Treatment of Haemorrhagic Telangiectasia with the Flashlamp-Pulsed Dye Laser

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Abstract The management of epistaxis in patients with hereditary haemorrhagic telangiectasia (Osler-Weber-Rendu) is often a perplexing problem. In the past, cautery, cryosurgery, and septodermoplasty have all been used with varying degrees of success. The Candela flashlamp-pulsed dye laser was developed for management of vascular lesions (port-wine stains and telangiectasia). The results of our prospective trial utilizing this laser in the treatment of intranasal telangiectasia in patients with hereditary haemorrhagic telangiectasia are presented.

Sommaire La conduite à tenir face aux épistaxis chez les patients avec télangiectasie hémorragique héréditaire (Osler-Rendu-Weber) est souvent un problème déroutant. Dans le passé, la cautérisation, la cryochirurgie et la septodermoplastie ont toutes été employées avec un degré variable de réussite. Le " Candela Flashlamp Pulsed Dye Laser" a été conçu pour la thérapie de lésions vasculaires (taches de vin et télangiectasies). Les résultats de notre recherche prospective utilisant ce laser dans le traitement des télangiectasies intranasales chez des patients avec télangiectasie hémorragique héréditaire seront présentés.

Hereditary haemorrhagic telangiectasia, or Osler-Weber-Rendu disease (OWR), is an autosomal dominant bleeding disorder with no sex or race predilection. The incidence of this disease has been reported to approach 2 per 100,000 population.[1] Problematic bleeding has been well known to occur in most parts of the body including the central nervous system, gastrointestinal tract, skin, and the nose. Epistaxis is usually the dominant clinical problem in OWR, and may give rise to numerous problems including minor (often daily) nosebleeds that may lead to a significant degree of psychosocial isolation, iron deficiency anemias, transfusions, and rarely, to life-threatening haemorrhages. The basic problem in this disease appears to lie at the level of the small subepithelial vessel.[2] Telangiectasias are the hallmark of OWR. These represent segmental lesions of thin-walled, fragile vascular channels, chiefly made up of capillaries and post-capillary venules, lined by a single layer of endothelium with no overlying protective coat of muscle or connective tissue.[3] The lack of elastic and muscular coats makes these telangiectatic lesions quite friable, rupturing with even minor trauma, and further unable to vasoconstrict effectively to control the haemorrhage when it does occur. Nasal telangiectasias are especially prone to haemorrhage due to local airflow and trauma effects.

A multitude of rather diverse treatments has been proposed for epistaxis control in OWR. Simple cauterization and nasal packing are the techniques used most often to control some of the acute manifestations of this disease but are unable to decrease, and in fact may exacerbate, future episodes of bleeding. Cryosurgery has had limited success. Surgical amelioration of the septal disease may be attempted with septodermoplasty. This consists of removing the most telangiectatic parts of the nasal and septal mucosa with subsequent skin grafting. In expert hands, this procedure has had some success[4,5] but, unfortunately, often cannot provide lasting relief due to further ingrowth of

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new telangiectatic vessels.[6] Exogenous estrogen therapy was believed to induce a squamous metaplasia of the nasal mucosa that, theoretically, may provide more protection than virgin mucosa for the underlying telangiectasias. Notwithstanding some of the potentially severe systemic effects of hormone therapy, a randomized trial using estrogen therapy in OWR did not show it to be of benefit.[7]

Laser photocoagulation has had some moderate success in the treatment of telangiectasias in OWR disease. Carbon dioxide, YAG, argon, and KTP lasers have all been utilized in the past.[8–11] The flashlamp-pulsed dye laser has been used widely in dermatology in the treatment of cutaneous vascular lesions because of its highly specific effect on blood vessels of the upper dermis. This laser can be adjusted to the maximum absorption spectrum of oxyhaemoglobin. It has been shown to have very high selectivity for small cutaneous vessels with minimal damage and no scarring of the surrounding tissue.[12] For these reasons, the flashlamp-pulsed dye laser (FPTD laser) is considered to be the laser of choice in the treatment of vascular lesions of the skin. It would also be expected to have a theoretic advantage over the other laser modalities in the treatment of epistaxis in OWR. One previous trial utilizing the FPTD laser was performed in Norway and found to be unsuccessful in ameliorating the frequency or severity of epistaxis in seven patients with OWR.[13] After analyzing their methodology, we felt that the energy levels used in that study were insufficient. Thus, with substantially greater energy levels, we undertook a prospective trial examining the effects, if any, of the FPTD laser in the treatment of telangiectasias in OWR.

Materials and Methods

A total of 11 patients with a diagnosis of Osler-Weber-Rendu disease were recruited into our study. Each had been diagnosed in the usual fashion based on the findings of typical telangiectasias, a positive family history, and recurrent epistaxis. A family tree was constructed that revealed that most of our study population was in fact somehow interrelated. Fully informed consent was obtained for all procedures performed.

All procedures were performed in the office setting of one of the authors (S.L.). Initially, each patient received a complete history and physical examination, with particular emphasis placed on the head and neck examination. Flexible fibre-optic endoscopy was used to first assess the extent of the disease in each case. Topical 5% cocaine solution was utilized to anaesthetize the nasal mucosa bilaterally. Protective eyewear was worn at all times by both patients and staff. A Candela flashlamp-pulsed dye laser with an experimental hand-held free fibre, provided by Candela Corporation, was used to photocoagulate all of the telangiectatic lesions under direct vision, with the use of anterior rhinoscopy. We were able to adjust the power delivered by adjusting the distance between the free end of the laser fibre and the mucosa (0.25–0.50 cm). Laser adjustments were performed by a qualified technician and were set at a wavelength of 585 nm, a pulse duration of 450 ms, a frequency of 1/5 sec, and an average spot size of 5 mm. A minimum energy level of 29 joules was utilized in each case. Following the procedure, patients were instructed to apply Polysporin ointment to each nasal vault. All patients were treated serially in 1 day and followed prospectively with a combination of telephone conversations, questionnaires, and 6-month follow-up examinations.

Results

The patient population consisted of 10 males and 1 female with a mean age of 41.8 years (range, 17–65 yr). Three of the 11 patients had had previous surgery performed in an attempt to control epistaxis (2 septodermoplasties and 1 rhinoseptoplasty). None had received previous transfusions. Almost all (n = 10) of the patients had undergone previous cauterization and/or nasal packing during acute episodes of epistaxis.

Flexible fibre-optic endoscopy was used immediately prior to the procedure in an attempt to qualify the severity of the nasal mucosal disease and to allow some broad localization of lesions. Most patients had disease localized primarily in the anterior septum and turbinates. The two patients with septodermoplasties had telangiectatic lesions scattered throughout their grafts. Topical cocainization did not affect the visibility of the telangiectasias on anterior rhinoscopy. The procedure was deemed universally painless by all patients. Three patients had very minor bleeding at the initial procedure that was adequately controlled with limited application of silver nitrate.

The mean number of discrete episodes of epistaxis experienced prior to the procedure was 6.8 per week (range, 1–25 per week). This number dropped substantially after laser photocoagulation to a mean of 2.7 per week (range, 0–12 per week) as noted at the 6-month follow-up.

Eighty-two percent (n = 9) of patients experienced a subjective improvement in their epistaxis (Table 1). Five of the 11 patients had a remarkable response, experiencing no further major nosebleeds after treatment. All but one patient felt that the procedure was of

Table 1 Subjective Change Post-Treatment (N = 11)

Significant improvement	7
Moderate improvement	2
No change	1
Worse	1

benefit to them. All patients, including the one that said he was worse after treatment, stated that they would like to have the procedure repeated on a regular basis. The one patient who experienced more episodes of epistaxis after laser photocoagulation was noted to have the most severe disease prior to the procedure. In addition, he had a substantial amount of posterior septal disease that was not treated due to the physical limitations of the equipment. In fact, the treated areas had improved substantially on the 6-month follow-up visit.

Physician assessment of the treated areas in the other subjects noted substantial improvement at the follow-up in every patient. There was a marked decrease in the numbers of telangiectatic lesions noted. The mean energy level used for photocoagulation was 30 joules (range, 29–32j). The mean number of pulses utilized was 89.3 (range, 58–153). All visible and accessible telangiectasias of the nasal lining on both sides of the septum were treated at the same sitting.

Discussion

With 82% (n = 9) of patients experiencing substantial relief from epistaxis after laser photocoagulation and all patients having significant improvement in the number of telangiectatic lesions in the treated areas, it may be reasonable to conclude that the flashlamp-pulsed dye laser is very effective in the treatment of epistaxis in OWR disease. The reduction in epistaxis would not be expected to be permanent, as new telangiectatic lesions will, undoubtedly, develop, as the underlying disease progression is not positively affected by any known treatment modality. The procedure would likely have to be repeated at regular intervals. The frequency of treatment would necessarily depend on the extent of disease and would have to be individually tailored. Most disease was noted to be localized to the anterior septum, which is easily accessible to treatment with a hand-held laser fibre under direct vision. Although the vast majority of patients experienced amelioration of symptoms, and 5 of 11 had no further major epistaxes, one cannot expect complete relief of epistaxis in all patients. Posterior disease was not accessible to treatment. This posterior disease would, naturally, be expected to contribute to nosebleeds.

The active component of all dye lasers is a complex organic dye that is dissolved in water or alcohol. The advantage of dye lasers is that the wavelength of the laser output can be selected or tuned according to the specific type of dye used. The wavelength of the Candela flashlamp-pulsed dye laser used in this trial is 585 nm. This corresponds quite well to one of the three known absorption peaks of oxyhaemoglobin (577 nm).[14] Theoretically, this should cause the flashlamp-pulsed dye laser to be more specific for the blood-filled telangiectatic lesions than for the surrounding nasal mucosa. This was clinically evident on the follow-up examination, with no evidence of atrophy or scarring of laser-treated nasal mucosa.

The major limitation of this procedure is the cost and availability of the flashlamp-pulsed dye laser. This is a rather expensive piece of equipment that likely would not be found in most otolaryngologists' offices. However, a number of dermatologists, with interests in treating vascular cutaneous lesions, have access to this equipment. Interspecialty cooperation is often needed in the optimum treatment of patients. Our present trial could not have taken place without this cooperation.

Conclusion

We believe it reasonable to conclude that the flashlamp-pulsed dye laser is a very effective tool in epistaxis control in Osler-Weber-Rendu disease. As a direct consequence of its unique specific absorption profile, the FPTD laser would be expected to provide better control than other laser modalities available (YAG, carbon dioxide, KTP, and argon). This latter point requires further study. If access to a FPTD laser is available, we believe that it should be considered early in the treatment of recurrent epistaxis in Osler-Weber-Rendu disease. It is a simple-to-perform and effective outpatient procedure that is of minimal discomfort to the patient and, generally, provides an excellent alternative in the treatment of troublesome epistaxis.

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