The Journal of Laryngology & Otology (2009), ***123***, *695–696.*

doi:10.1017/S0022215109004964

Clinical Record

A natural obturator in hereditary haemorrhagic telangiectasia

A Soni-Jaiswal, T J Woolford

Abstract

Objective: Most patients with hereditary haemorrhagic telangiectasia suffer with frequent episodes of epistaxis. The aim of this case report is to highlight the effect on epistaxis, occurring in hereditary haemorrhagic telangiectasia, when nasal airflow ceases.

Case report: We present the interesting case of a patient with hereditary haemorrhagic telangiectasia who experienced cessation of her recurrent, refractory epistaxis through the development of coexisting polyp disease. The patient's enlarged, grade three nasal polyps were behaving as physiological obturators, limiting airflow through her nose. This reduced the intranasal trauma and subsequent frequency of her nosebleeds.

Conclusion: Epistaxis is a debilitating part of hereditary haemorrhagic telangiectasia, and poses a frequent management challenge. Our patient was more tolerant of her grade three nasal polyps than of her recurrent epistaxis. This case highlights the importance of reducing nasal airflow when treating patients with hereditary haemorrhagic telangiectasia.

Key words: Hereditary Haemorrhagic Telangiectasia; Epistaxis; Nasal Polyps

Introduction

Hereditary haemorrhagic telangiectasia, or Osler–Weber–Rendu syndrome, first appeared in the literature in 1864, when Sutton described it as a disorder of epistaxis and degeneration of the vascular system.[1] It is a complex, systemic disorder characterised by telangiectases, arteriovenous malformations and aneurysms.[2] Hereditary haemorrhagic telangiectasia is a familial disorder with autosomal dominant, non-sex linked inheritance. Its incidence is about one in 8000 and it has equal sex preponderance.[3,4] Mutations on the long arm of chromosomes 9 and 12 are implicated. The resultant protein deficiencies produce an abnormal vascular architecture at the affected sites, prone to damage with minimal trauma.[4,5]

The telangiectases occur at multiple mucocutaneous sites. It is their presence within the nasal mucosa which brings them to the attention of the ENT surgeon. Epistaxis is the most frequent manifestation of hereditary haemorrhagic telangiectasia. The symptom tends to start in the third decade and in the majority of patients will progressively worsen over the ensuing years.[6,7]

Patients with moderate to severe nasal disease present a difficult management challenge. Various treatments are described in the literature, including those aimed at reducing nasal airflow. It is thought that eliminating airflow through the nose protects the mucosa from traumatic injury, thus decreasing the frequency of epistaxis.[8,9]

We present a case in which reduction in nasal airflow was produced by the patient's coexisting nasal polyp disease. The intranasal polyps mimicked a Silastic® obturator,[10] producing a dramatic reduction in bleeding.

Case report

A 56-year-old woman with type I familial hereditary haemorrhagic telangiectasia had suffered with severe, recurrent epistaxis for over 19 years. Incidentally, she had also suffered with nasal polyp disease. The epistaxis had progressively worsened over the years. The patient had tried a multitude of treatments, all with short-lived benefit, including electro-cautery under general anaesthesia, topical and systemic oestrogen therapy, and oral antifibrinolytics.

The patient had been fitted with a Silastic obturator in 1998, which she had successfully used until 2004, when her asthma worsened and she found wearing the obturator uncomfortable. Unfortunately, her daily epistaxis had resumed.

During an out-patients clinic appointment, the patient revealed that, for the last six months, her epistaxis had all but ceased. However, she did complain of nasal obstruction and anosmia.

Endoscopic nasal examination revealed bilateral nasal polyposis completely obstructing both nasal airways. The nasal polyps had enlarged over the year and were now acting as a physiological obturator, reducing the airflow through the nose.

The situation was discussed with the patient and a mutual decision made to leave the polyps untreated.

Discussion

Young's procedure and Silastic nasal obturators are treatment modalities which aim to reduce nasal airflow. Young's procedure, first described by Taylor and Young in 1961, involves complete surgical closure of one or both nasal cavities.[11] Bilateral surgical closure of the nasal cavities is the only known treatment modality to produce complete cessation of bleeding in cases of hereditary haemorrhagic telangiectasia. Bilateral closure eliminates airflow through the nose, protecting the mucosa from

From the Department of Otolaryngology, Manchester Royal Infirmary, UK.
Accepted for publication: 20 November 2008. First published online 11 March 2009.

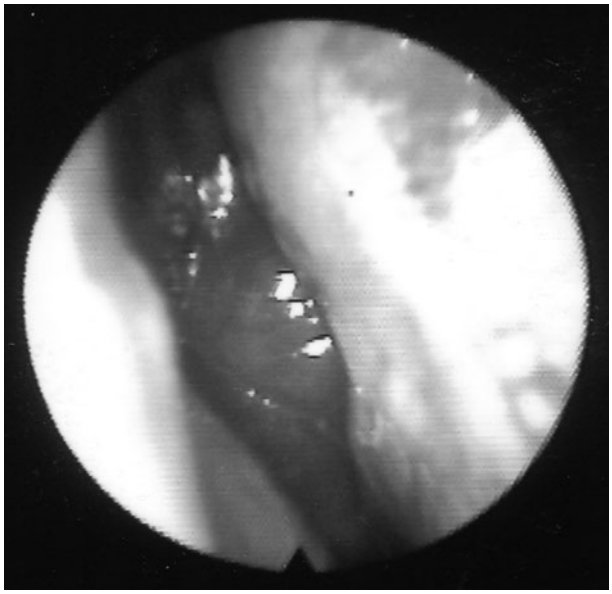

FIG. 1
Endoscopic view of the patient's polyps, which acted as physiological obturators. Note also the multiple mucosal telangiectases.

traumatic injury.[8,9] A study by Lund and Howard found that, despite the psychological stress of the procedure, patients reported an improved quality of life afterwards.[8]

- **Most patients with hereditary haemorrhagic telangiectasia suffer with frequent episodes of epistaxis**
- **The authors present a patient with hereditary haemorrhagic telangiectasia who experienced cessation of her recurrent, refractory epistaxis through the development of coexisting polyp disease**
- **The patient's enlarged, grade three nasal polyps behaved as physiological obturators, limiting airflow through her nose. This reduced the intranasal trauma and subsequent frequency of her nosebleeds**

Silastic nasal obturators provide an alternative to complete surgical closure. They eliminate nasal airflow in the same manner but are a temporary measure which the patient may remove at will, giving the patient some control over their illness.[10] Although Silastic obturators do not offer complete cessation of bleeding, they are a non-invasive, non-surgical technique offering vast improvement in the patient's frequency of epistaxis.

Our patient's bilateral, grade three nasal polyps mimicked the action of Silastic obturators. They occluded both nostrils, reducing the traumatic airflow through the nose, and produced complete cessation of our patient's severe, recurrent epistaxis. Despite the resulting anosmia and nasal obstruction, our patient decided to leave her polyps untreated, as she was more tolerant of these symptoms than the recurrent epistaxis to which she had been subject for many years.

Conclusion

This case highlights the effect on epistaxis in hereditary haemorrhagic telangiectasia if nasal airflow ceases. Our patient's symptoms improved because the polyps within her nose were behaving as physiological obturators. This also avoided the need for any further surgical treatment or recurrent nasal packing, improving her quality of life.

References

1 Sutton HG. Epistaxis as an indicator of impaired nutrition and degeneration of the vascular system. *Medical Mirror* 1864;**1**:769–71
2 Guttmacher AE, Marchuk DA, White RI. Hereditary haemorrhagic telangiectasia. *N Engl J Med* 1995;**333**:918–24
3 Cotran RS, Vinay K, Collins T. *'Robbins' Pathologic Basis of Disease 5th Edition. Published by W.B. Saunders Company, A Division of Harcourt Brace & Company, Philadelphia* 1999;531–2
4 Soriano PA, Petros J, Mckinsey JF. Osler-Weber-Rendu Disease. *'emedicine from WebMD'* http://emedicine.mediscape.com/article, updated 15th December 2008
5 Marchuk DA. The molecular genetics of hereditary hemorrhagic telangiectasia. *Chest* June 1997;**111**:79S–82S
6 Saunders WH. Hereditary Hemorrhagic Telangiectasia: Its familial pattern, clinical characteristics, and surgical treatment. *Arch Otolaryngol Head Neck Surgery* 1962;**76**: 245–60
7 Ross D, Jassin B. Current trends in the diagnosis and management of Osler Weber Rendu disease. *Curr Opin Otolaryngol Head Neck Surg* 1997;**5**:191–6
8 Lund V, Howard DJ. Closure of the nasal cavities in the treatment of refractory hereditary haemorrhagic telangiectasia. *J Laryngol Otol* 1997;**111**:30–3
9 Gluckman JL, Portugal LG. Modified Young's procedure for refractory epistaxis due to hereditary haemorrhagic telangiectasia. *Laryngoscope* 1994;**104**:1174–7
10 Woolford TJ, Loke D, Bateman ND. The use of a nasal obturator in hereditary haemorrhagic telangiectasia: an alternative to Young's procedure. *J Laryngol Otol* 2002; **116**:455–6
11 Young A. Closure of the nostrils in atrophic rhinitis. *J Laryngol Otol* 1967;**81**:515–24

Address for correspondence:
Miss A Soni-Jaiswal,
Apt 5, 22 Larke Rise,
Didsbury, Manchester M20 2UL, UK.

Fax: 01614348051
E-mail: archanasj@gmail.com

Miss A Soni-Jaiswal takes responsibility for the integrity of the content of the paper.
Competing interests: None declared