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Hereditary haemorrhagic telangiectasia: Young's procedure in the management of epistaxis

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Abstract

Two cases of hereditary haemorrhagic telangiectasia are presented. Epistaxis was intractable in both patients. Young's procedure proved successful in controlling nose bleeds. One of the patients has been followed-up for 14 years. Methods of treatment are discussed with a review of the literature.

Key words: Epistaxis; Telangiectasia, hereditary haemorrhagic; Surgery

Introduction

Hereditary haemorrhagic telangiectasia is an autosomal dominant familial disease manifested by the presence of vascular malformations throughout the body, commonly affecting the skin and mucous membranes. These malformations can bleed either spontaneously or following trivial trauma.

Epistaxis, the most common symptom, can be very difficult to manage. However, there is no generally accepted method of treatment. Closure of the nostrils (Young's procedure) is a well-established method of treatment for chronic atrophic rhinitis (Young, 1967; Shah *et al.*, 1974; Sinha *et al.*, 1977). Brooker and Cinnamond (1991) reported two cases of epistaxis due to nose picking: Young's procedure was successful in one case, the idea being to create a barrier of skin against the offending fingers. Reviewing the English literature, we could not find any other link between Young's procedure and epistaxis. The purpose of this paper is to advocate the use of Young's procedure for the treatment of intractable epistaxis in some patients with hereditary haemorrhagic telangiectasia.

Case reports

Case 1

A 53-year-old man presented to the Norfolk and Norwich Hospital, in January 1972, with recurrent epistaxis. In 1943 the patient had been diagnosed as suffering from hereditary haemorrhagic telangiectasia. Nose bleeds were the only manifestation of the disease. In the past he had been treated with repeated nasal packing and electrocautery. He had required multiple blood transfusions and was on regular iron tables to correct his anaemia. At one stage, he had been prescribed systemic oestrogen but this was given for only a short period due to intolerance of its side effects.

The patient's father, father's sister, grandfather and a brother were all affected but the patient was the worst affected in terms of epistaxes.

On examination, telangiectatic spots were present on the face and both the hard and soft palate. Anterior rhinoscopy revealed 18 telangiectatic lesions in the left nasal airway scattered over the septum and the lateral wall. A number of lesions were present on the right side and a small nasal septal perforation was present. There was no hepatomegaly or splenomegaly. Investigations revealed haemoglobin of 8.8 g/dl with a normal blood picture and clotting screen.

Initially the patient was treated with cryosurgery and chlortrianisene (a systemic pro-oestrogen which is not a steroid but which is stored in the body fat and slowly converted in the liver to oestrogen). Cryosurgery helped to reduce the amount of bleeding for only a short period, necessitating repeated applications at four to six weekly intervals. In May 1972 closure of the septal perforation was attempted using a local mucoperichondrial flap. In October 1973 the patient received an eight-day course of radiotherapy to his upper air passages but his nose bleeds continued.

In 1974 he suffered a posterior-inferior cerebellar artery thrombosis following which the oestrogen therapy was discontinued. The patient's haemoglobin level continued to fall and he was transfused when the latter dropped below 12 g/dl.

In July 1979, after the patient had received a total of 62 cryosurgical applications, it was decided to perform Young's procedure on both sides of the nose. A few weeks following surgery the closure on the left side became dehiscent at the suture line leaving a 2 mm opening. The right nostril remained completely closed. This situation has remained stable to the present day and the patient has had no further nose bleeds. It is worth reporting that when the patient sneezes he exudes through the hole clear mucus free of blood.

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After the operation it took the patient a few months to become accustomed to mouth breathing and to accept the consequent dryness of his throat. In the last 14 years he has been followed-up regularly on an annual basis and during this time he has not suffered any further significant nose bleeds, has not required blood transfusions, and is no longer taking oral iron. His haemoglobin has remained at a normal level.

Case 2

A 63-year-old, retired printer, with a known history of hereditary haemorrhagic telangiectasia presented to the Norfolk and Norwich Hospital, in November 1991. He had suffered from regular nose bleeds for many years but there was no history of bleeding from any other site. The patient's father, sister and a niece were also affected by the disease and also had nose bleeds. In 1978 the patient had undergone septal dermoplasty of the left nasal cavity but unfortunately the operation proved unsuccessful. The patient was on regular iron tablets to correct his anaemia.

On examination, telangiectatic lesions were visible on the face, ears, nose, lips and on the roof of the mouth. Figure 1 shows telangiectases over the tongue and lower lip.

Anterior rhinoscopy showed multiple discrete telangiectases on the right side of the nasal septum and much bigger vascular lesions of the left side of the septum. Investigations revealed a haemoglobin of 11.8 g/dl. Platelet count and clotting screen were normal. No abnormalities were seen on his chest X-ray.

It was decided to treat him with the CO_2-laser and this procedure was carried out on three separate occasions, with one episode of treatment using electrocautery to the nasal septal lesions. However, the nose bleeds persisted and the patient's haemoglobin dropped to 9.5 g/dl. Due to the failure of his treatment to date, coupled with the knowledge of the success of Young's procedure in the patient described in *Case 1*, it was decided to close the patient's nostrils surgically. This was performed under a general anaesthetic in October 1992. The post-operative period was uneventful and follow-up visits confirmed the success of the operation. However, the patient was unhappy with the bilateral closure, finding it difficult to cope with the complete nasal obstruction and requested that one side be reopened. Partial reopening was carried out in January 1993 and a year later when the patient was reviewed in the clinic he was delighted with the results of surgery. The opening in the right nasal airway, which is 3 to 4 mm in diameter, allowed us to inspect the mucosa of the right nasal cavity using the nasendoscope. The mucosa appeared much healthier and although telangiectatic spots were still present there were no crusts and no evidence of recent bleeding. The patient's haemoglobin had risen to 13.4 g/dl and remains at this level.

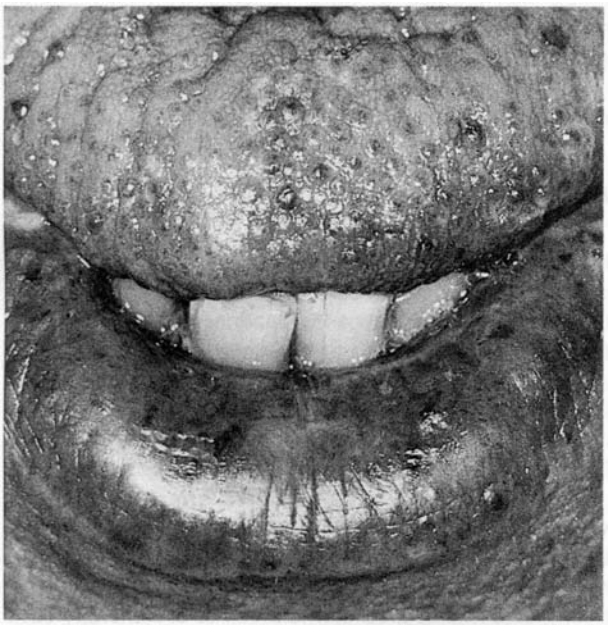

FIG. 1
Telangiectases on the tongue and lower lip.

Review of the literature

History

The first mention of this condition was by Sutton (1864) who described the association between epistaxis, internal bleeding and telangiectasias of the skin. The hereditary nature of the condition became an integral part following the publication of a letter by Babington (1865) to the editor of the *Lancet* in which he described five consecutive generations affected by epistaxis although there was no mention of telangiectasia. Further clarification of the disease was made by Rendu (1896), Osler (1901) and Weber (1907). The condition became known as Osler–Rendu–Weber disease. Hanes (1909) suggested the descriptive terminology of familial haemorrhagic telangiectasia.

Genetics

Hereditary haemorrhagic telangiectasia is an autosomal dominant disease. The patients affected are of the heterozygous type. The homozygous form of the disease is incompatible with life. Telangiectasia tends to be present in one generation and may skip certain generations before it reappears; such a tendency is called atavism. Atavism may account for the lack of family history of the condition in some patients. Gene mutation is another explanation of the lack of family history in other patients.

Pathology

In hereditary haemorrhagic telangiectasia there are two basic vascular abnormalities: firstly, there are localized areas in the capillaries that lack elastic tissue, and only have an endothelial layer; secondly, there are dilatations of arterioles and capillaries leading to arterio-venous fistulae. Electron microscopic studies have illustrated the presence of smooth muscle cells in the walls of the vascular cases and confirmed the absence of elastic tissue (Jahnke, 1970).

These smooth muscles were however elongated, irregularly-shaped, and did not form a continuous coat around the vessel. They were incapable of exhibiting adequate contraction. Bleeding always occurs from the summit of the telangiectases and never extravasates into the surrounding tissues. It thus appears likely that the factors which determine whether the lesions bleed or not are to be found in the state of the overlying epithelium and not in the vessels themselves (Harrison, 1964).

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Clinical picture

Telangiectases may occur on any of the mucous membranes or anywhere on the external surface of the body. The nasal mucous membrane is the most common site of involvement followed by the following sites in order of decreasing frequency: tongue, palate, lips, buccal areas, gum, pharynx and conjunctiva. On the skin, the lesions are found on the face, trunk, arms, fingertips and nail beds, but bleeding rarely occurs from these sites, probably because of the protective layer of squamous epithelium which usually covers the telangiectasis and minimizes damage from trauma (Harrison, 1964). The patient's blood is normal (except for anaemia) and the only defect is in the blood vessel. Often symptoms do not start until adulthood, when the number of telangiectases increases and these usually cause bleeding and anaemia. Epistaxis is the most common symptom followed by gastrointestinal bleeding. This is not surprising as the telangiectactic lesions in these two sites are covered by delicate mucous membrane and are subject to trauma. Telangiectases in the gastrointestinal tract can cause haematemesis and/or melaena. Pulmonary manifestations are not uncommon in patients with hereditary haemorrhagic telangiectasia. Arteriovenous fistulae are the basic pathological lesions.

Treatment

In 1969 Bradbeer listed the following methods of treatment for epistaxis in hereditary haemorrhagic telangiectasia:

(1) Electrocautery and application of various caustics
(2) Partial resection of the nasal septum
(3) Various forms of irradiation
(4) Hormone therapy with oestrogen
(5) Amputation of the nose
(6) Septal dermoplasty

To this list the following should be added:

(7) Arterial ligation
(8) Microembolization
(9) Cryosurgery
(10) Laser surgery
(11) Cultured epithelial sheets

Hormonal management of hereditary haemorrhagic telangiectasia was first proposed by Koch *et al.*, 1952. Animal experiments indicated that under the influence of systemic oestrogen, the ciliated columnar epithelium of the nose underwent metaplasia to squamous epithelium, and this was maintained as long as oestrogen was given. This squamous epithelium will provide a protective layer over the nasal lesions (Harrison, 1964). Oestrogen however has undesirable side effects. In view of the potential dangers of prolonged administration of large doses of oestrogen, it is probably wise to restrict such treatment to women after the menopause and to men after the male climacteric (Blackburn, 1963). The use of oestrogen–progesterone combination is effective in controlling bleeding. This beneficial effect lasts for many months after stopping hormonal therapy (Van Cutsem *et al.*, 1988; Van Cutsem, 1993).

Septal dermoplasty was described by Saunders (1962). Epistaxis in hereditary haemorrhagic telangiectasia occurs from the anterior part of the nose, the part subject to trauma, so providing thick skin cover over the telangiectactic lesions which should offer protection. Old mucosa but not perichondrium is removed from the anterior half of both sides of the nasal septum, from the top of the nose down to, and including, the floor. A split thickness skin graft is applied to the new area. This technique has proved to be successful in large numbers of patients with cessation of nose bleeding and return of the haemoglobin to normal values. Recurrence of telangiectasias within the skin graft has however been reported (McCabe and Kelly, 1972).

Different types of laser were used for the eradication of telangiectatic lesions in the nasal cavity. Neodymium–Yttrium-aluminium-garnet (Nd–YAG) laser photocoagulation is the most effective. In a study involving 19 patients treated with Nd–YAG laser, no complications occurred over a four-year period. However, three patients with the most severe epistaxis received minimal or no benefit (Kluger *et al.*, 1987). In another study (Illum and Bjerring, 1988) 10 patients with Osler's disease were treated for severe epistaxis. Three patients were treated with CO_2-laser and seven patients with argon laser. They found the argon laser to be more effective. This is not surprising as CO_2-laser is a cutting laser and argon laser is a coagulating laser.

Microembolization has been described, in which a small radioopague catheter is introduced via the femoral route and advanced into the carotid system. The maxillary artery is injected to obtain a series of films of the distribution of the vessel. First, tiny fragments of Gelfoam, about 1 × 2 mm, are used to penetrate the telangiectasia, and then progressively larger fragments are used to occlude the supplying arteries. This technique appears to have been successful for a time at least in one very severe case (Kendall *et al.*, 1977).

The use of cultured epithelial sheets is a new technique in which autografts of sheets of epithelium derived by culture from buccal epithelium were used to graft the nasal septum and inferior turbinates after surgical removal of the respiratory epithelium (Milton *et al.*, 1993). This technique was applied to three patients with encouraging results, although two of these required insertion of two grafts.

Exotic measures such as snake venom, vitamin K and intramuscular milk injections remain only as curiosities.

Discussion

Many different forms of medical and surgical therapy have been tried in futile efforts to control the severe, often daily epistaxis of patients with hereditary haemorrhagic telangiectasia. In some cases a patient may receive transfusions of huge amounts of blood during his/her lifetime. As one of Osler's patients put it 'I am in the habit of bleeding to death every day'. Others who bleed in lesser amounts still may be handicapped because they bleed so frequently that their social and working lives are affected.

In the two cases presented here, different treatments were tried unsuccessfully and the patients also suffered treatment complications. In *Case 1*, the patient had repeated electrocautery (which resulted in septal perforation), radiotherapy, 62 applications of cryotherapy each requiring hospital admission, systemic oestrogen which may have contributed to a posterior-inferior cerebellar

artery thrombosis and repeated blood transfusions. He had been badly affected by nose bleeds for 36 years. Following closure of his nostrils (one only partially), he has not been troubled with nose bleeds for over 14 years. In *Case 2*, repeated electrocautery, septal dermoplasty and also laser surgery were all ineffective in controlling nose bleeds and he also benefited from closure of his nostril.

Bleeding occurs chiefly from those epithelial surfaces where trauma is likely and where the mucosa is not of the squamous type (Saunders, 1964). Injury to the delicate telangiectatic areas in the nasal cavity may occur by drying, picking, sneezing or the mere changes in the intranasal pressure during breathing. Providing a tougher surface able to sustain trauma is the basic principle behind oestrogen therapy, dermal septoplasty and cultured epithelial sheets. Elimination of trauma is the aim of nasal closure.

Closure of the nostrils whether partial or complete is a well-established method of treatment for chronic atrophic rhinitis (Young, 1967; Shah *et al.*, 1974; Sinha *et al.*, 1977). In *Case 1*, one-layer closure was performed as advocated by Young (1967). In *Case 2*, we performed a two-layer closure technique. In this technique, the incision was made circumferentially in the vestibule 5 mm anterior to the muco-cutaneous junction. A skin flap posterior to the incision was elevated all round and sutured with absorbable material to form a deep layer. A further flap was raised by dissecting the skin anteriorly and sutured to form a superficial layer (Sinha *et al.*, 1977).

Although partial closure of one nostril was achieved in *Case 1* due to wound dehiscence and deliberately in one nostril in *Case 2* following the patient's request to reopen one side, partial closure proved as effective as complete closure in controlling nose bleeds in these two cases.

Although Young's procedure is not necessarily the final answer to epistaxis in hereditary haemaorrhagic telangiectasia, compared to other methods of treatment, it is an option worth considering.

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