ejpmr, 2024, 11(6), 657-659

SJIF Impact Factor 7.065

EUROPEAN JOURNAL OF PHARMACEUTICAL AND MEDICAL RESEARCH

www.ejpmr.com

Case Study

ISSN 2394-3211

EJPMR

ACQUIRED PORTWINE STAIN: A RARE OCCURRENCE: A CASE REPORT

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Article Received on 26/04/2024 **Article Revised on 16/05/2024** **Article Accepted on 06/06/2024**

ABSTRACT

Port wine stains (PWS) are congenital low-flow vascular malformations of the skin that represent a progressive ectasia of superficial cutaneous vascular plexus. Unlike haemangiomas, PWS do not involute over time, but often continue to progress and evolve into adulthood. Acquired PWSs are infrequently reported in the literature. Less than 75 cases of acquired PWS have been reported and the first case in India was reported in 2015. We report an acquired port wine stain in a 21-year-old female. The various theories, causative factors and treatment modalities have been discussed.

INTRODUCTION

Port wine stains are congenital low-flow vascular malformations of the skin that represent a progressive ectasia of superficial cutaneous vascular plexus. Over 40% of PWS are anatomically restricted to the cutaneous distribution of the trigeminal nerve. Furthermore, a strong association between PWS in the distribution of the ophthalmic (V1) branch of the trigeminal nerve and neuro-ocular pathology has been shown to exist, including Sturge–Weber syndrome (SWS).[1] Acquired PWSs are an uncommon entity developing later in life, but morphologically and histologically similar to the congenital lesions. The etio-pathogenesis of this rare entity has not been fully elucidated.

CASE REPORT

A 21-year-old female patient, presented to the dermatology OPD in our tertiary care institution with the complaint of a progressively enlarging, red coloured, asymptomatic, flat lesion over the right side of her forehead. The onset was 3 years back, from the upper part of her forehead. The lesion progressed to involve the whole width of her forehead (Figure 1). We could not elicit any history of antecedent mechanical or thermal trauma, drug intake, topical application or excessive ultraviolet exposure.

Dermatological examination revealed diffuse partially blanchable erythema extending throughout the width of the forehead on the right side. The lesion followed a dermatomal pattern involving the left V1 dermatome. On palpation, the local temperature was raised and no thrill was felt. Based on the history and clinical examination, a provisional diagnosis of acquired portwine stain and acquired elastotic haemangioma was kept. To confirm the same, a histopathological examination of the lesion was done.

On histopathology, the specimen showed a substantial increase in the number of dilated thin walled capillaries within the papillary dermis. A few capillaries in superficial and deep plexus were also dilated. Overlying epidermis was unaffected. The dermis showed sparse superficial and mid peri-vascular and peri-appendageal infiltrate of lymphocytes. The histopathological examination was suggestive of port wine stain (Figure 2 and 3).

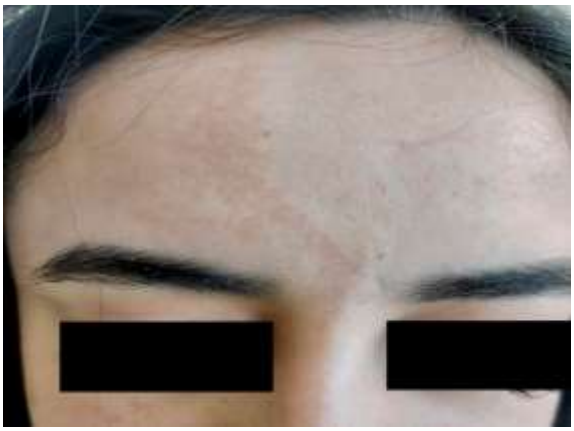
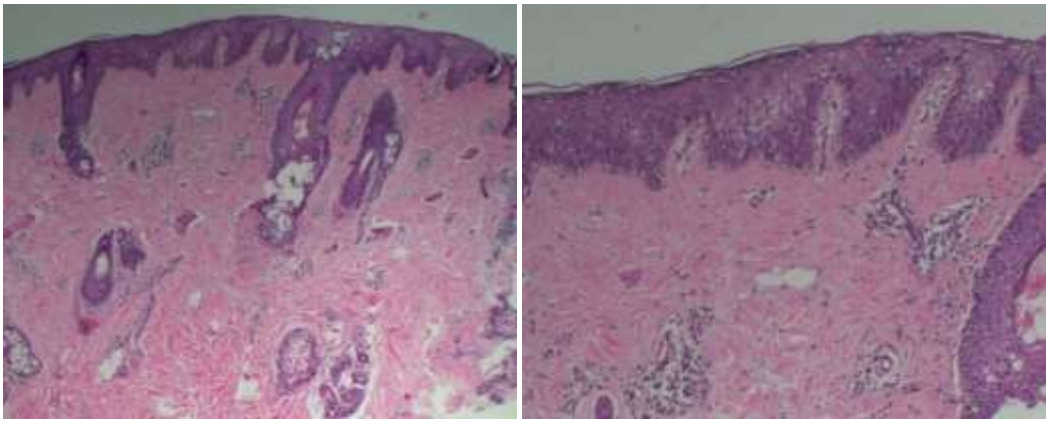


Figure 1: Portwine stain on the left side of forehead (V1 dermatome).



Figures 2 and 3: Histopathological examination of the lesion.

Overlying epidermis was unaffected. The dermis showed sparse superficial and mid peri-vascular and peri-appendageal infiltrate of lymphocytes. This was accompanied with a substantial increase in the number of dilated thin walled capillaries within the papillary dermis. A few capillaries in superficial and deep plexus were also dilated.

DISCUSSION

PWS are congenital vascular malformations usually presenting at birth in the form of pink-red to purple macules, which become darker, raised, and nodular as the person ages. Unlike haemangiomas, PWS do not involute over time, but often continue to progress and evolve into adulthood. Various theories have been hypothesized to explain this phenomenon.[1] The most accepted hypothesis is a defective embryological maturation of the sympathetic fibres, resulting in a loss of normal sympathetic control of the cutaneous vessels which leads to ectasia. Both acquired and congenital PWSs were the result of malformed sympathetic innervation. In the case of congenital PWS, there was a maturational defect in the local sympathetic nervous system, whereas loss of sympathetic innervation, possibly through trauma, could lead to acquired PWSs. It has also been shown that the vessels within a PWS do not react normally to vasoactive stimuli, further supporting the role of altered autonomic innervations. Some studies have implicated alterations in the surrounding supportive dermal structure, without vessel wall abnormalities. Rosen*et al.*, compared nerve densities in normal skin and PWS lesional skin and found significantly fewer nerves in the skin with a PWS. Discrepancy in density leads to altered neural modulation of vascular tone, thereby ultimately affecting the orientation and vasoactive properties of the vessels.[2] Trauma may lead to perivascular atrophy, which ultimately leads to vessel dilatation. Alternatively, the reparative process in vessels, after trauma, may proceed abnormally, resulting in dilated vessel walls.[4] A more recent study by Hibler*et al.*, has suggested that PWSs are due to intracranial circulation abnormalities and may result in cutaneous findings, implying that SWS is a product of "acquired venous obstruction rather than neural dysfunction."[4]

Acquired PWSs are infrequently reported in the literature. Various factors have been proposed for the occurrence of acquired PWS. Of these, trauma has been found to be the most prevalent causative factor in most of the cases. Trauma-induced PWS was first described by Fegeler in 1949 and thus, was named Fegeler syndrome. Few cases of acquired PWS have also been proposed secondary to oral medications like isotretinoin, oral contraceptive pills, simvastatin, and metformin. Isotretinoin causes skin fragility and frictional trauma, while simvastain and metformin promote angiogenesis by upregulation of vascular endothelial growth factor.[5] Chronic actinic exposure may also lead to acquired PWS. Isolated cases of acquired PWS following frostbite injury, obstruction of the peritoneovenous shunt, herpes zoster infection, cerebral arteriovenous malformation, spinal root compression, and solid brain tumor have also been reported.

Their presence near the eye or in the V1 or V2 distribution warrants an ophthalmology workup to rule out any associated malignant or other potentially serious sequelae.[4] This case report sheds a light on a rare dermatological presentation in the form of an acquired condition.

Figures

1. Portwine stain on the left side of forehead (V1 dermatome)
2. Overlying epidermis was unaffected. The dermis showed sparse superficial and mid peri-vascular and peri-appendageal infiltrate of lymphocytes. This was accompanied with a substantial increase in the number of dilated thin walled capillaries within the papillary dermis. A few capillaries in superficial and deep plexus were also dilated.

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