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Research Article

MODERN APPROACH TO THE DIAGNOSIS AND TREATMENT OF FLAMING NEVUS

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ABSTRACT

Capillary malformations (capillary angiodysplasia, CAD) in patients are an urgent problem of modern medicine due to functional, cosmetic, psychological disorders and social maladjustment of this category of patients in the absence of timely adequate treatment.

KEYWORDS

Flaming nevus, pathogenesis, neurotrophic factors, diagnosis, laser treatment, prognosis, malformation, mutation, percutaneous laser coagulation, capillary angiodysplasia.

INTRODUCTION

Transcutaneous laser photocoagulation (selective photothermolysis) is currently the "gold standard" for the treatment of port-wine stains. However, it is extremely rare to achieve complete removal of a vascular lesion, and resistance to laser exposure is noted in 20% of cases. The article presents clinical cases of the treatment of port-wine stains in two patients. Based on our own experience and literature data, we analyzed the results of treatment, side effects of the

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use of lasers, and also considered alternative promising methods of treatment.

A flaming nevus is a capillary angiodysplasia that occurs in the embryonic period of prenatal development. Occasionally, an acquired form of nevus is observed, associated with injuries, past infections, and taking medications. A flaming nevus has the appearance of a bright pink, red or purple spot of various sizes and bizarre shapes. It is predominantly located on the face, neck, and upper torso.

THE MAIN RESULTS AND FINDINGS

In the medical literature, two synonymous concepts are used: a flaming nevus (nevus flammeus) and a wine stain (port wine stains, PWS), which describe the same congenital anomaly of development. Pathology occurs in 0.3-0.5% of newborns, no significant gender, ethnic and racial differences were found.

The intensity of the color can change during the day, for example lighter in the morning, darker in the evening. The brightness of the spot increases with a rise in body or environmental temperature, during exercise or feeding a child. But these changes are reversible: when the effect of the irritating factor stops, the stain acquires its usual color.

As a result of the pathological expansion of capillaries on the skin of a person, an irregularly shaped spot appears, the color of which varies from light pink to purple, burgundy and violet. Most often, a flaming nevus is located on the skin of the face and neck, but in some cases it can be localized in other parts of the body. As a person grows older, the port wine stain increases in size.

Wine birthmark is not able to degenerate into a malignant tumor, so it does not pose a direct danger to the patient's life. However, it must be borne in mind that a flaming nevus can lead to a number of problems, so it must be treated.

Introduction. All vascular malformations of the skin are the result of a violation of the formation of the embryonic vascular system from the 4-8th to the 19-20th week of embryo development. They exist from birth, although they may not appear for a long time. The most common capillary malformations are described in the literature as port-wine stains, flaming nevus, capillary malformations, and capillary angiodysplasias (CAD). The prevalence of CAD is 3-5 cases per 1000 newborns; distinctive features - a large number of abnormally dilated blood vessels without signs of endothelial proliferation in the papillary and partially reticular layers of the dermis, do not undergo spontaneous involution and progress with age. The size of the spots varies from a few millimeters to half of the body surface, the area increases in proportion to the growth of the child, about 80% of the lesions occur in the head and neck region. Capillary angiodysplasia can be part of a number of pathological conditions, the most common are Sturge-Weber and Klippel-Trenaunay syndromes.

Characteristic external signs of a flaming nevus: Wine stains on the skin have an irregular shape, reminiscent of a geographical map. As a person grows older, their area proportionally increases.

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Color - from pinkish to maroon. They may turn blue with age. When lightly pressed with a finger, the spots lighten, then darken again.

In the initial stages of development, a flaming nevus does not cause physical discomfort; itching and inflammation are absent.

Over time, the surface of the nevus may become uneven, with convex nodules – angiomas.

Classification and stages of development of capillary malformation

The most convenient classification of vascular anomalies was created by the International Society for the Study of Vascular Anomalies (ISSVA). It is constantly supplemented with new nosologies and refined. Now in this classification the following types of capillary malformation are distinguished:

simple;

combined with other types of vascular anomalies: capillary-venous, capillary-lymphovenous, capillary-arteriovenous;

as part of syndromes, such as Sturge-Weber syndrome and Klippel-Trenaunay syndrome. The first syndrome is accompanied by damage to the skin, eyes, nervous system and internal organs. It is characterized by the presence of capillary malformation on the skin of the face, angioma of the choroid and pia mater. Klippel-Trenaunay syndrome is a rare congenital disorder in which the formation of blood and lymphatic vessels, skin, muscles, and bones is disrupted. It is accompanied by the appearance of a port-wine stain, malformations of the veins, proliferation of soft tissues and bones.

Etiology. There is no exact data regarding the causes of this pathology in modern medicine.

Genetically, the formation of wine stains is associated with capillary malformations. Patients with flaming nevus often have defects in the GNAQ6, RASA16, EPHB4, and PIK3CA genes. Genetically proven connection of the formation of malformations with the following syndromes:

Sturge-Weber syndrome - the location of port-wine spots on the face along the branches of the trigeminal nerve. In addition to skin symptoms, vascular anomalies of the brain and eyes occur, and atrophy of the nervous tissue is possible.

CLOVES syndrome. Pathology is manifested by multiple capillary, venous and lymphatic dysplasia. They are associated with epidermal nevi. Patients have lipomatosis, scoliosis, lesions of bones and joints.

Proteus Syndrome. Vascular malformations are combined with abnormally rapid and excessive growth of certain parts of the body, namely in the area of the palms and soles.

CLAPO syndrome. Capillary dysplasias are located in the region of the lower lip, perioral zone. Pathology is accompanied by macroglossia, deformation of facial features and limbs.

Klippel-Trenaunay syndrome. A distinctive feature of the flaming nevus with this defect is its location on the lower extremities. Pathology is accompanied by varicose veins in the legs, hypertrophy of bones and soft tissues.

The acquired flaming nevus, which was first described by the German dermatologist F. Fegeler in 1949, is distinguished into a separate category. Wine stains occur after trauma to the cervical or thoracic spine. Pathology can manifest itself at any age. Later, other triggers of acquired PWS were established: frostbite, hormonal and lipid-lowering drugs, and herpes zoster.

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Pathogenesis. The formation of congenital pathology falls on the 5-10th week of the prenatal period. In pathogenesis, two key hypotheses are distinguished: impaired vascular innervation and genetic mutations of angiogenesis. It has been established that the number of S-100 nerve fibers is sharply reduced in pathological dermal vessels. This causes a decrease in basal vascular tone and a decrease in the number of neurotrophic factors - the main causes of abnormal vascular growth.

Abnormal expression of arterial (EfnB2) and venous (EphB1) markers occurs in abnormal skin vessels. Such processes cause disturbances in the primary differentiation of dermal arterioles and venules, and contribute to excessive capillary growth. In the pathogenesis of the disease, pathological activation of the signaling pathways of enzymes is involved: activated protein kinases, phosphoinositide-3-kinases.

Pathologically, a flaming nevus is characterized by dilated capillaries and post-capillary venules, which are located in the papillary layer and the upper part of the reticular dermis. Signs of endothelial proliferation and cellular atypia are absent. Gradually, the number of ectatic vessels increases, and perivascular fibrosis progresses.

Diagnosis requires a clinical examination, Doppler ultrasound, MR angiography, and computed tomography. The decisive role in the diagnosis is played by the congenital nature of the spot, its typical localization and wine color. It is necessary to take into account the burden of the anamnesis with the genetic manifestations of the disease.

Doppler ultrasound. Ultrasound diagnosis of the affected area of the body is necessary to assess blood flow in pathological vessels, to exclude more dangerous arteriovenous malformations that can mimic a flaming nevus.

MR angiography. MRI can determine the presence of a vascular component in soft tissues. In the case of a simple capillary malformation, such changes are insignificant, so the information content of such a study is minimal. Given that in young children this study is performed under anesthesia or sedation, routine MRI is not recommended for children. Indications for MRI are: combined forms of vascular malformations and syndromic forms of the disease, such as Sturge-Weber syndrome.

CT scan. The study is carried out with malformations in order to assess the structure of the connective tissue, to identify pathologies of the musculoskeletal system. With concomitant neuralgic symptoms, a CT scan of the brain is indicated.

tissue biopsy. Sampling of biomaterial is required for differential diagnosis between congenital and acquired forms of port wine stain, for genetic research as part of the diagnosis of a hereditary syndrome.

Treatment of a flaming nevus. To date, laser exposure methods are used on the skin much more often than on any other tissues, due to the exceptional diversity and prevalence of skin pathology and various cosmetic defects, as well as the relative ease of performing procedures, which is associated with the superficial location of objects requiring treatment.

Due to its prevalence and pronounced cosmetic defect, flaming nevus is an important problem in practical dermatology. The efforts of specialists are aimed at developing minimally invasive and effective ways to remove stains.

Wine stains are subject to aesthetic correction by laser therapy. The effectiveness of treatment depends on the age of the patient, the size, number and depth of the vessels. The optimal age for the first procedure is

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from 3 months to 6 years. In this period, it is possible to achieve lightening by 55% in one procedure, while with age, the effectiveness of laser coagulation is 1.5-2 times lower.

The technique of laser coagulation with a pulsed laser with a wavelength of 595nM shows the maximum effectiveness in superficially located neoplasms. Wine stains on the lateral surfaces of the face are best corrected, since the vessels are located in the papillary layer of the dermis. Nevi in the central part of the face and on the body go away more slowly, require a long course of laser therapy.

20% of patients with giant and nodular vascular malformations show resistance to standard laser treatment. In such a situation, alternative hardware procedures are used: exposure to intense pulsed light (phototherapy), alexandrite and neodymium lasers. To achieve a lasting result, you will need about 10 sessions with intervals of 2 to 6 weeks between them.

Patients and methods. On the basis of the Department of Dermatovenereology of the Tashkent Medical Academy, 230 patients with CAD aged from 2 weeks to 36 years received treatment with a pulsed dye laser with a wavelength of 595 nm. Light waves emitted by laser devices, together with the use of a dye, penetrate the epidermis and are absorbed in the dermis by the target chromophore, in this case, hemoglobin. The peak absorption of a light pulse by hemoglobin depends on the degree of its saturation with oxygen. So, for oxyhemoglobin, the peak absorption is 577 nm, for carboxyhemoglobin - about 585 nm.

Wine stains are located on the right half of the woman's face.

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Wine stain on the right side of the neck.

(a) Before starting treatment. (b) After 5 sessions of laser therapy performed 1 month apart.

The final improvement was 70%.

Results. Analysis of the results of laser therapy showed that the total frequency of "excellent" and "good" effect of CAD lightening was 83%. Satisfactory results were observed in 18.9% of patients, unsatisfactory in 0.6%. Lack of response to treatment due to deep vascular involvement was noted in one patient with Klippel-Trenaunay syndrome. Complications are not registered.

A positive correlation was found between the skin phototype and the effect of laser treatment ($r=0.46$; $p<0.05$). In 96 and 86.3% of patients with phototypes I and II, respectively, the degree of clarification of the CAD zone exceeded 50%. In the vast majority (82.1%) of patients with skin phototype III, the regression of malformations was only 25-49%.

In addition, a significant positive correlation was found between the localization of the CAD and the degree of its lightening as a result of laser treatment ($r=0.56$; $p<0.05$). Laser exposure is especially effective in the projection of the 1st branch of the trigeminal nerve, for example, the forehead, and the 2nd branch, for example, in the area of the zygomatic bone. It should be emphasized that in all patients who received laser treatment of CAD in the facial area, the degree of clarification of the malformation exceeded 25%, there were no failures.

Of the side effects of laser treatment of CAD, it is necessary to note the appearance of temporary gray-blue spots - purpura or bruises at the site of laser exposure and edema of varying severity. In this study,

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we observed them in all 230 patients. The purpura persisted for 7-10 days.

CONCLUSION

Our own research results have shown high efficiency of CAD laser therapy in patients. An integrated approach to the diagnosis and treatment of patients with vascular malformations based on large medical institutions ensures individual treatment regimens and the effectiveness of the method.

Forecast and prevention. Laser coagulation can significantly lighten the flaming nevus, which has a positive effect on the appearance and psychological comfort of patients. With an isolated vascular anomaly, the prognosis is favorable. With a combination of vascular spots with genetic syndromes, the prognosis is determined by the severity of somatic symptoms. Measures to prevent the disease have not yet been developed.

CONCLUSION

"Port-wine stains" are a set of dilated capillaries of various diameters, occurring at different depths. The thickness of the epidermis over the vascular lesion and the thickness of the papillary dermis are different in different parts of the body. In addition, in the pathogenesis of this pathology, there is a violation of the innervation of blood vessels. Therefore, the use of "vascular" lasers for the treatment of this pathology has its own difficulties and nuances.

Despite the fact that laser photocoagulation remains the only effective method for the treatment of port-wine stains, new technologies are being sought to improve treatment results. The use of devices with different wavelengths (755 nm, 810 nm, 1064 nm), the use of epidermal cooling can in some cases solve the problem of PWS resistant to ILK 595 nm.

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