

Randy W. Lesh, MD, Vincent Desiato, DO, Kevin J. Kovatch, MD, Nicholas C. Purdy, DO, and Thorsen W. Haugen, MD
 Department of Otolaryngology - Head and Neck Surgery, Geisinger Health System, Danville, Pennsylvania, USA

Introduction

Hereditary hemorrhagic telangiectasia (HHT) is an autosomal dominant, multisystem angiomyolipomatous disorder¹

Recurrent epistaxis occurs in nearly 100% of cases by age 40

Epistaxis related to HHT can be life-threatening and decreases QoL

Numerous topical, ablative, systemic, and surgical treatments have been utilized with varying degrees of success²

Septodermoplasty decreases the need for blood transfusions and improves quality of life (QoL)

Current medical and surgical options are limited, particularly in the advanced stages of the disease

Nasal resurfacing using radial forearm free flap through bilateral lateral rhinotomy incisions has been described³

Contact

Randy W. Lesh, MD
 Otolaryngology – Head & Neck Surgery
 Geisinger Health System
 100 N. Academy Avenue, Danville PA
 rlesh2@geisinger.edu

Case

73-year-old man with HHT

History of HTN, BPH, Afib s/p ablation, left RCC s/p nephrectomy in 2017, lung AVMs embolized in 2017

Epistaxis started around age 25, up to 5x per day, choking on nightly bleeding, IV iron PRN

Declined nasal closure

Resurfacing endoscopic free flap (REFF)

1. Total septectomy
2. Bilateral endoscopic medial maxillectomies
3. Inferior and middle turbinate resections
4. Left Caldwell-Luc antrostomy
5. Left neck exploration for donor vessels
6. Right radial forearm fasciocutaneous free flap used to resurface nasal cavity
7. Nasal cavity packed with foley catheter balloon and NasoPore

Results/Discussion

Minimal epistaxis with significantly improved QoL

Epistaxis Severity Score improved from 8.9 to 3.5

Underwent revision surgery with debulking of flap and DCR for epiphora

Future work will revise inset technique using 3D printed skull to map placement

Treatment History

Medical:

Topical moisturization

Doxycycline trial, had initial improvement but benefit waned

Avastin (bevacizumab) trial, did not have improvement

PO TXA 1,300 mg TID with improvement

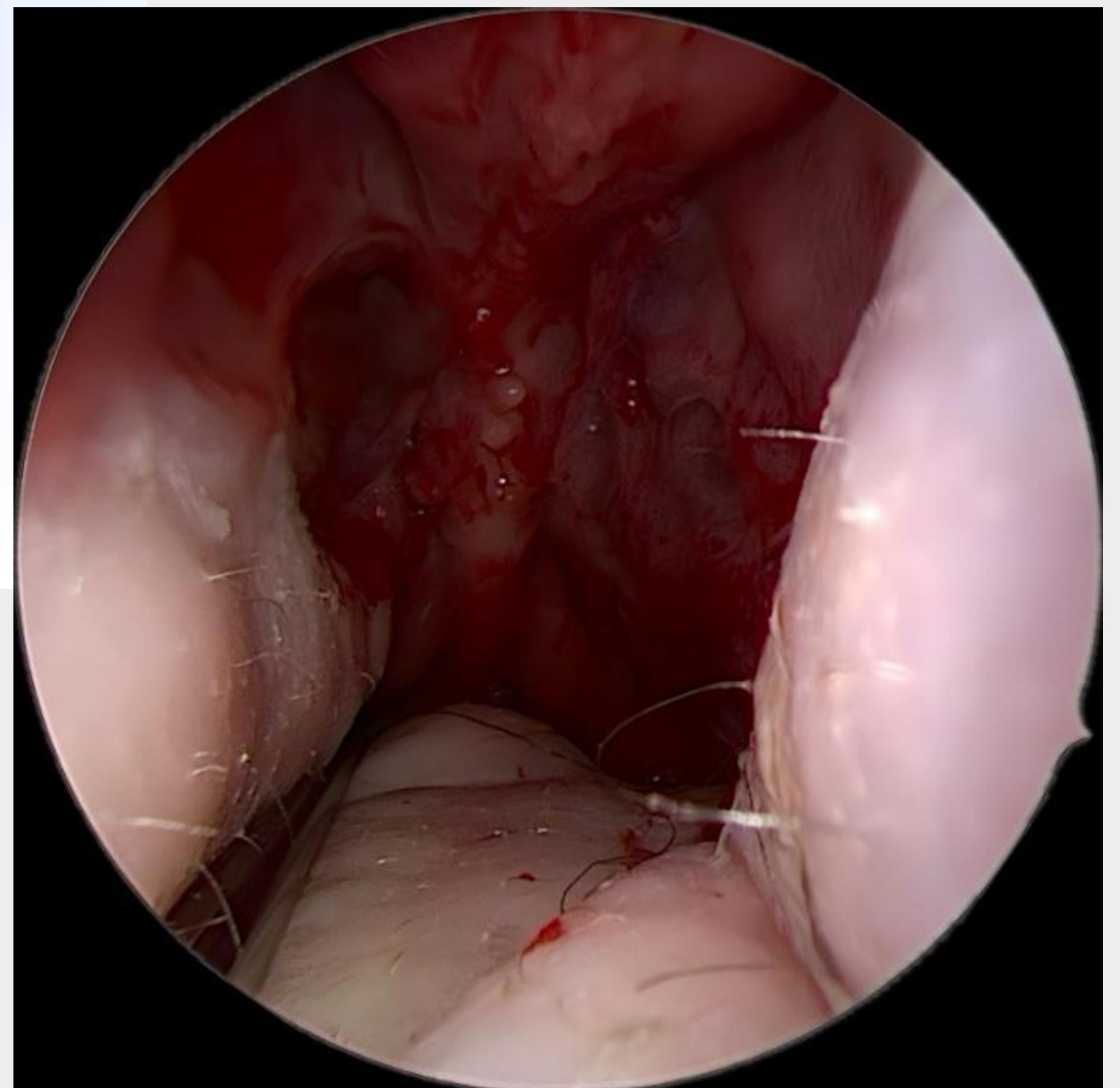
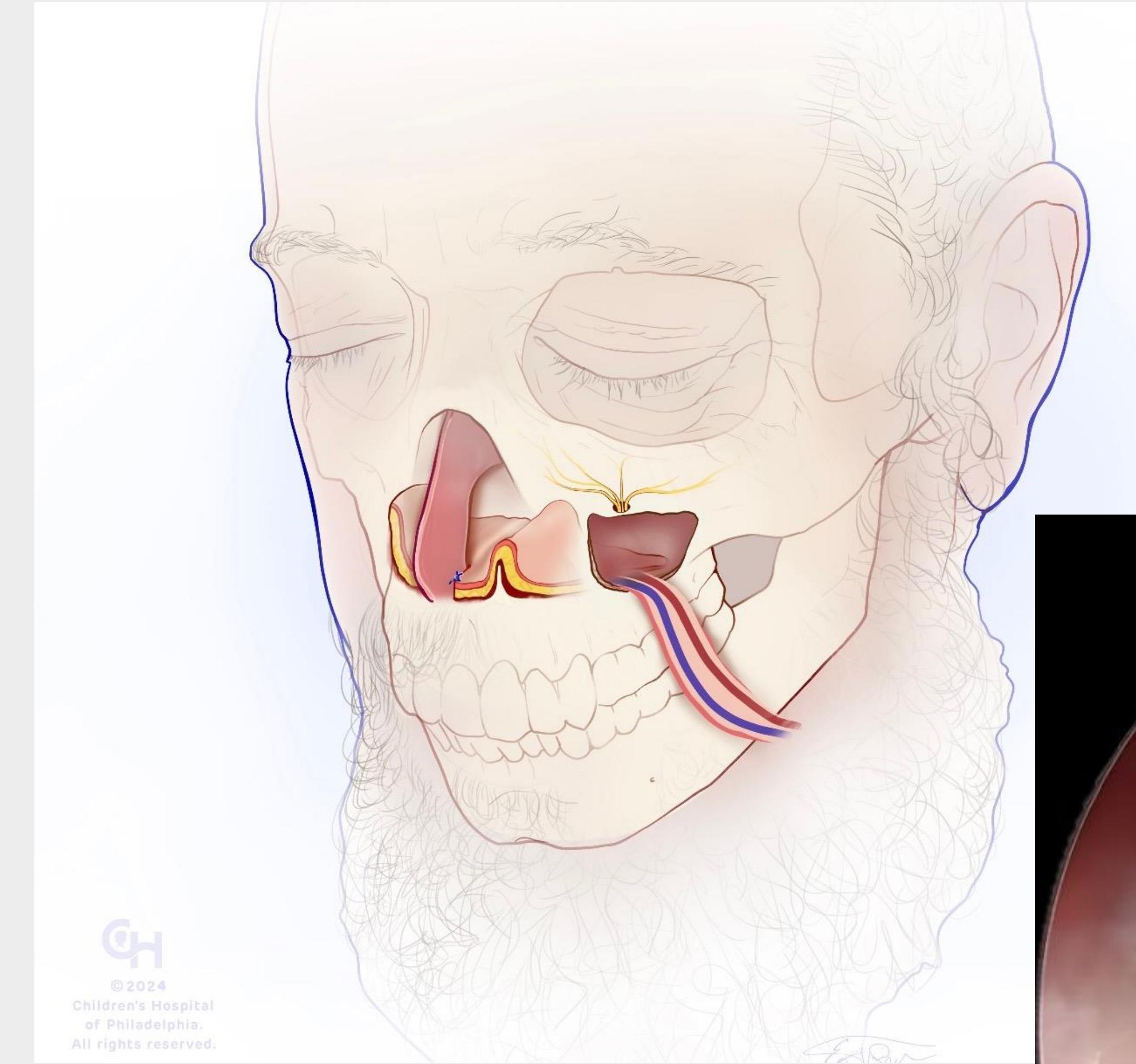
Topical TXA PRN

Surgical:

Numerous cauterizations

Sclerosing agents

Septodermoplasty



References

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