

Nevus Flammeus

Nevus flammeus, also known as a port wine stain (PWS), is a birthmark caused by a capillary malformation in the skin. It usually appears as a flat, pink to red patch at birth, and the lesion is typically painless. Over time, the lesion may darken, become thicker, and even form nodules. These lesions are non-blanchable, meaning they do not fade when pressed.

Port wine stains can appear anywhere on the body, but they are often found on one side (unilateral) or in a specific area (segmental). When they appear on the face, they often follow the path of the trigeminal nerve, and sometimes the lesions can extend to the mucous membranes, like the mouth.

Clinical Features and Progression

Nevus flammeus typically appears at birth as a flat, pink to red lesion, caused by dilated capillaries that increase blood flow in the affected area. Over time, the lesion often darkens and can thicken. In some cases, nodular changes may develop, further affecting its appearance.

The lesion does not regress on its own, and it tends to grow larger as the individual ages. When located on the face, especially along the areas of the trigeminal nerve, it is important to closely monitor the lesion. This is because it can sometimes lead to complications, such as ocular involvement or an increased risk of glaucoma.

Differential Diagnosis and Syndromic Associations

While nevus flammeus often occur on their own, they can sometimes be part of a syndromic condition, which may require further investigation. Some of the most common syndromes linked to port wine stains include:

- **Sturge-Weber Syndrome:** This condition is marked by a port wine stain, especially in the V1 (ophthalmic) branch of the trigeminal nerve. It is often accompanied by neurological issues, like seizures and developmental delays.
- **Klippel-Trenaunay Syndrome:** A rare disorder characterized by port wine stains, varicose veins, and soft tissue growth.
- **Parkes-Weber Syndrome:** A variant of Klippel-Trenaunay syndrome with abnormal connections between arteries and veins.
- **Servelle-Martorell Syndrome:** Characterized by one side of the body growing larger, venous malformations and port wine stains.

- **Proteus Syndrome:** A rare condition causing overgrowth of various tissues, including skin, bones, and fat, along with port wine stains.
- **Bannayan-Riley-Ruvalcaba Syndrome:** Associated with multiple benign growths, intellectual disabilities, and port wine stains.
- **CLOVES Syndrome:** A rare overgrowth syndrome with characteristic features of port wine stains, vascular malformations, and lipomatous overgrowths.

If a person has multiple port wine stains, particularly if they follow a segmental or dermatomal pattern, it is important to consider these syndromes and seek further evaluation.

Diagnosis

The diagnosis of nevus flammeus is primarily clinical, meaning it's based on a careful history and physical examination. The typical appearance of the lesion, its location, and how it changes over time are key to making the diagnosis.

In cases where the lesion might be associated with a more complex syndrome, additional tests such as imaging may be necessary. For example, if there are concerns about Sturge-Weber Syndrome, brain imaging (like an MRI or CT scan) may be ordered to look for any neurological issues. Moreover, if the port wine stain involves the eyelids or the area around the eyes (periocular region), it's important to refer the patient to an ophthalmologist. This is because glaucoma can be a complication of Sturge-Weber syndrome, and early detection is crucial for managing potential eye issues.

Treatment Options

While nevus flammeus is benign, its persistent and progressive nature often leads to concerns regarding cosmetic appearance and possible functional impairment. Various treatment options are available, with laser therapy being the gold standard for management.

- **Pulsed Dye Laser (PDL):** The pulsed dye laser is the most commonly used and most effective treatment for port wine stains. It works by targeting the blood vessels in the lesion with a specific wavelength of light, causing the blood vessels to constrict and the lesion to fade. This treatment can be particularly effective in preventing the thickening and nodularity that often occurs over time.
- **Laser Therapy with Nd:YAG Laser:** In certain cases where deeper vascular lesions are present, an Nd:YAG laser may be employed to penetrate more deeply into the skin and target larger or more persistent vessels.
- **Cryotherapy and Electrocautery:** These methods, which involve freezing or burning the lesion, are generally not recommended for nevus flammeus. These modalities can lead to significant scarring and tissue damage, making them less suitable than laser treatments.

- **Surgical Excision:** Surgical excision is rarely used for port wine stains due to the potential for scarring and the tendency for the lesion to recur. It is generally reserved for cases with small, well-localized lesions that are refractory to laser treatment.

Prognosis and Follow-Up

The prognosis for individuals with nevus flammeus is generally good, especially with early treatment. Starting laser therapy early can greatly reduce the appearance of the port wine stain and prevent it from becoming thicker or more raised. Regular follow-ups with a dermatologist are recommended to track the progress of treatment and manage any potential complications, like the development of additional lesions or vascular malformations.

For individuals with nevus flammeus as part of a syndromic condition, it's important to closely monitor other related health issues. For example, those with Sturge-Weber syndrome may need regular neurological checkups, and ophthalmologic evaluations should be done to screen for glaucoma.

Conclusion

Nevus flammeus, also known as port wine stain, is a congenital skin condition caused by a vascular malformation. While generally harmless, it can lead to cosmetic concerns as it may enlarge or develop nodular changes over time. Though typically benign, nevus flammeus can be linked to certain syndromes, making thorough evaluation important. Pulsed dye laser therapy is the most effective treatment, helping to prevent the lesion from thickening or darkening. Regular follow-ups and referrals for any related health concerns are essential for ensuring the best care for individuals with this condition.

References

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