third of the face and neck region, since birth. Her parent's gives a history of red patches since birth and as age increases the intensity of the red patches was decreasing and also gives no history of trauma and any other systemic complaints.

On examination a well-defined asymptomatic erythematous to violaceous blanching patches seen on the lower third of the face and neck region involving right and left cheek, right ear, mental region, lower lip and neck extending approx. from right preauriclar to left preauriclar region. [Fig. 1 a and b]

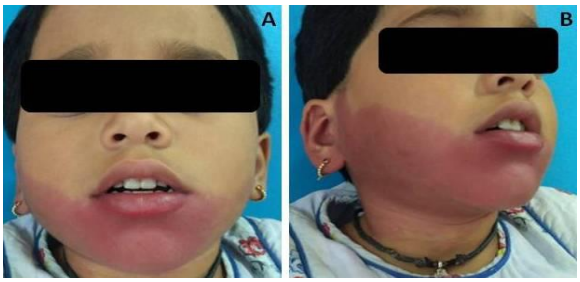

Fig. 1: Erythematous to violaceous blanching patches seen on the lower third of the face (a); neck region (b)

On intra oral examination erythematous to violaceous patches seen on the floor of the mouth, lower labial and lingual gingiva, ventral surface and tip of the tongue and right side of the palate. [Fig. 2 a, b and c]

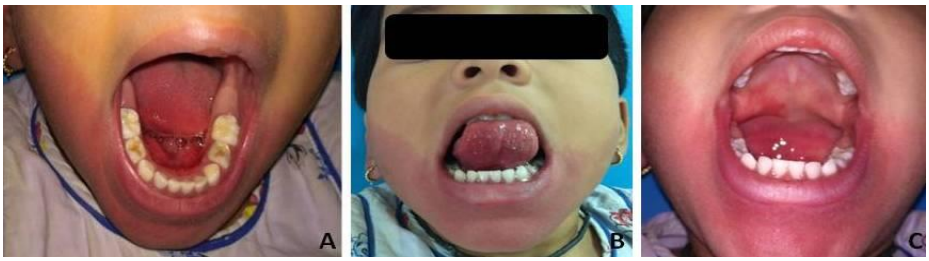

Fig. 2: Erythematous to violaceous patches seen on the floor of the mouth (a); ventral surface and tip of the tongue (b); and right side of the palate (c)

Baseline investigations are non-contributory. Based on the history and typical clinical presentation a provisional diagnosis of Port-Wine Stains was made with a differential diagnosis of arteriovenous malformations, cutis marmorata telangiectatica congenital and Sturge-weber syndrome.

Discussion

Port-Wine Stains (PWSs) are congenital vascular lesions, usually manifested in newborn infants. Clinically represented a well-defined pink to purple in colour macules. As the patient age increases it become darker, raised and nodular or cobble stone appearance.[2] In the present case, the size and colour of the lesions are decreasing as the child ages. PWS lesions are more commonly seen among Caucasians than African Americans and Asians. There is no gender predilection.[5]

The exact etiology and mechanism behind formation of these lesions remains unknown. The most But most of the authors widely accepted hypothesis proposed by Rosen and Smoller[6] stating that the congenital PWSs results from maturational defects in sympathetic nerve system, where the development of PWS is the deficiency or absence of surrounding neurons regulating blood flow through the ectatic postcapillary venules. As a result, the blood vessels are unable to constrict normally and remain permanently dilated. It is believed that PWSs develop within the first 2 to 8 weeks of gestation,[3] whereas acquired PWSs results from loss of sympathetic innervations caused by trauma. In the present case no definite cause was found for the development of the lesion and the lesion was present since birth, so considered it as congenital port-wine stains. Acquired PWSs are also reported in the literature, which manifest rarely. Various factors play an important role for the occurrence of acquired PWSs. Of these, trauma considered the most important causative factor in majority of the cases. In 1949, Fegeler is the first person to describe Trauma-induced PWS and hence named Fegeler syndrome.[4,7] The trauma-induced PWSs reported by Adams BB et al (2000) in his review, a total of 59 patients with acquired PWS, in which 17 (29%) cases found trauma to be a causative factor.[1] Few of the authors reported some of the cases induced by trauma.[8,9] Few cases of drug-induced acquired PWSs are reported in the literature by Cobb and Goldman (1990),[10] Traub (1939)[11] Hoque S et al (2005)[12] and Rose RF et al (2011)[13] which are secondary to oral medications such as isotretinoin, oral contraceptive pills, simvastatin and metformin. The reason behind drug-induced PWSs is Isotretinoin causes skin fragility and frictional trauma, while simvastain and metformin have been shown to promote angiogenesis by upregulation of vascular endothelial growth factor.[13] Pasky KA (1993)[14] reported in his study that chronic actinic exposure plays a important role in the development of PWSs. [Table 1]

Table 1: Reports of Acquired Port Wine Stains

Authors (Years)	Number of Cases	Age/Sex	Suspected Antecedent Factor
Adams and Lucky (2000)[1]	59	Mean age =	Trauma (17 cases)
Senti and Trüeb (2000)[7]	1	24+16 yrs	Trauma
Piaserico *et al*., (2004)[8]	1	--	Trauma
Hoque & Holden (2005)[12]	1	4/F	Isotretinoin
Kirkland *et al*.,(2011)[9]	1	18/M	Trauma
Rose e*t al*.,(2011)[13]	1	67/M	Simvastatin, Metformin
Bansal S *et al*., (2013)[4]	1	69/M	None
Pasky KA (1993)[14]	1	41/M	chronic actinic exposure
Present report (2018)	1	-- 5/F	None

Clinically, unilateral representations of the PWSs are most commonly seen on the face and neck region, but can occur anywhere on the body and most of the lesions persist throughout life, unlike hemangiona.[15] 50% of all facial PWSs lesions are located along the distribution of the trigeminal nerve.[1,16] The most

common orodental manifestations represented were hyperplasia of the lip (53.3%), stained gums (46.7%), malocclusion (30%), bleeding of gums (26.7%), spacing in between the teeth (23.3%), gingival hyperplasia (20%) and maxillary and mandibular hypertrophy. Orodental manifestations were more common among patients with darker and thicker PWS.[17] The Potential complications of facial port wine stains include association with neurocutaneous syndromes (Sturge-Weber syndrome), Ocular complications, Hypertrophy of the jaws, Oral involvement, and negative psycho-social impact.[15,17] In the present case orodental abnormalities not found and lesions seen bilaterally involving face and neck along the distribution of trigeminal nerve.

Histopathologically, characterized by capillaries that have multiple ectasias and dilatations, with time both increases. Vessels are generally located superficial dermis associated with loosely arranged collagen.[18]

The management of the PWSs includes surgical excision with skin grafting,[19] cryotherapy, ionizing radiation, dermabrasion, electrotherapy or tattooing. All of these have met with limited success and often left cosmetically unacceptable secondary scarring. These are no longer considered viable treatment options.[3] Recently, treatment with lasers such as argon, argon-pumped, krypton, neodymium: Yttrium-aluminum garnet, copper vapor and pulsed dye lasers. Pulsed dye laser is most preferred due to its minimal risk of scarring; as well acquired PWS tends to have a faster and better response to pulsed dye laser than congenital PWS.[20]

Conclusion

The present case report demonstrates a rare entity of a congenital vascular malformation in a 5-year old girl affecting face and neck region bilaterally along the distribution the trigeminal nerve. Although no definite cause could be appreciated for the development of lesion in our patient and presents since birth. Further reports and future insights into the pathogenesis of such cases would broaden our knowledge regarding this infrequently reported phenomenon.

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