

Novel Use of Nasal Septal Button for Epistaxis in HHT Syndrome: An Observational Study

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Abstract

Osler-Rendu-Weber or HHT (Hereditary Hemorrhagic Telangiectasia) is a genetic disease that causes patients to bleed, especially from the nose (epistaxis). The management of epistaxis in HHT is often very complex and frustrating. Epistaxis affects patient's quality of life but can also be life threatening highlighting the importance of finding a permanent solution for this problem. Multiple management strategies have been proposed: packing, chemical and electrical cauterization, septodermoplasty, embolization, SPA ligation, antifibrinolytic agents. In the present study, we discuss the case of three patients with HHT and recalcitrant epistaxis who failed most of these strategies. The placement of nasal silicone septal button was shown to incidentally decrease significantly the incidence of bleeding in one patient by isolating the septal mucosa. The same procedure was then offered to the two other patients with similar success in decreasing bleeding and transfusion rate over 4 years of follow up.

Keywords: Osler; Rendu; Weber; Epistaxis; Silicone button

Introduction

Osler-Rendu-Weber, or Hereditary Hemorrhagic Telangiectasia (HHT) is an autosomal dominant disorder that affect blood vessels in mucocutaneous tissues and visceral organs, and results in a tendency for bleeding [1]. The underlying pathology is a defect in the vascular wall that begin with dilation of postcapillary venules and leads to expansion of the upstream vessels causing arteriovenous shunts [2]. While epistaxis is the most encountered symptom for otolaryngologists, there are many other manifestations such as arterio-venous malformations of the lungs, and Gastrointestinal (GI) tract, central nervous system, liver, spleen and urinary tract. Recurrent and severe epistaxis is the most common presentation, frequently leading to severe anemia that requires multiple blood transfusions. These episodes can range from minimally bothersome to life-threatening. Epistaxis affect almost 100% of the patients by the age of 40. [3]. As such, otolaryngologist: Head and Neck surgeons are frequently involved in the management of these patients. The Epistaxis Severity Score is a validated and useful outcome measure that was designed specifically for HHT and can be easily used to gauge disease severity and the effect of treatment. There are various treatment options available for epistaxis in HHT, with many new advances and innovations being developed: Nasal packing, hospitalization, cauterization in the operation room, and frequent blood transfusion limiting patient's quality of life. Diagnostic criteria, referred to as the Curaçao criteria, include epistaxis, telangiectases, visceral lesions, and affected first degree relatives. Relative frequencies of HHT are not discussed in this article. Nasal septal button has been commonly and safely used since 1970, for non-surgical closure of septal perforation. The aim of this study is to shed the light on a potential role of nasal septal button in decreasing the rate of epistaxis in patients suffering from Osler- Rendu syndrome.

Case Report

We present the case of 3 patients diagnosed with Hereditary Hemorrhagic Telangiectasia according to Curaçao criteria, who end up having nasal septal button, after multiple trial of many treatment strategies. The first patient is a 46-years- old female, suffering from epistaxis since the age of 14. She had multiple severe epistaxis treated with electrical cauterization in the operation room and had multiple hospitalizations and blood transfusions. She has a positive family history of Osler-Rendu. A total body MRI did not reveal any other visceral

involvement (except for the tongue). She had a silicone nasal septal button inserted through a septal perforation four years ago, initially as an attempt to provide additional packing material after failure of both electrical cauterization and conventional pope packing. After the button placement, the patient experienced a significant decrease in her nose bleeds both in rate and abundance. The septal button was changed three times ever since with some complains of crusting and irritation of the surrounding mucosa reported by patient. This experience has led us to consider the potential role of septal button in decreasing the rate of serious epistaxis in HHT patients.

The second patient is a 40-years-old man, who suffered from heavy epistaxis since adolescence. He has a positive family history of Osler-Rendu. On clinical exam, he presented with multiple lingual and facial telangiectasia. Because of the common occurrence of epistaxis, patient was even instructed to do self-packing with pope to reduce his visits to the Emergency Department. He received blood transfusions on average every 6 months to compensate for blood loss. Considering our previous experience with nasal septal button, patient was offered nasal splitting 2 years ago. After this procedure, the patient presented with no episode of epistaxis, and this has significantly improved his quality of life, by reducing blood transfusion, and self-packing to none.

The third patient is a 75-years-old male, with a history of Abdominal Aorta Aneurysm replacement, multiple liver hemangiomas, lung arteriovenous malformation, pulmonary hypertension, and moderate heart failure. He has a positive family history of HHT having a sister and two daughters suffering from the disease. He had suffered from epistaxis for more than 10 years, treated with multiple chemical and electrical cauterizations and had also turned to self-packing to reduce the cost of his recurrent hospitalizations. An urgent endoscopic SPA ligation (sphenopalatine artery ligation) was performed on one occasion to stop

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a severe epistaxis. He was also offered an endoscopic septodermoplasty only to have the bleeding episodes recur a few months later. He was prescribed Etamsylate as well as Bevacizumab to control of bleeding with little to no benefits, decreasing the rate of transfusions from 7 to 5 times a year. A nasal septal nasal septal button was inserted bilaterally through a perforation created de novo for this purpose. After 16 months of the surgical procedure, the patient present with no serious episodes of epistaxis. He comes to clinic every month for a follow up and debridement. He has not had a blood transfusion ever since.

Discussion

According to the guidelines published by the French society published in 2017 in the European Annals of otolaryngology, the first-line treatment for acute epistaxis in Rendu-Osler-Weber disease is bidigital compression after nasal cleansing. In case of failure, resorbable packing should be used in association with antibioprophyllaxis, Embolization is reserved for persistent epistaxis and signs of anemia and decrease in hemoglobin. Non resorbable packing and chemical or electrical cauterization are not recommended [4]. Many trials were done on estrogen-related hormones and antifibrinolytic agents, pulsed dye laser treatment, thalidomide [5], septal dermoplasty [6], endovascular embolization, N-acetylcysteine and bevacizumab with variable rates of success. According to Al-Samkari et al. Epistaxis control was achieved in 85% with bevacizumab versus 0% before treatment and reduced red cell units transfused by 92% units vs. 0%. [7] However many other studies, aka Dupuis-Girod et al. found that bevacizumab did not reduce epistaxis compared with placebo (59 patients treated with different doses of bevacizumab did not present any decrease in the epistaxis rate compared to placebo group) [8]. To our knowledge, this is the first description of a potential role of septal button in decreasing nose bleeds

In the present report, the case of three HHT patients with severe recurrent epistaxis were presented. All these patients underwent multiple interventions in order to control of bleeding with little to no success. All three has a septal button placed through an existing or especially created septal perforation. This procedure has resulted in significant decrease in the rate of serious epistaxis or blood transfusion with a the longest follow up being of 4 years. We believe that septal

button achieves this by isolating the septal mucosa thereby preventing desiccations and trauma to the fragile septal vasculature. There is also a potential role in decreasing (by compression) the blood flow reaching the anterior septal area. All three patients presented in the study tolerated the septal button placement well despite the long follow up. Nevertheless, potential colonization with bacteria or erosion or necrosis of the septal mucosa cannot be dismissed although the benefit of having a significant decrease in bleeding seems to outweigh these risks.

Conclusion

Based on this observational study on three patients, bilateral silicone button placement might be a promising alternative for refractory epistaxis in HHT patients who have failed more conservative measures.

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