Case Report

Long Term Complete Epistaxis Relief in Hereditary Hemorrhagic Telangiectasia: A Case Report

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Abstract

Hereditary hemorrhagic telangiectasia (Osler-Weber-Rendu disease), is an autosomal dominant genetic disorder in which telangiectasias, arteriovenous malformations, and aneurysms may be widely distributed throughout the body vasculature. The most prominent clinical manifestation is recurrent epistaxis that present considerable variability and severity, which often leads to alterations in social functioning and quality of life. Various treatments are recommended for clinical management to reduce frequency, duration, and intensity of epistaxis including surgical and non-surgical methods. New approaches are warranted as current methods do not provide a cure or guarantee lasting relief. Here we present the case of a 40-year-old female with a history of over three decades of consistent HHT-related epistaxis, who has found long-term complete relief due solely to dietary modifications.

Keywords: epistaxis, hht, hereditary hemorrhagic telangiectasia, diet, osler-weber-rendu

Introduction

Hereditary hemorrhagic telangiectasia (HHT), also known as Osler-Weber-Rendu syndrome, is an autosomal-dominant disorder that leads to the development of abnormal vessels [1]. This disease affects nearly one in every 5,000 individuals and represents a significant health burden [2]. The most common symptom of HHT is recurring epistaxis, often severe, and occurring in over 90% of HHT patients [3]. Diagnostic criteria are outlined by the Curacao criteria, consisting of the following: 1) epistaxis 2) family history 3) intra nasal telangiectasias and 4) AVMs. A patient with 3 out of 4 of these constitutes a probable diagnosis of HHT, and 2 out of 4 constitutes a possible diagnosis of HHT [4].

At the present time, there is no cure for HHT or the associated recurrent and life-long epistaxis. Current treatment modalities range from non-invasive treatments that frequently fail to achieve even short-term control to surgeries and systemic therapies that carry significant risk of complications [5]. Lifestyle and dietary influence have been shown to influence occurrence and severity of epistaxis from HHT [6], and certain foods have also shown to be triggers for onset of epistaxis and related symptoms [7]. Certain dietary ingredients which have been reported to exacerbate nosebleeds include foods with high salicylate content (such as red wine, spices, chocolate, and coffee), with natural antiplatelet activity (such as ginger, garlic, ginkgo biloba, and ginseng), and with high levels of omega-3 acids (such as salmon) [6,7]. No studies, however, have identified a dietary intervention or protocol for specific food elimination which completely resolves incidence of epistaxis for HHT. Further exploring a possible association between
epistaxis and specific dietary intake or avoidance may immediately provide HHT patients with a feasible treatment for self-management.

In this article, the authors report a case of a typical HHT patient to contribute to the literature for treatment of epistaxis caused by HHT, which currently lacks clinical reports on the same. The long-term outcome for this patient was absolute relief from HHT related epistaxis.

**Case Report**

A 40-year-old female with HHT ACVRL-1 diagnosis confirmed in 2011 by genetic analysis recently presented to a tertiary care Rhinology clinic. She had a history of recurrent epistaxis since age 6 with average frequency of 3-5 episodes per week steadily through the years. Physical exam revealed telangiectasias on the lips, tongue, fingers and interior of nose which do not currently bleed. Previous screening for arteriovenous malformations in the brain and lungs have come back negative.

Patient has history of two KTP laser surgeries for intra-nasal and tongue telangiectasias. These were successfully completed in 2011 and 2015 under general anesthesia. Following each treatment, patient reported short term relief from epistaxis lasting approximately 3-6 months; however, epistaxis eventually recurred.

Evaluation included a screening trans-thoracic bubble echo which was negative for lung arteriovenous malformations. In conversation, patient mentioned she has not had any epistaxis in the past three years. This finding was extraordinary considering her history of consistent epistaxis for over the last three decades. The patient explained she had drastically changed her diet as a therapeutic modality for symptoms of an autoimmune disorder, Hashimoto’s Thyroiditis, which is unrelated to HHT. No other changes were reported for lifestyle or medical treatment.

The dietary intervention consisted of total elimination of all gluten protein and dairy products. The patient started this diet in May 2016 and had noticed relief of epistaxis by June 2016. Since introduction of the elimination diet the patient has stated there has been absolute relief from epistaxis and no new telangiectasia formations on her body that she has noticed in the past three years.

**Discussion**

Recurrent epistaxis can severely affect quality of life and present precarious medical outcomes for HHT patients. Currently, treatments aim at reducing the frequency and severity of the epistaxis. Non-surgical treatments include moisturization, topical ointments like mupirocin or Vaseline, topical estrogen, and bevacizumab [8]. Adjuvant non-surgical treatments include blood transfusions and iron dextran infusions. Another aggressive non-surgical treatment is arterial embolization. Surgical treatments include laser treatments, cauterization, sclerotherapy, septodermoplasty, and the most aggressive treatment is nasal closure, also known as the Young’s procedure [9]. The Young’s procedure is reserved for severe, transfusion dependent epistaxis or life-threatening epistaxis refractory to other aggressive treatments. The senior author advocates for performing unilateral Young’s procedure on the most severe side prior to bilateral, given the drastic side effects like complete nasal obstruction, loss of sense of smell, and possible obstructive sleep apnea.

None of the above-mentioned treatments provide a cure or even sustained relief from HHT-related epistaxis, and many treatments like embolization and surgery have significant potential side effects. Thus, given the improvement in epistaxis in the current patient with what can be deemed a low-risk or even no-risk treatment (lifestyle and dietary modification), it stands to reason that this approach warrants further investigation. Given the shortcomings and potential drawbacks of current treatments for epistaxis in HHT, there is a need for new therapeutic options. Although food restrictions may not be acceptable to some, dietary intervention for relief of epistaxis could be an inexpensive alternative to pharmaceutical and surgical techniques and offers self-management for patients.
Sharing the findings from this case report in practice to HHT patients may be a valued suggestion before intensive measures are offered. Dietary modification should be a matter for patient choice, however, if epistaxis has significant impact on patients’ lifestyle and general health then there is an argument that dietary advice could be part of clinical management. Advisement to try a risk-free strategy of food avoidance and monitoring of dietary intake and epistaxis episodes could serve as a marker to protect against other sequelae from HHT.

This case report calls for investigation into exactly which foods may need to be avoided for patients with HHT who suffer from epistaxis. This patient removed two major foods, gluten and diary, and it is unknown which, or the combination of both, produced complete epistaxis relief for nearly three years. It is also unknown if internal telangiectasias were affected or prevented by the exclusion of dairy and gluten products in this patient, however, a recent test cleared this patient of lung arteriovenous malformations. Controlled studies are recommended to determine which food(s) specifically may need to be excluded for optimal outcome.

Ethical Considerations
- The confidentiality of the identity of the patient has been ensured
- The patient has been informed of the publication of the case
- The case report is in the best interest of the community and to create awareness among otolaryngologist and general health professionals
- Our institution does not require ethical approval for case reports

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References