OPEN ACCESS
ISSN: 2835-9496
Schenck NL, et al. (2025): 1320
Ameri J Clin Med Re, 2025; 5(11): 100245
DOI: 10.71010/2835-9496/ajcmr-e245

Research Article
American Journal of Clinical and Medical Research

Recurrent Epistaxis: A Rare Complication of Dermatomyositis

(***Running Title:*** *Recurrent Epistaxis in Dermatomyositis*)

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Citation: Schenck NL, Moradzadeh D, Alvarado S (2025) Recurrent Epistaxis: A Rare Complication of Dermatomyositis. Ameri J Clin Med Re: AJCMR-245. *https://doi.org/10.71010/2835-9496/ajcmr-e245*.

Received Date: 24 October, 2025; **Accepted Date:** 03 November, 2025; **Published Date:** 07 November, 2025

Case Presentation

We present the case of a 53-year-old female with known dermatomyositis, who presented with recurrent episodes of epistaxis. She initially experienced several mild nosebleeds from the left nostril, which progressed to severe, spontaneous epistaxis from the right nostril. Examination revealed diffuse bleeding from the entire Kiesselbach's plexus rather than a single focal vessel. Hemostasis required electric cauterization and nasal packing for five days. She denied any recent upper respiratory infection, nasal trauma, or anticoagulant use. Coagulation studies, including prothrombin time (PT) and partial thromboplastin time (PTT), were within normal limits. At presentation, her medications included prednisone 25 mg daily, hydroxychloroquine 200 mg daily, and regular intravenous immunoglobulin (IVIG) infusions. Physical examination revealed characteristic dermatomyositis findings, including erythema over the face and hands. She reported a history of dysphagia, chest rash, and fatigue. Post-epistaxis treatment included saline spray, saline gel, and 2.5% hydrocortisone ointment applied into the nostrils twice daily; however, these measures did not prevent recurrent nosebleeds.

Discussion

Dermatomyositis (DM) is a rare autoimmune inflammatory disease characterized by muscle weakness and skin rashes [1]. It primarily affects endomysial capillaries, which surround individual muscle fibers, and perimysial capillaries, which surround bundles of muscle fibers in skeletal muscle, as well as superficial dermal capillaries in the skin. This leads to the deposition of C5b-9, a membrane attack complex that mediates immune-driven cell damage, resulting in capillary loss and vessel wall thickening [1]. These vascular changes contribute to the muscle weakness and characteristic skin rashes seen in DM. In some cases, dermatomyositis acts as a paraneoplastic syndrome triggered by an underlying malignancy. Studies estimate that adult patients with DM have a six-fold increased risk of associated cancers, with breast, ovarian, lung, and gastrointestinal malignancies being the most common [2].

Beyond muscle and skin involvement, dermatomyositis can cause systemic complications including pulmonary disease, cardiac inflammation, and gastrointestinal dysmotility [3]. Oropharyngeal and esophageal dysfunction are well-documented, with esophageal involvement occurring in up to 50% of patients [4]. This often presents as dysphagia secondary to inflammation and weakness of the upper esophageal musculature.

Although vascular involvement is a hallmark of dermatomyositis, manifestations affecting the nasal mucosa are infrequently described [1,2,4,5]. In the references cited for dermatomyositis, recurrent epistaxis is rarely mentioned as a complication [1,2,4,5]. When it occurs, epistaxis may reflect fragile nasal blood vessels or inflammation of the nasal mucosa [1]. However, the precise pathophysiology and incidence of this manifestation remain poorly understood, highlighting its underrecognized clinical significance.

Epistaxis is a common condition with many causes, most of which are benign and self-limiting. Common local contributors include trauma, dry air, upper respiratory infections, allergic rhinitis, and use of nasal sprays or anticoagulant medications. Systemic causes encompass coagulopathies, hypertension, vasculitis, and, rarely, autoimmune diseases such as dermatomyositis [3].

In our patient, recurrent and ultimately severe epistaxis occurred without any identifiable common triggers. There was no history of infection, trauma, environmental irritants, or structural nasal abnormalities, and her coagulation profile was normal. The bleeding originated diffusely from Kiesselbach's plexus, a highly vascularized area of the inferior nasal septum. See Figure 1 below [5].

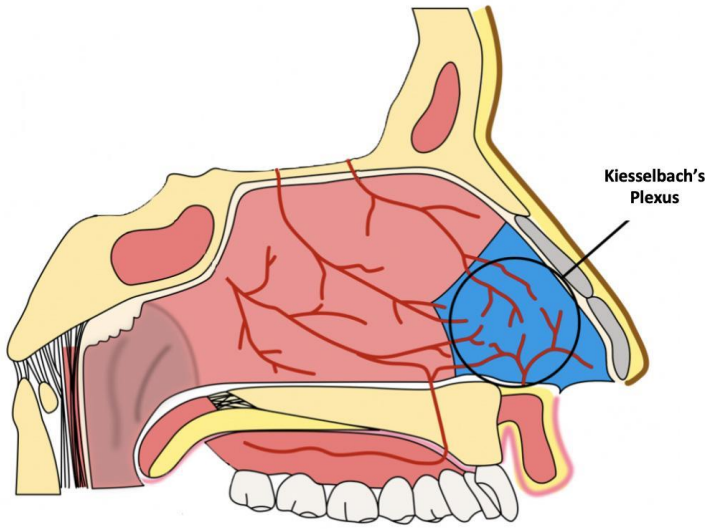

Figure 1 (5). Illustration of Kiesselbach's Plexus showing the site of the patient's most massive bleed.

Unlike most epistaxis cases that arise from a single ruptured vessel, involvement of the entire plexus suggested diffuse mucosal fragility. Although epistaxis is an uncommon manifestation of dermatomyositis, underlying vascular inflammation may increase the susceptibility of the nasal mucosa to bleeding. Intravenous immunoglobulin (IVIG), a standard treatment for dermatomyositis shown to improve dermatological symptoms, could theoretically contribute to epistaxis, though evidence for this remains unclear [6].

Conclusion

Epistaxis in this patient was persistent and unresponsive to conservative management, necessitating multiple interventions including extensive cauterization and prolonged nasal packing. Although rare, this case suggests that recurrent epistaxis may represent an atypical manifestation of dermatomyositis, especially in patients without known coagulopathies or other predisposing factors. The involvement of the nasal mucosa likely reflects underlying small-vessel vasculopathy, a recognized feature of the disease. This unusual presentation underscores the importance of considering dermatomyositis as a potential diagnosis in patients presenting with unexplained recurrent epistaxis.

Conflicts of Interest: The authors declare that there are no conflicts of interest regarding the publication of this case report. All authors have reviewed and approved the final manuscript and affirm that there are no financial, personal, or professional relationships that could be perceived as influencing the work reported in this paper.

Clinical Trial Registry and Grants: Not applicable

Funding: None

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