

Atrophic rhinitis

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ABSTRACT

A case of atrophic rhinitis complicated by nasal septum perforation is presented, followed by a discussion of the clinical characteristics, pathophysiology, diagnosis, and management of this disease process. Clinical pearls and pitfalls are emphasized for the use of the practicing allergist and fellow in-training. The diagnosis of atrophic rhinitis is guided by a careful clinical history and evaluation to exclude other possible etiologies for the patient's symptoms and physical findings. Because atrophic rhinitis is a poorly understood process, it is surrounded by considerable contention in the literature and its management is not well defined. Atrophic rhinitis often carries significant morbidity and is best addressed by a multidisciplinary approach.

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Question

A 29-year-old white man presents with a 4-year history of anterior nasal septum perforation and long-standing rhinosinusitis (Fig. 1). He also reports green and black nasal crusts, anosmia, episodic epistaxis, and has been told that his nose exudes a fetid odor. Two-years before presentation, a nasal septum button was placed by otorhinolaryngology with subsequent enlargement of the perforation and increased symptoms. Laboratory evaluation, including antineutrophil cytoplasmic antibodies and rheumatoid factor, was unremarkable. We referred the patient for a biopsy of the septal tissue, which was notable for growth of *Staphylococcus aureus* and an absence of dysplasia or granulomas. The patient denies prior nasal trauma or surgery, cocaine use, occupational exposure to caustic materials, history of sexually transmitted infection, autoimmune disease, or prolonged use of intranasal corticosteroids or of intranasal decongestants. Other than a large perforation of the anterior nasal septum, the patient's physical examination is unremarkable. Which of the following is true?

- A. This patient's condition is more common among male subjects.
- B. This condition indicates autosomal dominant inheritance.
- C. Surgical management is curative.
- D. Atopic processes, such as allergic rhinitis, are commonly associated with this condition.
- E. This condition has only recently been recognized and characterized.

Atrophic rhinitis is a poorly understood, chronic disease process characterized by atrophy of the nasal mucosa, formation of nasal crusts, and a characteristic foul odor.^{1,2} It shows a female predominance (5.6:1; other sources state 6:1.5, female/male) and is endemic to subtropical and temperate climate zones where its incidence ranges from 0.3 to 1% of the population.^{3–5} Diagnosis is based on history and exclusion of other pathological entities; however, standardized diagnostic criteria have recently been proposed.⁶ Although known since ancient times and first characterized in the medical literature by Fraenkel in 1876, the underlying etiology and management of this disease remains a contentious issue in the medical literature.^{2,3} Partly to blame is the use of several terminologies to refer to atrophic rhinitis, including atrophic rhinosinusitis, rhinitis sicca, rhinitis fetida, and ozena.^{7,8} Regardless of the moniker, atrophic rhinitis is a progressive disease in which its management, whether medical or surgical, largely serves to mitigate but not arrest this process.^{2–5,8}

CLINICAL CHARACTERISTICS AND PATHOPHYSIOLOGY

The hallmark features of atrophic rhinitis include progressive atrophy of the nasal mucosa, the development of hardened nasal crusts, anosmia, and the presence of a foul odor or fetor emanating from the patient's nose.^{4,8} It is frequently noted in the medical literature that the patient is often unaware of this latter symptom and is only alerted to its presence by the reluctance of others to

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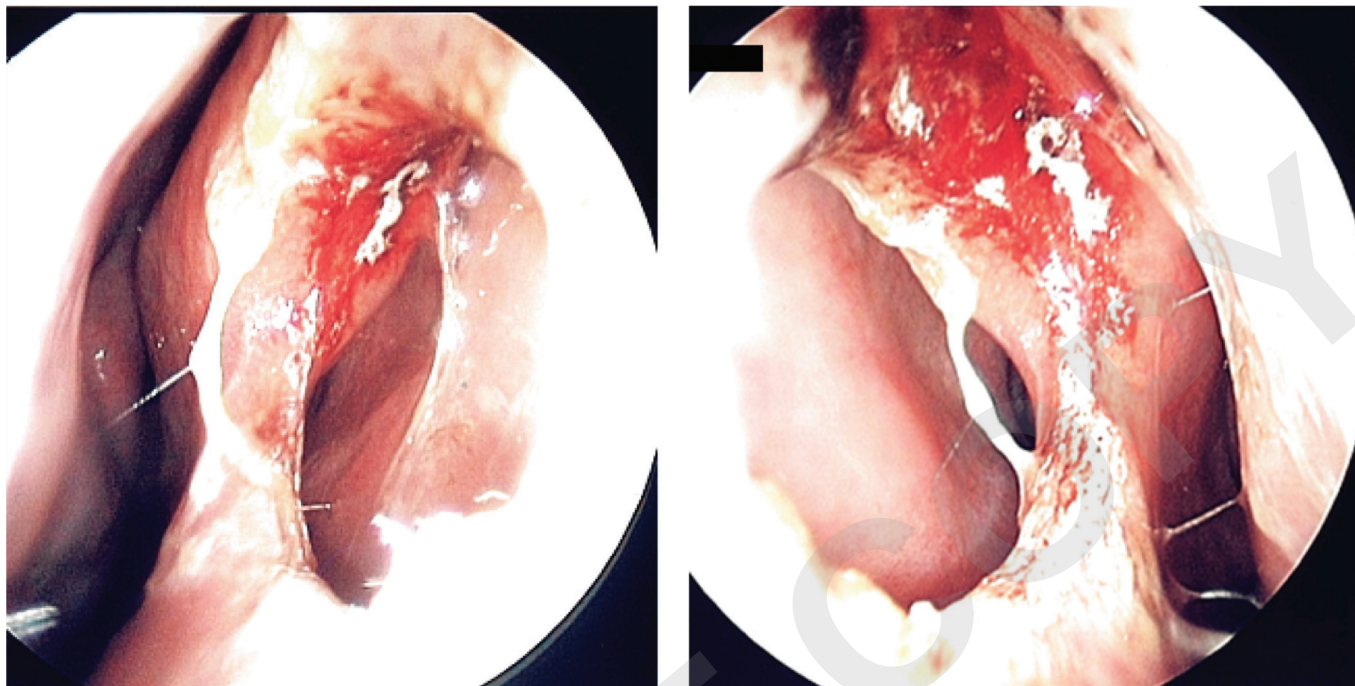


Figure 1. Bilateral view (oriented as if patient is facing the observer) of our patient's significant anterior nasal septum perforation.

remain in close proximity.³ Additional symptoms include a sensation of dryness in the nose, nasal obstruction, episodic epistaxis, and, in some cases, perforation of the nasal septum.^{2,4,6,7} A recent study by Garcia *et al.* proposes that increased mucus evaporation and water fluxes in the noses of atrophic rhinitis patients may lead to these symptoms by disrupting the warming and humidification of inspired air.⁹

Although the underlying etiology driving atrophic rhinitis has not been elucidated, a refinement of the disease process into primary and secondary forms was first made by Ruskin in 1932.¹ Primary atrophic rhinitis is principally evident in Saudi Arabia, China, Philippines, and developing countries in the subtropical and temperate climate zones. Although there is no clear genetic basis for this form of the disease, up to one-half of patients have a family history of atrophic rhinitis.^{3,4} Primary atrophic rhinitis is also closely related to infectious pathogens; however, the nature of the relationship is not clear. Such infections or bacterial colonization may be either a cause or could represent an opportunistic process. Preeminent among these organisms is *Klebsiella ozaenae*, but other organisms have also been noted such as *S. aureus*, *Pseudomonas aeruginosa*, *Proteus mirabilis*, and *Escherichia coli*.^{1,2,10} Finally, it is notable that concurrent allergic rhinitis is diagnosed in as many as 85% of atrophic rhinitis patients.⁴

Secondary atrophic rhinitis is the more prevalent form in the United States and developed countries. It also remains incompletely understood but has been linked with a number of underlying causes. Diseases associated with granulomatous changes are strongly linked with the

secondary form of atrophic rhinitis. Implicated processes include infections such as tuberculosis, leprosy, and syphilis, as well as autoimmune processes such as sarcoidosis and Wegener's granulomatosis.¹ There are also reports of atrophic rhinitis representing a rare presentation of anhidrotic ectodermal dysplasia.⁴ Other causes include nasal or sinus surgeries, often called "empty nose syndrome" in the literature.^{10,11} There is some data suggesting that antiangiogenic drugs may contribute to this secondary form of the disease.¹ Unlike the previously cited autoimmune and infectious processes, cocaine, prolonged use of intranasal corticosteroids and decongestants, and digital trauma are implicated in nasal septum perforation, but are not generally associated with atrophic rhinitis.

DIAGNOSIS

Primary atrophic rhinitis is largely a diagnosis of exclusion focused on evaluation for the causes of secondary atrophic rhinitis. Long bereft of uniformly accepted diagnostic criteria, Ly *et al.* have offered seven signs and symptoms associated with 95% sensitivity and 77% specificity in their study of 22 patients.⁶ Each of the criteria must have been present for at least 6 months and include patient report of chronic nasal obstruction, recurrent epistaxis, and episodic anosmia or documentation by a physician of nasal purulence, nasal crusting, and two or more sinus surgeries. A final element, which more clearly identifies secondary atrophic rhinitis, is the identification of a chronic inflammatory disease associated with granuloma formation.⁶

Although not required for diagnosis, a number of other findings are suggestive of atrophic rhinitis. Drawing from Moore and Kern's (2001) landmark article reviewing 242 atrophic rhinitis patients, a nasal biopsy specimen showing loss of the normal pseudostratified columnar epithelium and atrophy of the mucus glands was seen in 80% of patients. A nasal culture identifying *K. ozaenae* strongly suggests the diagnosis and isolation of other associated organisms is also helpful. Computed tomography showing resorption or destruction of the ethmoid sinuses, mucosal thickening of the paranasal sinuses, or maxillary sinus hypoplasia is also linked with atrophic rhinitis.²

MANAGEMENT

There is no definitive treatment for atrophic rhinitis; therefore, the primary goal of therapy is the reduction of symptoms to improve quality of life. Medical management principally consists of nasal hygiene and the use of antibiotics. The former approach makes use of irrigation with hypertonic saline solutions with some protocols calling for the addition of a corticosteroid to the lavage solution.¹ Antibiotic administration may take the form of lavage solutions containing tetracycline, aminoglycosides, Bactroban, levofloxacin, or ciprofloxacin.^{1,2,10} Along similar lines, oral courses of rifampin, 600 mg daily for 12 weeks, or ciprofloxacin, 500–750 mg twice daily for 8–12 weeks, have been described.^{4,8}

There are a wide variety of surgical approaches to atrophic rhinitis; however, studies have been hampered by small numbers of patients and some sources cite methodological concerns.⁵ A number of techniques either seek to reduce the size of the nasal cavities or allow the nasal mucosa to “rest” through temporary closure of the nostrils. The former strategy has used injections, synthetic implants, and autologous grafts to decrease the size of the nasal cavities.^{4,11} Surgical techniques for closing or narrowing the nostrils have recently been joined by the development of synthetic nasal stents that maintain a restricted, but patent, nasal airway.^{4,12} Treatments may ameliorate but will not cure atrophic rhinitis.

Allergists should seek to become familiar with atrophic rhinitis because of the overlap of symptoms with other forms of chronic rhinosinusitis as well as the common presence of allergic rhinitis in patients. The progressive nature, poorly understood etiologic mechanisms, and several contributing or concurrent disease processes related to atrophic rhinitis render a multidisciplinary approach to management essential.

Answers to the Question

- A. False—Atrophic rhinitis is female predominant with a female/male of 5.6:1.

- B. False—Although a family history is often reported, there is no clear inheritance pattern.
C. False—There is no cure, only a means to ameliorate the symptoms.
D. True—Up to 85% of atrophic rhinitis patients have allergic rhinitis.
E. False—Reports of atrophic rhinitis extend to the Edwin papyrus (1700 BC).

Pearls

- Atrophic rhinitis should be suspected in the presence of atrophy of the nasal mucosa, nasal crusts, anosmia, and nasal fetor.
- Primary atrophic rhinitis is a diagnosis of exclusion, particularly of processes associated with granuloma formation.
- Atrophic rhinitis requires a multidisciplinary management team.

Pitfalls

- Primary atrophic rhinitis must be differentiated from secondary forms, because it is important to treat any underlying disease process.
- The common presence of atopy, specifically of allergic rhinitis, should prompt involvement of the allergist with atrophic rhinitis patients.

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