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Case Study

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PORT WINE STAIN WITH NODULOSIS: A CASE REPORT

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ABSTRACT

Port wine stains (PWS) are congenital vascular malformations with dilated capillaries or venules in dermis. It is a manifestation of vascular ectasia that can develop nodules or thickening with time. Thickening is due to further dilatation of already ectatic vessels whereas nodules arise due to vascular hyperplasia or neoplasm.

INTRODUCTION

Port-wine stains are congenital cutaneous capillary malformations that usually present on head and neck region. Its incidence ranges from 0.3-0.5% among newborns. It does not have any gender predisposition. Usually, the lesions are limited upto dermis but may affect the underlying subcutaneous tissue and bone, and extend on to adjacent mucous membrane and conjunctiva. If left untreated, they can darken in color, expand or regress over time. In severe cases, they can form nodules or thicken causing cosmetic disfigurement. Port-wine stain has favourable prognosis with various treatment modalities.

CASE REPORT

A 62-year-old female came to dermatology department for port-wine stain on left side of her forehead since childhood. Over time after age of around 40 years, lesions started to darken in color, thicken and developed nodules on previous flat PWS. It neither involved any mucosa nor had any other neurological deficits associated.

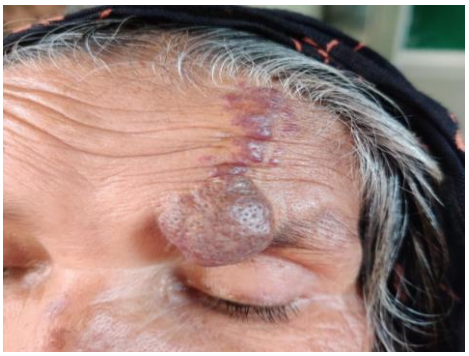

Figure 1: Female patient with portwine stain and thickening and nodulosis on the right side of forehead.

DISCUSSION

Portwine stain is a common vascular malformation with benign nature and is generally flat. It is a capillary malformation in dermis. With time, certain changes namely darkening, thickening and nodules can develop on PWS. Pathomechanisms behind these changes are largely unknown. Further dilatation of capillary leads to thickening and nodule formation. One report suggested that PKCα and PI3K signalling pathways contribute to the development of hypertrophy and nodularity.[1]

Portwine stain is typically first treated with propranolol with variable results. Pulsed dye laser has been proven to be a highly effective and safe treatment method but its results in adults are also variable. Pulsed-dye laser (PDL) is the gold standard for treating PWS patients, but at present the main modality of choice is hemoporfin photodynamic therapy (PDT).[2] In PWS with thickening and nodulosis, photodynamic therapy and pulsed dye laser both have been used successfully. However, in severe cases, surgery may also be combined with these treatment modalities for better outcome. Because of financial issues, our patient denied any kind of treatment modality.

Early treatment should be recommended to people born with PWS in order to avoid hypertrophy and nodulosis to prevent any significant threat to physical and psychological health. For future perspectives, studies involving histological and histochemical analysis can be done for better understanding of pathogenesis which can influence direction for better treatment options for PWS.

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