

A novel treatment for patients with hereditary haemorrhagic telangiectasia

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Abstract

Hereditary haemorrhagic telangiectasia (HHT) is an autosomal dominant disorder characterized by dermal, mucosal, and visceral telangiectases as well as pulmonary and cerebral arteriovenous malformations. Recurrent epistaxis occurs in the majority of patients, and by the very nature of the thin walled vessels involved it is often refractory to conventional forms of treatment. We present the case of an 82-year-old lady with intractable epistaxis secondary to HHT, that was successfully controlled by the application of fibrin glue.

Key words: Telangiectasia, Hereditary Haemorrhagic; Fibrin Tissue Adhesive

Introduction

HHT results from the inheritance of a single autosomal dominant gene producing localized areas of thin-walled capillaries. In mature telangiectases, the venules are markedly dilated and convoluted, have excessive layers of smooth muscle without elastic fibres, and often connect directly to dilated arterioles.¹ It is thought that the lack of elastic tissue leaves the damaged vessel incapable of effective constriction and direct connection to arterioles makes the control of venous haemorrhage difficult. Given that the pseudostratified respiratory epithelium of the nasal cavity is susceptible to drying crusting and the local trauma of nose blowing and digitation, it is no surprise that epistaxis is the commonest form of presentation of this disease.¹

Case report

This 84-year-old lady has been attending the local ENT department with recurrent epistaxis for the past 20 years. During this time she has been treated with repeated nasal cautery, anterior and posterior nasal packing, ligation of both internal maxillary arteries and both facial arteries, septodermoplasty, insertion of a septal button to a perforation and laser photocoagulation.

Her condition had deteriorated in the 133-day period before the application of fibrin glue. During this time she spent 113 days in hospital for management of her epistaxis. Her longest continuous period out of hospital was 10 days and the average length of stay was 22.6 days. She was treated with further nasal packing using both bismuth iodoform paraffin paste on ribbon gauze and Algosteril® rope packs. She had open ligation of the anterior ethmoidal artery and on two occasions was referred to the department of neuroradiology at the Royal Victoria Hospital in Belfast. On the first occasion angiography demonstrated abnormal and tortuous beaded vessels bilaterally in the nasal cavity and tongue, typical of telangiectases. The internal maxillary artery was selec-

tively catheterized and the vessels supplying the telangiectatic lesions were occluded with contour particles. The internal maxillary artery was occluded with VortX coils. A complete angiographic obliteration of the telangiectatic lesions of the right distal IMAX was achieved. She was discharged four days after this procedure, but returned three days later with further right-sided bleeding. Cerebral angiography was then attempted, but unfortunately was unsuccessful.

After further epistaxis the patient was taken to theatre for examination under anaesthetic and application of fibrin tissue sealant. The septal button was removed and bleeding was noted from the edge of the perforation, the floor of the nose and the anterior end of the inferior turbinate. The bleeding stopped immediately with the application of fibrin glue and no nasal packing was required. The nasal cavity remained dry post-operatively and the patient was discharged four days later. At four-week review there has been only one episode of mild epistaxis, that did not require hospital admission. Examination of the nose, however, caused further bleeding which was controlled with another application of fibrin glue in theatre. This resulted in a three-day stay in hospital. It is now our intention to apply the sealant in the outpatient setting on a regular basis to avoid any further hospital admission.

Materials

The fibrin sealant used in this case was from the Tisseel® kit. The kit contains two components, which are kept in separate syringes prior to administration. One syringe contains the steam-treated sealer protein concentrate including bovine aprotinin solution, the other contains lyophilized thrombin and calcium chloride solution. The bovine herd used for extraction of aprotinin has been bred selectively for this purpose to preclude the risk of transmission of known prion-related diseases. The human plasma products used in the kit are taken from healthy

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donors who have tested negative for antibodies to human immunodeficiency virus (HIV) 1 and 2 and hepatitis C virus (HCV) as well as hepatitis B virus surface antigen (HBsAg). The ALT level must not exceed a certain threshold and a sample of the plasma pool is also tested for HIV and HCV antibodies, along with HBsAg. In addition a test for virus genome sequences of HIV, HBV and HCV is carried out by the polymerase chain reaction.

The two components are mixed during application. This results in a viscous thrombin solution that quickly sets to form a white, elastic mass, which firmly adheres to the tissue. This process simulates key features of the physiological coagulation process and is used to achieve haemostasis, to seal or glue tissue and to support wound healing. In the course of wound healing the solidified fibrin sealant is completely absorbed.

Discussion

This case highlights some of the difficulties associated with the management of refractory epistaxis in HHT and demonstrates a new method for the control of such bleeding. A thorough search of the English medical literature has revealed no other case where fibrin glue has been topically applied in a patient with HHT.

Treatment of such patients includes simple measures such as packing and cautery. Cautery in HHT can exacerbate bleeding and therefore is best performed by an experienced otolaryngologist.¹ For more difficult cases embolization of the vascular tree, or laser photocoagulation may be useful, but these techniques are not available in all centres. Surgical treatments include endoscopic and open arterial ligation, septodermoplasty, Young's procedure² and even the auto-grafting of cultured epithelial sheets derived from the buccal mucosa.³

Surgical intervention can carry significant morbidity by disrupting the normal anatomy and physiology of the nose and should only be used as a last resort. Control of

epistaxis is best achieved at the point of bleeding. More proximal intervention paves the way for collateral vessel formation and anastomotic supply of the bleeding point.

This technique of fibrin sealant application in the management of intractable epistaxis in HHT avoids the trauma of nasal packing, preserves normal nasal function and provides good distal control of the bleeding vessel. We recommend that the use of fibrin sealant should be considered before surgical intervention in the management of refractory epistaxis in patients with HHT.

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