Intractable Epistaxis and Systemic Lupus Erythematosus:
High-Dose Intravenous Pulse Steroid Therapy

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Abstract

Objectives:
To describe a novel therapy for the treatment of intractable and refractory epistaxis in a subset of patients with poorly controlled systemic lupus erythematosus. Clinical presentation, treatments/interventions, and a discussion of the limited relevant literature are presented.

Study Design:
Case report and review of the literature

Methods:
A patient's case was reviewed. A MEDLINE search was performed using the terms: epistaxis AND steroid ADN systemic lupus erythematosus. All existing literature on the treatment of vasculitides in relation to systemic lupus erythematosus was extracted.

Results:
This case report describes the hospital course of a patient with severe SLE and intractable epistaxis. We discuss classic management options for epistaxis and offer a novel treatment option for patients with SLE-related vasculitides goal-directed medical therapy with high-dose intravenous pulse steroid therapy.

Conclusion:
To our knowledge, this report is not only the first description of targeted treatment options for intractable epistaxis in patients with SLE, but serves to augment the traditional algorithm with the addition of a goal-directed medical therapy control of vasculitis through high-dose intravenous pulse steroid therapy. We demonstrated that six milligrams of intravenous dexamethasone given every six hours can be highly effective in controlling epistaxis in patient with uncontrolled SLE. The presumed mechanism is through control of associated vasculitides.

Case Report

Introduction

Epistaxis is common and generally self-limited, but can quickly become an otolaryngic emergency. The etiology of epistaxis is vast and includes causes from digital manipulation, inflammation and neoplasia to coagulopathies and systemic disease [1]. Intractable epistaxis can become life-threatening, as hypotension and hypoxia can easily occur. Although 60% of people may experience an episode of epistaxis in their lifetime, only 6% of cases will require medical intervention [2].

Conservative treatment options include nasal vasconstrictors, pressure to anterior and/or posterior nasal packing, and electrocautery. Treatment of epistaxis refractory to conservative management includes arterial ligation and/or embolization of the maxillary, sphenoidal, and/or external carotid arteries [2]. Because severe epistaxis can cause devastating sequelae, it is important to identify the etiology and source of bleeding in order to construct a customized treatment plan for each patient.

Intractable epistaxis is defined as bleeding of unidentified origin and bleeding not controlled by at least one trial of standard nasal packing [3]. These bleeds can be even harder to manage in the setting of potential comorbidities, such as those encountered with iatrogenic, idiopathic, acquired, and/or genetic coagulopathies. Specifically, those with chronic inflammatory disorders like systemic lupus erythematosus represent an understudied population at high-risk of intractable epistaxis.

Systemic lupus erythematosus (SLE) is an autoimmune inflammatory disorder, capable of affecting almost any organ in the body. Bleeding is relatively common in patients with lupus, as autoantibodies and immune complexes formed can deplete platelets, initiate peripheral hemolysis of red blood cells and cause potentialing inflammation. Acute lupus vasculitis is classically characterized by an involvement of small vessels with foci consisting of mononuclear cells, perivascular infiltrates and fibrinoid deposits [4]. Acquired coagulopathies are also common as patients can develop antiprothrombin antibodies and lupus anticoagulant autoantibodies.

Despite the significantly elevated bleeding diathesis, cases of severe and intractable mucosal bleeding in patients with SLE are rarely described. Systemic lupus erythematosus (SLE) represents an understudied population at high-risk of intractable epistaxis. There is a paucity of research on treating epistaxis associated with lupus-related vasculitides, and data for treating hemorrhage in SLE is limited only to the management of lupus enteritis. Gastrointestinal literature suggests bleeding caused by lupus enteritis is best treated with high-dose intravenous pulse steroid treatments. Tan et al reports success in obtaining control of an extensive bowel hemorrhage using 500-mg of intravenous methylprednisolone daily for three days [7]. Additionally, Hiraishi et al describes a case of massive gastrointestinal bleeding in a 47-year-old female with lupus enteritis who recovered rapidly after administration of 1500-2000-mg methylprednisolone daily for five days [8]. Both investigators emphasize the efficacy of steroid pulse therapy in rapid resolution of clinical signs and diagnostic studies.

Although surgical intervention is a definitive option, high-dose intravenous pulse steroid responsiveness indicates corticosteroids could become a potential first-line treatment for lupus enteritis [9]. Although no current studies exist that prove a similar result for epistaxis associated with lupus vasculitis, empiric evidence as well as the anatomic and physiologic similarities between the two systems and the associated vascular territories may support the use of corticosteroids. Both disease processes involve systems with a central lumen/cavity lined with mucosal cells, robust vascularity, and contain a strong presence of lymphoid tissue. As a result, gastroenterologic research provides a potential foundation on which to base treatment for acute lupus vasculitis causing epistaxis [10].

Discussion

The most important aspect of managing epistaxis is identification of the bleeding source and control of exacerbating factors, in this case vasculitis secondary to systemic lupus erythematosus (SLE). Classic methods of initially controlling epistaxis are well known and frequently reviewed, but the proactive management of patients with specific blood dyscrasias is rarely discussed. Management therefore tends to be reactive in nature, instituted on a case-by-case basis only. By definition, this strategy condemns the otolaryngologist to always being one step behind in the treatment algorithm.

Early surgical intervention may be especially important in high-risk patients, such as those with coagulopathies or vasculitis, in an attempt to avoid exsanguination and resultant blood transfusions [3, 5, 6]. Despite multiple surgical options for refractory/intractable epistaxis, effective medical treatment options are few.

To our knowledge, this report is not only the first description of targeted treatment options for intractable epistaxis in patients with SLE, but serves to augment traditional treatment strategies with the addition of a goal-directed medical therapy – control of vasculitis through high-dose intravenous pulse steroid therapy. We demonstrated that six milligrams of intravenous dexamethasone given every six hours can be highly effective in controlling epistaxis in patient with uncontrolled SLE. The presumed mechanism is through control of associated vasculitides.

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References