A rare bilateral presentation of port wine stain of the face and neck

Yüz ve boynundaki doğum lekelerinin yaygın olmayan iki taraflı yerleşimi

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Abstract

Structural anomalies of blood vessels without endothelial proliferation are termed ‘vascular malformations’. These malformations are present at the time of birth and continue their presence throughout the life of the individual. Most of these Port wine stains are unilateral in nature or typically showing segmental distribution that respects the midline. Although Port wine stains are considered to be common capillary malformation, their presentation bilaterally is a rare occurrence. Here we report a case of a rare bilateral presentation of Port wine stain involving both the face and neck, extending from the hairline on the scalp to the angle of the mouth and tragus of the ear on both the sides.

Key words: Hemangioma, port wine stain, vascular malformation

INTRODUCTION

Vascular malformations are structural anomalies of blood vessels without endothelial proliferation. By definition, vascular malformations are present at birth and persists throughout life. They can be categorised according to the vessel involved (capillary, venous, arterial) and according to hemodynamic features (low flow or high flow). Port wine stains are relatively common capillary malformations that occur in 0.3% to 1% of the newborns. They are most common on the face particularly along the distribution of the trigeminal nerve¹. A port wine stain is defined as a macular telangiectatic patch which is present at birth and remains throughout life. They may be localized or extensive. It may affect a whole limb. Port wine stain is often associated with an underlying disorder. These are best considered as low-flow vascular malformations that may occur on any part of the body but commonly affect the face in the distribution of the trigeminal nerve. Initially, the lesions are pale pink patches. Eventually they evolve into a violaceous colour. They can remain static or even lighten. Facial port wine stain typically evolve into thicker areas with vascular blebs, occasionally pyogenic granulomas and underlying tissue hypertrophy². This paper documents a case of Port Wine stain crossing the midline and involving both the left and right side of face and neck.

CASE

A 19 year old male patient reported to the Department of Oral Medicine and Radiology with a chief complaint of forwardly placed teeth since the time of the eruption of his permanent teeth. History revealed that he had visited an Orthodontist for the
same problem previously. He has difficulty in chewing food and closing his lips. Medical history revealed that the reddish discoulouration of the face was present since birth. It was initially lighter in shade which eventually turned darker and has reached the present shade. He also was epileptic during childhood. He was treated for his epilepsy and right now is not under any medication. No known drug allergy is reported. The patient has never been treated for the discolouration.

Figure 1. Clinical photograph showing Port wine stain bilaterally involving the face and neck with the presence of papulo-nodular lesion on the left lateral and right lateral region of forehead.

On extraoral examination there was reddish discoulouration of the skin bilaterally on the face extending from the hairline on the scalp superiorly to the region above the line joining the angle of the mouth to the tragus of the ear on both the sides, laterally involving the auricular and post auricular region. On the dorsal side of the head and neck region the reddish discoulouration is seen involving the neck on both the sides. On the face there is a small patch of normal skin in the frontal region measuring approximately 3cm x 2cm. A single papulo-nodular lesion was seen overlying the reddish discoulouration on the right forehead region measuring approximately 2cm x 2cm in dimension. Two papulo-nodular lesions were seen on the left side in the region of the forehead measuring 1cm x 1cm and 0.5 x 0.5 cm respectively (Figure 1). All the growths were reddish in colour, soft to firm in consistency and non tender with no evidence of surface discharge or ulceration.

Figure 2. Clinical photograph of Port wine stain on the sclera, Bulbar conjunctiva and palpebral conjunctiva of the left and right eye.

Sclera, Bulbar conjunctiva and palpebral conjunctiva on the left and right eye was involved (Figure 2). On intraoral examination, reddish discoulouration of the labial mucosa, buccal mucosa bilaterally and soft palate was seen. There was decay w.r.t 16, 17, 27, 36 and posterior open bite on the left side (Figure 3). Blanching was observed on diascopy (Figure 4). Based on the classical clinical features a provisional diagnosis of Port Wine stain was made. Panoramic radiograph, lateral cephalogram and a postero-anterior view of the skull was done to rule out intracranial extension that eliminated possibilities of a syndromic association. Radiographic examinations showed no gross abnormalities (Figure 5).

Figure 3. Port wine stain involving labial mucosa, buccal mucosa and soft palate.

Figure 4. Photograph showing blanching on diascopy.
DISCUSSION

Vascular malformations are errors of morphogenesis whereas hemangiomas and other vascular tumors grow by cellular proliferation. Vascular anomalies are common birthmarks. A classification system, first proposed by Mulliken and Glowacki was revised in 1996 by the International Society for the Study of Vascular Anomalies based on clinical, radiological and hemodynamic characteristics, into vascular malformations and vascular tumors. A port wine stain is defined as a macular telangiectatic patch which is present at birth and remains throughout life. They represent hamartomatous capillary malformations and are named so due to the deep red hue that they leave on the skin or mucosa. These are best considered as low-flow vascular malformations which may occur on any part of the body but commonly affect the face in the distribution of the trigeminal nerve. Initially, the lesions are pale pink patches, but with time they may mature into a violaceous color, remain static or even lighten and may become nodular because of vascular ectasia (cobblestone formation). Port wine stains are associated with the following syndromes. It is very essential to differentiate the occurrence of isolated Port Wine stain with Sturge-Weber-Dimitri syndrome. This syndrome is characterized by noninherited and nonfamilial port wine stain and ipsilateral leptomeningeal angiomatosis leading to cerebral calcifications, epileptic convulsive crisis, contralateral hypertrophy, hemiparesis and mental retardation. Another syndrome associated with Port Wine stain is Klippel-Trenaunay syndrome characterized by port wine stain, angiomatosis of the extremities.

Several treatments options have been proposed. Pulsed tuneable dye laser (PDL) has become the treatment of choice. Laser therapy has been the most successful among all the treatments. It is the only method that can destroy the tiny blood vessels in the skin without significantly damaging the skin. Cryosurgery may be used to correct lip and other soft tissue deformities. Port wine stains on the face can be a cosmetic problem and should be treated by a plastic surgeon. High dose of hydrocortisone given orally in infancy results in regression of well localized small lesions.

Over time, as the blood vessels become more and more dilated, they become more susceptible to spontaneous bleeding or hemorrhage following minor trauma. Bleeding can be difficult to control, may necessitate hospitalization, and may also increase the likelihood of skin infection. The hypertrophy (increased tissue mass) of underlying soft tissue that occurs in approximately two-thirds of lesions further disfigures the facial features of many patients. For all of the above reasons, most medical specialists agree that it is essential to begin treatment of Port wine stains as early as possible and to maintain treatment to prevent the development of vascular nodules and hypertrophy in later years.

Port Wine stains are a common variety of vascular malformations, but, it generally affects the face in the distribution of trigeminal nerve. The occurrence of Port Wine stain crossing the midline involving the skin bilaterally, the eyes, and the neck is rare. Since Port Wine stains are associated with syndromes it is essential to elicit a detailed medical history, do a thorough clinical examination and radiographic evaluation to rule out such possibilities. Port wine stains also cause cosmetic problems to the patients. As the patient grows, the possibility of increased bleeding tendencies, permanent disfigurement due to hypertrophies can be anticipated. Early recognition and prompt treatment can help the patient to lead a relatively normal and healthy life.

![Port Wine stain of face and neck](image)

**REFERENCES**

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