Laser Treatment of Port-Wine Stains

M. J. Boffa
Dermatologist

Introduction

A state-of-the-art pulsed dye laser machine to treat port-wine stains and other vascular lesions has been available in the Malta Health Service since 1999. This article reviews the pathophysiology and clinical features of port-wine stains and describes the principles of laser treatment for this condition.

Pathophysiology

A port-wine stain *(syn. naevus flammeus)* is a vascular capillary malformation characterised by ectasia of superficial dermal capillaries[1]. Increased blood flow through these dilated vessels produces the persistent erythema which is seen clinically. The malformation represents a developmental anomaly and is not inherited; lesions occur familially no more frequently than by chance.In two large series the incidence of port-wine stains in the newborn was 0.5%[2] and 0.3%[3], with an equal sex distribution. Disturbances of neural supply and reduced vasoactive responses have been demonstrated in port-wine stain skin. Compared to normal skin there is a reduction in the density of perivascular nerves around the superficial dermal vascular plexus and it is likely that reduced influence on vascular tone contributes to the development of port-wine stains[4,5].

Clinical features

Port-wine stains are usually congenital i.e. present at birth, although their presence may initially be masked by the normal pink colour of neonatal skin. Rarely they may be acquired, appearing in childhood or adult life[6]. Clinically they present as well-demarcated areas varying in colour from light pink to deep red or purple (hence the term 'port-wines stain'[7]) and in size from a few millimetres to several centimetres across. Occasionally lesions can be very extensive and cover a large area, for example a whole limb. Port-wine stains may involve any part of the skin surface but occur most commonly on the face and upper trunk and are often unilateral. Port-wine stains usually persist throughout life with the surface area affected remaining unchanged relative to body size. With increasing age they tend to become darker purple in colour and more noticeable. Raised, thickened and nodular areas may also develop. There is no association with skin cancer.

Complications and associated conditions

The main consequence of port-wine stains is cosmetic. Lesions are often unsightly and, especially when on the face, may cause profound psychological disturbance[8]. Even small lesions at other sites may cause significant embarrassment and most patients who have port-wine stains would like to have them removed. Facial port-wine stains may be associated with abnormalities of the ocular vasculature including dilated conjunctival vessels and an increased risk of glaucoma. Glaucoma is more likely if both upper and lower eyelids are affected and such patients should have regular ophthalmic review. In the Sturge-Weber syndrome a unilateral facial port-wine stain is associated with ipsilateral encephalmeningeal angiomatosis which represents a developmental malformation of the vasculature of the leptomeninges. Affected individuals may suffer from epilepsy, mental retardation and glaucoma. The rare Klippel-Trenaunay syndrome consists of the association of a usually extensive port-wine stain on a limb with soft-tissue swelling, with or without bony overgrowth.

Treatment of port-wine stains

Until recently management of port-wine stains has been very unsatisfactory. Treatment modalities which have been used over the years include excision and grafting, dermabrasion, tattooing, Thorium X, red phosphorus, radiotherapy, electrocautery and cryotherapy[1]. Unfortunately none of these treatments produced acceptable results and for most patients the only option was cosmetic camouflage.

The situation has changed dramatically over past few years with the advent of pulsed dye lasers which are now the treatment of choice for port-wine stains. This is an exciting development which means that for the first time effective and safe treatment may be offered to affected patients. The term laser is an acronym for *l*ight *a*mplification by *s*timulated *e*mission of *r*adiation. Laser light has certain unique properties which are responsible for its clinical effects. Laser light is intense, coherent, collimated (perfectly parallel beam) and monochromatic. Sophisticated technology allows the laser light to be emitted in brief pulses.

Treatment is based on the principle of selective photothermolysis whereby pulses of light are selectively absorbed by the target chromphore (in this case haemoglobin in the abnormal blood vessels of the port-wine stain) to produce selective, thermally-medi-

ated injury[9]. By using short pulses, energy is deposited in the targets before they can cool off, producing extreme localised heating. This results in destruction of the target with minimal damage to intervening structures. The laser light is delivered in small circular spots commonly up to 10 mm in diameters. The parameters of the pulsed dye laser currently available locally include a pulse duration of 1.5msec and four wavelenghth options - 585nm, 590nm,595nm and 600nm. The longer wavelengths penetrate further into skin thus reaching vessels located more deeply. On the other hand because the longer wavelengths are further away from the absorption peak of haemoglobin (577nm) a bit of selectivity may be lost and there may be more competitive absorption by melanin. It is important to note that there are several different types of laser with medical applications yet only a few are suitable for treating port-wine stains. Although the parameters mentioned appear to be the best available at present for treating port-wine stains no single laser is completely effective for all cases. The pulsed dye laser is a major advance over the argon laser which was used in the past to treat port-wine stains but which produced unacceptable scarring in many cases.

Pulsed dye laser treatment for port-wine stains is usually carried out as an out-patient procedure. Laser pulses are applied carefully with minimal overlap to cover all the affected area. With big lesions this may be quite time consuming. Each pulse produces some discomfort if large areas are treated. In selected cases topical anaesthesia e.g. EMLA® cream may be helpful. Infants and young children require general anaesthesia[10].

Hazards, precautions and complications

In general, the pulsed dye laser has a low incidence of reactions and can be used even in infants and children[11,12]. Nevertheless it is a powerful energy source which has to be handled carefully. The laser should be used only in properly controlled sites with precautions taken to protect operators and patients from accidental exposure. In particular, laser light can damage the eyes if these are unprotected during treatment. Special protective eyewear must therefore be worn by both operators and patients during treatment to prevent the laser light from entering the eyes.

Purpura commonly appears in the treated area immediately after treatment. This may appear alarming but is harmless. It is caused by haemorrhage from the targeted blood vessels in the port-wine stain and typically resolves within 7-10 days. Blistering and crusting may also appear in the treated area especially if high fluences are used.

Potential cutaneous complications of pulsed dye laser treatment for port-wine stains include scarring and pigmentary changes (hypo- and hyper-pigmentation). There is a risk with every treatment and it is important to note that lack of scarring or pigmentary changes with previous treatment does not completely exclude this risk in future. Nevertheless the incidence of significant texture or pigment change following pulsed dye laser treatment is less than 5% in most published series[10,11] and the local experience appears to be in keeping with this or better. The laser machine available locally has a recently developed feature called dynamic cooling whereby a spurt of cryogen is sprayed on the skin just before the laser pulse. This reduces discomfort while protecting the epidermis and superficial dermis by selective cooling allowing safe use of higher laser fluences. There is evidence that laser treatment with dynamic cooling is safer and more effective than with earlier lasers which did not incorporate this feature[13]. The risk of pigmentary changes is greater in patients with darker skin types and those with sun-tanned skin. Treatment should therefore be avoided in those with tanned skin and patients instructed to protect the skin from sunlight before and after laser treatment. If blistering or crusting develop in the laser-treated area this may increase the risk of scarring and should be treated with a topical antibiotic ointment to reduce the chance of infection.

At what age should patients be treated?

Port-wine stains may be treated with the pulsed dye laser at any age. Ideally however treatment should start in infancy or early childhood with a view to completing it before school age and thus minimising psychological consequences of disfigurement. This is especially important with large, visible lesions on the head and neck. Early treatment has other advantages in that lesions usually respond better and require fewer laser treatments than those in older patients and furthermore fewer laser pulses are required because the area to treat is smaller. However advantages of early treatment have to be balanced against the risk and inconvenience of general anaesthesia which is required to treat young children[10].

Results

It is usual to treat a small test area first to assess response. Most lesions require multiple treatments, usually 6-10, for maximum improvement. Treatments are repeated every 6-8 weeks until maximum clearing becomes evident. In practice the first treatment produces proportionately most improvement. Response to treatment varies according to which particular laser equipment and parameters are utilised and therefore results in published studies are not always comparable. In general however more than 80%

of port-wine stains can be expected to lighten by at least 50%[14]. Not all lesions respond equally well[14]. Several studies have demostrated a better treatment outcome in younger patients and those with smaller lesions. Body site is also important - for example port-wine stains on the midface involving the medial cheek, upper lip and nose (corresponding roughly to the distribution of the second branch of the trigemminal nerve) usually respond more slowly and less completely than other facial areas. In addition there may be structural differences in microvascular patterns in different port-wine stains which could also contribute to differences in treatment outcome[17]. The presence of associated glaucoma, Sturge-Weber or Klippel Trenauney abnormalities are not contraindications to treatment however it should be emphasised that the laser will correct only the cutaneous component.

Pulsed dye laser treatment for port-wine stains has now been available in the Malta Health Service since 1999. The laser machine is installed at St Luke's Hospital. Several patients have been treated and results so far are encouraging. Although by no means all port-wine stains will disappear completely it should be possible in most cases to at least lighten the colour and thus produce worthwhile cosmetic improvement. Patients should be referred to the Dermatology Department in the usual way. It is important to note that the pulsed dye laser is effective only for vascular lesions. Pigmented birthmarks and other pigmented disorders do not respond to treatment with this laser.

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