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ScienceDirect
Medical Laser Application 24 (2009) 27–51
Medical Laser Application
www.elsevier.de/mla

Vascular tumors and malformations of the pelvic and genital region – Classification and laser treatment

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Received 22 October 2008; accepted 5 November 2008

Abstract

Hemangiomas in infancy and vascular malformations (VM) such as port-wine stains, venous, arteriovenous, lymphatic or mixed malformations of the anogenital region may present considerable problem for patients at different stages of their individual development. A clear differentiation between the different entities is important for the choice of the proper therapy (from none to immediate intervention). Whereas hemangiomas are vascular tumors which arise during the first weeks of the newborn period, VM may be present at birth or be recognized later, but usually show little or no progress in infancy. Their growth tendency is related to the vessel types involved but is mainly influenced by the anatomical and functional situation of the malformed and ''normal'' regional vessels involved. Both types of vascular anomalies may present an ''iceberg phenomenon'' where the clinically visible portion is only a small part of the whole picture.

As hemangiomas show a spontaneous regression, the treatment is aimed at preventing complications such as ulcerations and excessive growth which could lead to secondary problems, e.g. in urine flow or defecation and infection. Treatment will usually be completed during the first year of life, employing laser treatment, corticoid medication and sometimes surgery.

The treatment of VM is symptomatic. Complete healing is usually not possible with non-mutilating strategies; long-term recurrences are frequent. Swelling, bleeding and obstruction may occur during adolescence or later (e.g. during pregnancy) and usually mark the onset of therapy. An individualized and effective therapy requires a complete understanding of the anatomy of the malformation and employs various techniques such as laser sclerotherapy and surgery, with either a direct or endoscopical approach.

Keywords: Laser; FPDL; KTP; Pulsed Nd:YAG laser; Nd:YAG laser; Diode laser; Hemangioma; Vascular malformation; Angiodysplasia; Selective photothermolysis; Interstitial; Intraluminal

Introduction

A clear classification of vascular anomalies is necessary in order to choose the right therapeutic option as different types of angiomas require different treatment strategies. This article consists of a short introductory classification and therapeutic guidelines for the treatment of both hemangiomas and vascular malformations (VM), with special emphasis on laser therapy.

The clinical appearance of hemangiomas, other vascular tumors and VM with their individual colors, shapes, sizes and vascularization types varies within a

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1615-1615/$ - see front matter © 2008 Elsevier GmbH. All rights reserved.
doi:10.1016/j.mla.2008.11.003

wide range and it is for this reason, that there cannot be only one laser for all different types.

To choose the right system, it is imperative to properly categorize the "angioma" and to determine which laser effect is desired. According to the different clinical appearance, the correct laser type with its specific and characteristic tissue effect has to be chosen for the best treatment and the most satisfying results. This may vary through different treatment steps. Furthermore, the patient's skin type and the localization of the lesions have to be considered.

Vascular tumors – classification and typical clinical appearance

Infantile hemangiomas

Hemangiomas in infancy, also referred to as infantile hemangiomas (IH), are the most common tumors in the postnatal phase. They are histologically classified as proliferating embryonic tumors that possibly stem from placental tissue or resemble it (they are GLUT 1-positive). Their clinical classification refers to the stage, localization and growth pattern. They are present at birth in 2–3% of newborns, and at the end of the first year of life, are present in about 10% [1]. Hemangiomas also occur more frequently in female than in male infants, with a female-to-male preponderance of between 3 and 5:1 [2].

The head and neck region is the site of predilection for IH (60–70% of all cases), but hemangiomas can appear anywhere on the body surface. Hemangiomas may involve the mucous membranes of oral and genital regions. In this case, and the case of multiple appearance ($n > 7$) associated deeper internal lesions have to be excluded. Anogenital hemangiomas are not frequent but are usually cumbersome as they tend to ulcerate and cause secondary problems or may be associated with further anomalies.

IH usually develop on the skin with a typical coloration ("strawberry") and subcutaneously with a progression in typical stages [3]. The cutaneous portion often precedes the subcutaneous part, which in fortunate cases can be entirely lacking. IH of the subcutaneous space are usually recognized later by an elastic swelling with a bluish shine.

All hemangiomas develop in typical phases.

Growth phases of infantile hemangiomas

- *Prodromal phase*: At the prodromal phase of development, the tumors are highly cellular and characterized by plump endothelial cells that line vascular spaces with small inconspicuous lumina. Color-coded duplex sonography (CCDS) often displays only a diffuse hyposonic structure, lacking any visible capillarization.
- *Initial phase*: In the early or initial phase, infantile hemangiomas may partially appear within a few days. Depending on the type of growth, limited or infiltrative (see below), they are diffuse, infiltrating the surrounding tissue or in case of limited growth are sharply demarcated. The latter frequently clearly protrude from the skin, are light reddish in color and shine brightly, causing parents to show these to a physician more readily than the infiltrative ones. In CCDS, often only a diffuse hyposonic structure will be seen like the image of a fresh hematoma still without visible vessels or capillarization. In intracutaneous hemangiomas, the typical double lamina structure of the skin has vanished.
- *Proliferation phase*: During the proliferation phase, IH usually proliferate at a fast rate while spreading in size, by exophytic, tangential or subcutaneous growth. Some IH develop only cutaneously (Fig. 1), but the majority has subcutaneous portions. The development of such a subcutaneous portion may continue for several months, leading to a much greater subcutaneous extension than is cutaneously visible ("iceberg phenomenon") (Fig. 2). Hypercapillarization can be seen in the CCDS image, and when further growth occurs, more and more vessels become distinguishable forming feeder arteries, a wide net of tumor vessels and draining veins with more (localized) or less (infiltrative) borders (Fig. 3).

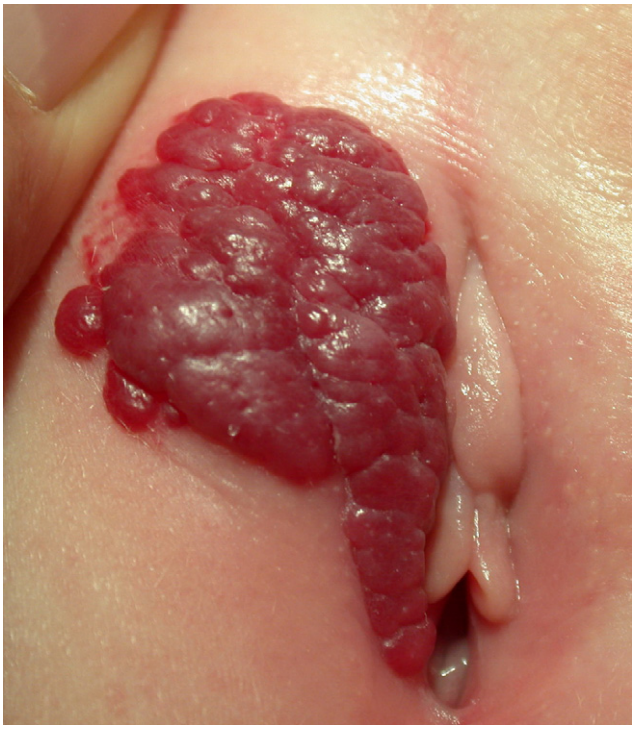

Fig. 1. Infantile hemangioma (labia maiora) with cutaneous exophytic manifestation in the proliferation phase.