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**Case Report**

# Hereditary hemorrhagic telangiectasia presenting with severe nasal bleeding: an ENT perspective

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## ABSTRACT

Hereditary hemorrhagic telangiectasia (HHT), also known as Osler–Weber–Rendu syndrome, is a rare autosomal dominant vascular disorder characterized by mucocutaneous telangiectasia and visceral arteriovenous malformations. Recurrent epistaxis is the most common and often the earliest manifestation, frequently bringing patients to the otorhinolaryngologist. We report a case of a patient presenting with recurrent, spontaneous, and progressively severe nasal bleeding. Anterior rhinoscopy and diagnostic nasal endoscopy revealed multiple, discrete, bilateral telangiectatic lesions over the nasal septum, floor of the nasal cavity, and inferior turbinates, which bled on touch, making cauterization difficult at certain sites. Mucocutaneous telangiectasia over the lips and oral cavity with a positive family history supported the diagnosis. Based on clinical findings and established diagnostic criteria, a diagnosis of definite Hereditary Hemorrhagic Telangiectasia was made. The patient was managed with local nasal measures and supportive therapy. This case highlights the importance of early recognition of HHT by ENT specialists to enable timely diagnosis, appropriate management, and prevention of systemic complications.

**Keywords:** Hereditary hemorrhagic telangiectasia, Epistaxis, Telangiectasia, ENT manifestations

## INTRODUCTION

Hereditary hemorrhagic telangiectasia (HHT), also known as Osler–Weber–Rendu syndrome, is a rare autosomal dominant vascular disorder characterized by abnormal angiogenesis leading to mucocutaneous telangiectasia and visceral arteriovenous malformations. The disease exhibits considerable clinical heterogeneity, with manifestations ranging from mild mucosal bleeding to potentially life-threatening systemic complications. The estimated prevalence of HHT is approximately 1 in 5,000–8,000 individuals.[2-5]

Recurrent epistaxis is the most common and often the earliest manifestation of HHT, occurring in more than 90% of affected individuals.[1-3,8] Nasal bleeding typically begins in childhood or early adulthood and shows a progressive increase in frequency and severity with advancing age.[2,3,8]

The nasal septum, particularly the anterior septal region, along with the inferior turbinates and floor of the nasal cavity, are commonly involved due to the presence of fragile telangiectatic vessels within the nasal mucosa.[10,11]

Patients with HHT frequently present with spontaneous, unilateral or bilateral epistaxis that is disproportionate to local inflammatory findings and may be resistant to conventional nasal cauterization.[6,8,10]

As epistaxis often precedes the recognition of systemic arteriovenous malformations by several years, otorhinolaryngologists are commonly the first clinicians to encounter these patients.[1-3,5] Early recognition of HHT is essential, as timely diagnosis permits appropriate screening for visceral involvement and reduces the risk of serious complications.[1,2,5]

## CASE REPORT

A 54-year male patient presented to the otorhinolaryngology outpatient department with complaints of recurrent, spontaneous nasal bleeding for several years. The episodes were bilateral, sudden in onset, and not associated with trauma, infection, or use of anticoagulant medications. The frequency and severity of epistaxis had increased over time, leading to symptoms suggestive of anemia.

General examination revealed pallor. Cutaneous and mucosal examination demonstrated multiple telangiectatic lesions over the lips and oral cavity. Anterior rhinoscopy and diagnostic nasal endoscopy revealed multiple, discrete, bilateral telangiectatic lesions over the nasal septum, floor of the nasal cavity, and inferior turbinates, which bled on touch, making cauterization difficult at certain sites.

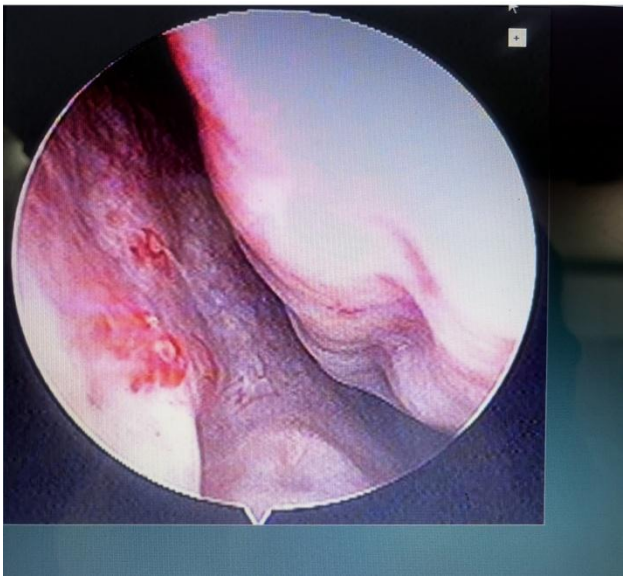

**Figure 1: Multiple telengectatic vessels on the nasal septum.**

Laboratory investigations revealed reduced hemoglobin levels with normal platelet count and coagulation profile. A positive family history of recurrent epistaxis was elicited. Based on recurrent epistaxis, mucocutaneous telangiectasia, and positive family history, the patient fulfilled the diagnostic criteria for definite HHT.

## DISCUSSION

HHT is a genetically determined vascular disorder resulting from mutations affecting normal angiogenesis.[2-5] These abnormalities lead to the development of fragile, dilated vessels that are prone to rupture, particularly at mucocutaneous surfaces. The nasal mucosa is especially vulnerable due to its rich vascularity and exposure to repeated minor trauma and mucosal dryness.[10,11]

Epistaxis remains the hallmark and most frequent clinical manifestation of HHT. Nasal bleeding is often the initial symptom and may progressively worse, resulting in chronic blood loss and iron-deficiency anemia.[2,3,8] Characteristic telangiectatic lesions are commonly located over the nasal septum, inferior turbinates, and floor of the nasal cavity, as observed in the present case.

Epistaxis in HHT is unilateral or bilateral, spontaneous, and recurrent, and may be resistant to routine therapeutic measures.[6,8,10] Telangiectatic lesions tend to bleed on minimal manipulation, posing challenges during endoscopic cauterization and often necessitating repeated interventions.[6,8] Early identification of HHT is crucial, as epistaxis may be the sole presenting feature for several years before systemic involvement becomes apparent.

The diagnosis of HHT is primarily clinical and is based on established diagnostic criteria that include recurrent epistaxis, mucocutaneous telangiectasia, visceral arteriovenous malformations, and a positive family history.[1,12] Prompt diagnosis allows appropriate screening for pulmonary, cerebral, and gastrointestinal arteriovenous malformations, thereby reducing long-term morbidity and mortality.[1,2,5]

Management of epistaxis in HHT is aimed at symptom control and improvement of quality of life.[1,6-9] Local measures such as nasal packing, endoscopic cauterization, and nasal moisturizing agents remain the mainstay of treatment. Given the chronic and recurrent nature of the disease, a multidisciplinary approach and long-term follow-up are essential.[1,2,5]

## CONCLUSION

Recurrent spontaneous epistaxis should prompt consideration of HHT, particularly in the presence of mucocutaneous telangiectasia and positive family history. Early recognition by otorhinolaryngologists is essential for timely diagnosis, effective management, and prevention of potentially serious systemic complications.

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