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Hereditary hemorrhagic telangiectasia treated by laser surgery

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SUMMARY

Hereditary hemorrhagic telangiectasia (Osler's disease) is a rare, inherited abnormality of the subepithelial vessels causing severe nose bleed as the most predominant symptom. The most efficient treatment has, until now, been dermoplasty of the nasal septum and cavity. There have been a few reports on treatment with CO_2-, argon- or Nd-YAG-lasers, which have been encouraging.

Ten patients with Osler's disease were treated for severe epistaxis. Three were treated with CO_2-laser, two of these under general anaesthesia. Seven patients were treated with the argon laser under local anaesthesia. A variety of other sites were treated at the same time for functional or cosmetic reasons (mouth, face, fingers, rectum).

Good results were obtained in eight cases. In three of the cases the observation period was more than two years. A second treatment has been necessary in three cases. Two other patients were treated recently, and it is too early to judge the results. The argon-laser is more convenient for this purpose than the CO_2-laser. The method is quick and reliable and there is no need for hospitalization. The treatment can be repeated without permanent damage to the function of the nose.

INTRODUCTION

Hereditary hemorrhagic telangiectasia (Osler's or Rendu-Osler-Weber's disease) is a rare, general abnormality of the small subepithelial vessels inherited in an autosomal, dominant pattern. Bleeding occurs from many of the surfaces of the body, but nose bleed is the most predominant symptom, often causing considerable morbidity and leading to repeated hospitalization and need for blood transfusions. There is a clear aggravation with increasing age, as new telangiectases develop.

Many modalities of treatment have been proposed in the past. Cauterization has proved to be of little value. Removal of the affected parts of the nasal mucosa on the septum and lateral walls, followed by free transplantation with a split skin graft as described by Saunders (1960), has been shown to control the bleeding efficiently in approximately 75% of the cases, also in long-term studies (Harrison, 1982; Ulso et al., 1983). Modifications using amniotic membrane as the trans-

planted material have been described (Laurian et al., 1979). No surgical procedures can, however, control the bleeding totally, and formation of new telangiectases in the transplant can occur (Harrison, 1982; Ulso et al., 1983).

There is a great deal of confusion concerning high dose estrogen treatment in this disease. This treatment has been shown efficient in 100% of cases in some series (Harrison, 1982), but it has also been reported by others to have no effect at all (Vase, 1981). No estrogen-binding and a few progesterone-binding receptors have been found in the nasal mucosa (Richtsmeier et al., 1984).

Laser treatment of hereditary haemorrhagic telangiectasia has been performed during the last decade, using CO_2- (Ben-Bassat et al., 1978), Neodymium-YAG- (Nd-YAG-) (Parkin and Dixon, 1981; Shapshay and Oliver, 1984) and argon-lasers (Parkin and Dixon, 1981). We have tried laser treatment in a series of patients with Osler's disease, as it seems to offer some advantages compared to the conventional surgical methods.

PATIENT MATERIAL

Ten patients with hereditary hemorrhagic telangiectasia have been referred to our clinic for nose bleed during the last three years, five of these from the same family (pt.no. 1-5, Table 1). Only one patient did not have a family history of Osler's disease. All patients had multiple telangiectases in the nose, most pronounced in the three elderly patients and pt.no. 9, all of whom had been hospitalized many times for haemostasis or blood transfusions. Pt.no. 9 had had a Saunders' operation performed five times previously, and she had been hospitalized for nose bleeding some 40 times, six times in the previous four month period. Estrogen treatment had been shown of no value.

Two of the patients had had an intracranial bleeding, and two had large pulmonary hemangiomas. One patient had had gastric bleeding and two suffered from rectal bleeding.

All patients had daily nose bleeds. All but one patient visited the emergency department or ENT-specialist unit at close intervals, and only three patients had never needed hospitalization.

Six of the patients were on constant treatment with iron, and they had had severe anaemia.

As the laser treatment under local anaesthesia was done in the out-patient clinic and was regarded as a minor procedure, the hemoglobin level was only examined in half of the cases prior to the treatment and was found to be in the low normal range.

METHOD

Three patients were treated with a Sharplan 733 CO_2-laser coupled to a Zeiss operating microscope with a micromanipulator. The eyes were protected with

Table 1. Sites treated, result of treatment, and observation time. All patients had daily nose bleeds before treatment.

pt.no.	sex	age	laser	regions treated	result	observation time, months
1.	M	80	argon	nose lips, fingers	minor bleeding twice since operation	24
2.	F	78	argon	nose	minor bleeding twice since operation	17
3.	F	71	CO_2 argon	nose fingers, lips	minor bleeding 5 times since operation	18
4.	M	47	argon ×2	nose	no change after treatment, new treatment recently	4
5.	F	40	argon	nose	minor bleeding a few times since operation	26
6.	F	40	CO_2	nose, left side	minor bleeding a few times since operation	25
7.	F	29	argon ×2	nose	recidiv after 3 weeks, rare bleeding since new argon-laser treatment	12
8.	M	42	argon ×2	nose fingers, rectum, lips, face, mouth	recidiv after 12 months, no bleeding since new argon-laser treatment	20
9.	F	40	CO_2	nose	no bleeding	4
10.	M	37	argon	nose, face	?	–

aluminium tape, and the face covered with wet gauze to prevent undesired burns. The telagiectases were treated individually using laser power of 10 watts and defocused beam, until vaporization of the abnormal vessels was obtained. Patient no. 3 was treated under general anaesthesia, as severe adhesions through the nose as a result of many previous attempts to control the bleeding by conven-

tional methods were found. These adhesions were vaporized. A piece of X-ray film was placed in the nasal cavity for three weeks postoperatively to restore a nasal airway and prevent reformation of adhesions. Patient no. 9 was also treated under general anaesthesia. Patient no. 6, who had rather few telangiectases, was treated under topical anaesthesia with cocaine 2%. Only one side of the nose was treated, as bleeding occurred only rarely from the other side.

Seven patients were treated with a Lexel Aurora 150 argon-laser. All patients were treated in the out-patient clinic. Topical anaesthesia with cocaine 2% was used. Eye-protecting glasses were used by patient and doctor, and the telangiectases were treated one by one using a nasal speculum. The beam was transmitted through a naked 300 μ quartz-fiber. Powers of 1 to 2 watts in continuous mode were used, until blanching of the abnormal vessel was seen.

Bleeding or cosmetically troublesome telangiectases in a variety of other sites were additionally treated in the same way in five of the patients. One patient was treated in the rectum and anal region, four patients on the lips and in the face, one in the mouth, and two on the hands and fingers. Local infiltration anaesthesia with lidocaine 1% was used for these treatments.

RESULTS

The results of the treatment are summarized in Table 1. Nose bleeding has stopped or been held at a very low level in eight of the patients, although repeated treatment was necessary in two of these patients.

Treatment with the CO_2-laser seemed peroperatively to be less satisfactory than with the argon-laser, as bleeding occurred from all treated spots. The bleeding, however, could easily be controlled, and the three patients treated with the CO_2-laser are completely satisfied with the results, two of them having had minor bleeding only a few times after 18 and 25 months respectively, while patient no. 9 has had no bleeding at all for four months.

Treatment with the argon-laser was a minor procedure, well tolerated by the patients, and no per- or postoperative bleeding occurred. The effect has so far been very good in three patients, who have had only minor epistaxis for 26, 24, and 17 months respectively. Repeated bleeding occurred in patients no. 7 and 8 after three weeks and 12 months respectively, but a new treatment with argon-laser has had good effect for almost a year. Patient no. 4 had no benefit from the treatment, and a new treatment has been given recently. Patient no. 10 has likewise been treated recently.

A better efficacy of the CO_2-laser than of the argon-laser might be suspected from the results, but the material is too sparse for statistic evaluation. Repeated hemoglobin estimations were not done at the follow-up examinations.

The argon-laser treatment of lesions in other sites was also satisfactory, as no bleeding occurred from the treated spots during the observation period. Many of

the telangiectases have disappeared completely, although a few elements recurred.

DISCUSSION

The results from treatment with CO_2- and argon-laser are quite satisfactory. The bleeding from the nose can usually be controlled and held at an acceptable level, and the daily life of the patients can be normalized.

Lesions in the posterior part of the nasal cavity may be expected to be more difficult to treat than lesions in the anterior half of the nasal cavity with both the CO_2- and argon-laser, as they can be difficult to visualize. However, most of the lesions in Osler's disease are situated in the anterior half and the nasal cavity, predominantly on the septum, and none of the patients, who needed repeated treatment had nose bleeds from the posterior part.

The work with the CO_2-laser is to some extent impeded by bleeding from the mucous membrane caused by the cutting effect of the laser, which on the other hand makes minor resections possible, thus permitting visualization of more inaccessible parts of the nasal cavity or restoration of a lumen in an obstructed air passage through the nose. The laser beam will hit the mucous membrane tangentially, especially when directed towards the posterior parts of the nasal cavity. This will lead to a defocused beam, which enhances the hemostatic effect and diminishes the cutting effect of the treatment. General anaesthesia is often required.

The rather limited depths of coagulation of the argon-laser makes this laser well suited for use in hereditary hemorrhagic telangiectasia, as the lesions are superficial. The beam is absorbed by hemoglobin, and the effect is therefore concentrated in the telangiectases. The treatment is well tolerated on an out-clinic basis. The laser treatment can be repeated without permanent damage to the function of the nose.

ZUSAMMENFASSUNG

Erbliche hämorrhagishe Teleangiektasie (Oslers Krankheit) ist eine seltene erbliche Abnormität der subepithelischen Kapillaren, die als vorherrschendes Symptom schweres Nasenbluten verursacht. Die bisher effektivste Behandlung war Dermoplastik von Septum und Nasenhöhle. Es hat einige versprechende Berichte von Behandlungen mit CO_2-, Argon- oder Nd-Yag-Laser gegeben.

Zehn Patienten mit Oslers Krankheit wurden gegen schwere Epistaxis behandelt. Drei wurden mit CO_2-Laser behandelt, zwei von ihnen in Vollnarkose. Sieben Patienten wurden mit dem Argon-Laser in örtlicher Betäubung behandelt. Verschiedene Regionen wurden gleichzeitig aus funktionalen oder kosmetischen Gründen behandelt (Mund, Gesicht, Finger, Rektum).

In Acht Fällen wurden gute Resultate erzielt. In drei Fällen dauerte die Beobachtungszeit mehr als zwei Jahren.

Eine wiederholte Behandlung war in drei Fällen notwendig. Zwei andere Patienten wurden neulich behandelt, und das Ergebnis lässt sich so früh nicht beurteilen.

Der Argon-Laser ist für solche Zwecke besser geeignet als der CO_2-Laser. Die Methode ist schnell und zuverlässig und Krankenhausaufenthalt ist unerforderlich. Die Behandlung kann ohne bleibenden Schaden an der Nasenfunktion wiederholt werden.

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