Dieulafoy’s Lesion of the Nasal Mucosa: A Case of Recurrent Epistaxis From Submucosal Arterial Malformation

Prithwijit Roychowdhury¹, Ali Akalin, MD², and Christopher J. Ito, MD³

Keywords
Dieulafoy's lesion, epistaxis

Received February 25, 2020; accepted April 23, 2020.

Dieulafoy’s lesions (DLs) were first described by the French surgeon Georges Dieulafoy in 1898 in his report of 3 cases of upper gastrointestinal (GI) bleeding in the proximal stomach, resulting in fatal gastric hemorrhage in otherwise asymptomatic young men.¹ While most cases present in the stomach or along the GI tract, there are reports of the lesion in extra-gastrointestinal locations such as the bronchus and head and neck.² In the head and neck, the pathology is conserved but is referred to as either cirsoid aneurysm (on the scalp) or submucosal arterial malformation.³ DLs are a source of profound bleeding, and to our knowledge, this is the first report of a DL as a source of recurrent epistaxis.

Case Report
This report was exempt from review by the University of Massachusetts Institutional Review Board.

Presentation
A 55-year-old man with no significant medical history presented to a tertiary care otolaryngology clinic for evaluation of recurrent unilateral epistaxis. A vascular 1-cm submucosal lesion was identified on nasal endoscopy and was suspected to be the source of the epistaxis, often triggered by coughing and sneezing. The lesion was excised under endoscopic guidance and the recurrent epistaxis resolved. A gross specimen and a hematoxylin and eosin–stained specimen were examined. Histopathology confirmed the mass to be a DL.

Endoscopy
Endoscopically, a DL typically appears as an isolated protruding vessel surrounded by normal mucosa, without an associated ulcer.¹ In this case, endoscopy revealed a friable raised lesion with surrounding hyperemic mucosa on the right nasal floor (Figure 1). The lesion was located beneath the inferior turbinate and required infracture and superior displacement of the turbinate to allow room for excision.

Gross Pathology
A DL is a histologically normal vessel with an abnormally large diameter (1-3 mm) that completes an indirect course within the submucosa and protrudes through a mucosal defect (2-5 mm) without surrounding ulceration.¹

Figure 1. Endoscopic image of the Dieulafoy’s lesion on the nasal floor.

¹University of Massachusetts Medical School (UMMS), Worcester, Massachusetts, USA
²Department of Pathology, University of Massachusetts Medical School, Worcester, Massachusetts, USA
³Department of Otolaryngology, University of Massachusetts Medical School, Worcester, Massachusetts, USA

This article was presented at the AAO-HNSF 2019 Annual Meeting & OTO Experience; September 15-18, 2019; New Orleans, Louisiana.

Corresponding Author:
Prithwijit Roychowdhury, University of Massachusetts Medical School, 55 Lake Avenue North, Worcester, MA 01655, USA.
Email: Prithwijit.roychowdhury@umassmed.edu

This Creative Commons Non Commercial CC BY-NC: This article is distributed under the terms of the Creative Commons Attribution-NonCommercial 4.0 License (http://creativecommons.org/licenses/by-nc/4.0/) which permits non-commercial use, reproduction and distribution of the work without further permission provided the original work is attributed as specified on the SAGE and Open Access pages (https://us.sagepub.com/en-us/nam/open-access-at-sage).
Histopathology
Histology shows a large tortuous vessel underneath an area of thinned sinonasal mucosa with seromucinous glands (Figure 2).

Pathophysiology
While the pathogenesis is not well understood, the prevailing theory is as follows: pulsations in a large, tortuous, submucosal arteriole can lead to the disruption of overlying epithelium (which may be intrinsically vulnerable). Epithelial disruption results in areas of localized ischemia and allows for further superficial mucosal erosion and resulting rupture. Of note, DLs are pathologically distinct from other vascular lesions; they do not contain arterial and venous intercommunications that characterize arteriovenous malformations, and unlike hemangiomas, DLs consist of a single circuitous large-caliber vessel rather than a collection of numerous small vessels. In addition, they are generally unifocal and noninherited, unlike hereditary hemorrhagic telangiectasias (HHT).

Treatment
The management of DL depends on the specific presentation and location of the lesion. In the GI tract, endoscopic combination therapies (epinephrine with either fulguration or mechanical monotherapies) are preferred initially, followed by angiofurgy with embolization. Failure of either of the former approaches necessitates surgical excision.

In this case, the lesion was excised endoscopically in the operating room, and on follow-up, the patient showed no signs of recurrent epistaxis. Fulguration in the nasal cavity would have been a possible initial treatment, but excision allowed for pathologic examination to confirm the diagnosis. Electrocautery was still required to achieve postsurgical hemostasis.

Discussion
DL is a rare vascular lesion that has been identified primarily in the stomach and GI tract and is recognized as a source of GI hemorrhage. Given our experience here, DL is a clinical entity that should also be considered in the differential diagnosis of recurrent epistaxis for patients with a history negative for trauma, inflammation, underlying hematologic pathology, and structural abnormalities of the nasal mucosa and septum. Therefore, the case presented here demonstrates the necessity of endoscopic evaluation for recurrent and unilateral epistaxis in the clinic. If appropriately recognized, management of DLs can be completed with surgical excision, avoiding additional procedures such as embolization or sphenopalatine artery ligation.

Author Contributions
Prithwijit Roychowdhury, study design, drafting of manuscript, final approval, and accountable for the work; Ali Akalin, case review/study design, drafting of manuscript, final approval, and accountable for the work; Christopher J. Ito, study design, drafting of manuscript, final approval, and accountable for the work.

Disclosures
Competing interests: None.
Sponsorships: None.
Funding source: None.

References
1. Chaer RA, Helton WS. Dieulafoy’s disease. J Am Coll Surg. 2003;196(2):290-296.
2. Qian X, Du Q, Wei N, et al. Bronchial Dieulafoy’s disease: a retrospective analysis of 73 cases. BMC Pulm Med. 2019;19(1):104.
3. Gurkanlar D, Gonul M, Solmaj I, et al. Cirsoid aneurysms of the scalp. Neurosurg Rev. 2006;29(3):208-212.
4. Nguyen DC, Jackson CS. The Dieulafoy’s lesion: an update on evaluation, diagnosis, and management. J Clin Gastroenterol. 2015;49(7):541-549.
5. Beck R, Sorge M, Schneider A, Dietz A. Current approaches to epistaxis treatment in primary and secondary care. Dtsch Arztebl Int. 2018;115(1-2):12-22.