

Sinonasal Tumor Presenting as Chronic Nasal Congestion

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A 56-year-old Hispanic man with a history of hypercholesterolemia was referred to the Allergy Clinic with complaints of worsening nasal congestion for nine months. The patient had seen his primary care physician who prescribed several medications, including antihistamines, intranasal steroids, and decongestants, which did not provide relief. For the previous three months the patient had complained of blurry vision, and his family had noted that his left eye seemed to be red and protuberant at times. One month prior to his clinic visit, he had begun to experience left-sided facial pain. The patient admitted to a ten pound weight loss, but denied fever, chills, night sweats, epistaxis, cough, breathing difficulties, rashes, or joint complaints. His review of symptoms was otherwise negative.

His only medications included intranasal fluticasone two sprays in each nostril daily and montelukast 10 mg orally daily for chronic rhinitis symptoms. He denied any drug or food allergies, and there was no family history of atopy. His home had central air conditioning and heating. He did not have any pets. He worked as a janitor in a chicken food plant and denied use of tobacco, alcohol, or drugs.

On physical examination, the patient was a well-developed, well-nourished man with normal vital signs. He had left eye proptosis with mild chemosis and left lower lid edema (Figure 1). Pupils were equal, round, and reactive to light, and extraocular movements were intact. Tenderness to palpation was present over the left maxilla. The left nasal passage was barely patent with brown mucus present on anterior rhinoscopic examination. The nasal mucosa was pale and edematous. Left posterior cervical chain and posterior auricular lymph nodes were palpable, albeit small, doughy, and non-tender. Chest auscultation revealed clear lung fields. Cardiac, abdominal, and skin examination revealed no abnormalities. Rhinolaryngoscopy demonstrated very edematous nasal mucosa bilaterally. The left nasal passage was very narrow with edematous middle and inferior turbinates. The opening to the maxillary sinus could not be visualized on the left. The vocal cords appeared normal.

Because the patient was referred to the Allergy Clinic with a possible diagnosis of allergic rhinitis, he underwent skin testing to inhalants. Skin testing was negative with a positive histamine control. A complete blood count (CBC) revealed a white blood cell count of 5,200/mL

TARGET AUDIENCE

The September/October Clinical Case of the Month is intended for primary care physicians, general internists, otorhinolaryngologists, surgeons, radiologists, and oncologists.

EDUCATIONAL OBJECTIVES

After reading the article, the healthcare provider should be able to discuss the differential diagnosis and management of chronic nasal congestion.

CREDIT

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CME INFORMATION

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DISCLOSURE

Dr. Kewalramani has nothing to disclose.

Dr. Aresery has nothing to disclose.

Dr. Hedberg has nothing to disclose.

Dr. Wild has nothing to disclose.

Dr. Lopez discloses that he is a member of the *Journal* Board of Trustees and the *Journal* Editorial Board.

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Figure 1. The patient presented with proptosis of the left eye.

(3,800-10,800/mL) with 66% neutrophils, 26% lymphocytes, 6% monocytes and 2% eosinophils. His hemoglobin was 15 g/dL (13.2-17.1 g/dL) with a hematocrit of 43% (38.5-50%). His platelet count was 157,000/mL (140,000-400,000/mL).

Because of the proptosis of this left eye, the patient was sent for a computed tomogram (CT) of the sinuses and orbits to look for sinusitis, anatomical abnormalities, nasal polyps, or a tumor. His CT showed a 5-cm-in-diameter soft tissue mass in the left nasal cavity. The mass eroded into the right nasal cavity, left maxillary sinus, left orbit, both ethmoid sinuses, and the cribriform plate. It displaced the inferior and medial rectus muscles and filled the entire left and part of the right frontal sinuses (Figure 2). The patient underwent an endoscopically-guided biopsy, which revealed a large fleshy, polypoid mass. The histology was consistent with adenoid cystic carcinoma. He subsequently underwent tumor debulking via CT-guided endoscopic sinus surgery. All visible tumor mass was removed from the nose and left maxillary sinus. The tumor was adjacent to the optic nerve, which was decompressed. The patient then completed a 6-month course of chemotherapy and radiation. He currently is doing well without nasal or visual complaints.

DISCUSSION

The differential diagnosis of chronic nasal congestion is lengthy and broad and includes allergic rhinitis, vasomotor rhinitis, chronic sinusitis, anatomic abnormalities, a foreign body, nasal polyps, autoimmune or granulomatous diseases, and benign and malignant tumors.

Allergic rhinitis is the most common form of rhinitis and affects 20 to 40 million people in the United States, including 10 to 30% of adults.¹ Continuous exposure to allergens, such as dust mites, animal dander, pollen, and molds, can lead to the presentation of the allergen by antigen presenting cells to CD4+ T lymphocytes which secrete cytokines that promote allergen-specific IgE production. Upon inhalation, allergens are deposited in the nasal mucus, with subsequent diffusion into nasal tissues.² Within minutes of contacting an allergen, IgE-sensitized mast cells degranulate and discharge preformed and newly synthesized mediators. Many of these mediators, such as leukotrienes, prostaglandins, hista-



Figure 2. Computed tomogram of the sinuses demonstrates a large sinonasal tumor originating in the left nasal cavity.

mine, and cytokines lead to the typical early-phase symptoms of sneezing, pruritus, rhinorrhea, and congestion.³ In the late phase of the allergic rhinitis response, congestion becomes the predominant symptom. Mediators cause dilation of arteriole-venule anastomoses with consequent edema, pooling of blood in the cavernous sinusoids, and occlusion of nasal passages. As a result of cytokine or mediator release in the early phase, the nasal mucosa becomes infiltrated with inflammatory cells, including basophils, eosinophils, neutrophils, mast cells, and mononuclear cells. Over the next 4 to 8 hours, these cells become activated and release their pro-inflammatory mediators to sustain the nasal mucosal inflammatory response.¹

Vasomotor rhinitis occurs in 5 to 10% of patients, and usually refers to an idiopathic, perennial nonallergic rhinitis.⁴ Patients tend to have negative allergen skin tests, normal serum IgE levels, and no apparent inflammation on nasal cytology. Nasal blockage and obstruction are frequent symptoms, although many report rhinorrhea. Little is known regarding the pathophysiology of vasomotor rhinitis. Although increased neural efferent flow to the blood vessels supplying the nasal mucosa was once believed to play a role, there is a lack of evidence to support this.² Symptoms may result from nonimmunologic stimuli, such as changes in temperature or relative humidity, alcohol ingestion, strong odors, and other airborne irritants.⁵

Chronic rhinosinusitis includes continuous symptoms for 12 or more weeks. Symptoms include nasal obstruction, purulent nasal discharge, postnasal drainage, hyposmia or anosmia, headache, halitosis, fatigue, dental pain, cough, and ear pain.⁶ Chronic rhinosinusitis can be infectious or hyperplastic. Chronic infectious

rhinosinusitis is usually associated with bacteria, including anaerobes such as *Fusobacterium* and *Prevotella*, *Staphylococcus aureus*, and Gram-negative rods such as *Pseudomonas aeruginosa*. Neutrophils are frequently seen. Chronic hyperplastic eosinophilic rhinosinusitis is accompanied by an influx of eosinophils and mononuclear cells. It is often seen in conjunction with aspirin intolerance, asthma, and nasal polyps. Fungi may also play a role in chronic rhinosinusitis. Patients with allergic fungal sinusitis (AFS) usually present with longstanding nasal congestion and/or obstruction with or without nasal polyps. Inhalation of fungal spores may lead to an immunologically mediated hypersensitivity reaction.⁴ There may be encroachment on surrounding structures, such as the orbit or central nervous system, leading to visual loss or complete nasal obstruction.^{4,7} Patients with AFS usually demonstrate an allergy to the fungus colonizing the allergic mucin present in their sinuses.⁷ Fungi present in the sinuses may also result in a fungus ball or invasive fungal sinusitis. A fungus ball usually occurs in the maxillary or sphenoid sinus and is generally unilateral. Chronic nasal obstruction and headaches may be seen. Invasive fungal sinusitis is typically seen in immunocompromised individuals such as people with diabetes, those with malignancies, those on chronic high-dose steroids, and transplant recipients. These individuals present with fever, headaches, epistaxis, and possibly mental status changes.⁴

Rhinitis symptoms are seen with systemic autoimmune diseases such as systemic lupus erythematosus, relapsing polychondritis, and Sjögren's syndrome. Granulomatous diseases such as sarcoidosis or Wegener's granulomatosis can also produce nasal manifestations in many patients.⁵ It is not unusual for systemic symptoms to be absent or undiagnosed when individuals first present with nasal symptoms. In Wegener's granulomatosis the nose and sinuses are the second most common sites for presenting symptoms in 67% of patients. Ninety-one percent of patients eventually will have nasal and sinus involvement. Symptoms frequently include nasal obstruction associated with clear rhinorrhea or purulent drainage. This may progress to foul-smelling crusting with friable ischemic tissues, and then two nasal septal perforation, and eventually two saddle-nose deformities secondary to cartilaginous and bony loss. Relapsing polychondritis involves recurrent episodes of inflammation of cartilage of the auricles, larynx, trachea, and nose, as well as many other sites throughout the body. Nasal involvement is secondary to inflammation and fibrotic changes of the nasal cartilage. Recurrent nasal pain and erythema are frequent. Cartilage is soon replaced by fibrous connective tissue with supporting structures losing their integrity, which leads to painless nasal collapse and a saddle-nose deformity. Nasal mucosal involvement in sarcoidosis is common and may be the only presenting sign. Patients may present

with no symptoms or varying degrees of nasal occlusion, including rhinosinusitis with nasal obstruction and hyposmia. To make the diagnosis of systemic lupus erythematosus, at least four of eleven criteria must be met. One criterion is the presence of nasopharyngeal ulcerations. Nasal mucosal ulcerations and anterior septal perforations have been reported and are secondary to ischemia. The mucous membrane lesions flare with other systemic manifestations. Xerorhinia, or dry nose, may be seen in conjunction with xerostomia and xerophthalmia in those with Sjögren's syndrome. Up to 100% of patients report xerorhinia. Decreased nasal secretions lead to impaired olfaction and nasal crusting, which then causes nasal obstruction.⁸

Anatomic abnormalities account for 5 to 10% of chronic nasal disorders. In adults, anatomical causes include nasal septal deviation and turbinate hypertrophy. Patients usually present with obstructive symptoms rather than rhinorrhea. Nasal septal deviation is common, but usually well tolerated when mild. Moderate-to-severe septal deviation associated with nasal mucosal edema secondary to underlying rhinitis can result in symptoms of blockage.⁵

Although nasal polyps occur in only 1% of the general population, they should always be considered in the differential diagnosis of constant nasal congestion and obstruction.⁵ They are benign inflammatory outgrowths arising from inflamed paranasal sinus mucosa. Common symptoms are nasal congestion, rhinorrhea, and a decreased sense of smell.² Nasal polyps can often (though not always) be visualized with anterior rhinoscopy. They are usually bilateral, multiple, freely movable, and pale gray.⁵ Eosinophils are present in moderate-to-large numbers in the polyps of 77% of individuals. Interestingly, neutrophils tend to be the primary cells noted in polyps associated with cystic fibrosis. Although complaints of rhinorrhea, sneezing, and itching, elevated histamine and IgE levels in polyp fluid, degranulated mast cells, and marked tissue eosinophilia may lead one to believe that the formation of nasal polyps is related to allergic rhinitis, fewer than 5% of allergic rhinitis patients have nasal polyps.^{2,9} They are more frequently associated with aspirin intolerance, non-allergic asthma, chronic sinusitis, cystic fibrosis, Churg-Strauss syndrome, and allergic fungal sinusitis.⁵

Foreign bodies in the nasal cavity can lead to unilateral foul smelling, mucopurulent discharge and nasal obstruction which may lead to a usually painless sinusitis.² Nasal foreign bodies can be found in any portion of the nasal cavity, but are usually found around the floor of the nose immediately below the inferior turbinate or just anterior to the middle turbinate. However, these objects are usually inserted by young children or institutionalized patients. Some foreign bodies may remain in the nose for years without mucosal changes; however, most objects initiate congestion and swelling of the na-

sal mucosa, with the possibility of pressure necrosis accompanied by ulceration, erosion, and epistaxis. Retained secretions, the decomposed foreign body, and ulcerations will lead to the foul odor.¹⁰

Although neoplasms are uncommon, one must consider the possibility of a benign or malignant tumor in the nasal passage, paranasal sinuses, or nasopharynx when a patient presents with refractory nasal congestion, cranial nerve abnormalities, and/or orbital involvement.¹¹ Lesions may occlude the nasal airway, and are frequently unilateral.¹ Depending on the histology, the origin of the tumor, and extent of growth, bleeding, hyposmia or anosmia, pain, and otalgia may be seen.¹² Invasion can result in diplopia, pain, Horner's syndrome, and proptosis.¹³

Sinonasal tumors, which represent tumors of the nasal cavity and paranasal sinuses, are uncommon and make up 3% of all head and neck tumors. Approximately 50% of these tumors are benign.¹⁴ The major risk factors for sinonasal malignancies include exposure to dusts and chemicals, including wood or leather dusts, nickel, chromate, formaldehyde, and chlorophenol.¹ Smoking has been implicated, but studies have not confirmed this assertion.¹⁴

Presenting signs and symptoms of sinonasal tumors are nonspecific and consist of nasal obstruction, pain, epistaxis, swelling, and mass lesions. Many patients will report months-to-years of these symptoms before the diagnosis is considered, and on average a six-month delay in diagnosis is reported.¹⁴ Seventy-five percent of malignant paranasal and nasal sinus neoplasms have tumor extension beyond the sinuses. Some malignancies may present with signs suggestive of perineural or orbital invasion. Orbital invasion is seen in up to 45% of cases and patients may present with proptosis, globe displacement, decreased extraocular movements, conjunctival chemosis, or a palpable mass within the orbit.¹³ Certain malignancies, such as adenoid cystic carcinoma, invade the nerves and may cause facial or eye pain or numbness, diplopia, Horner's syndrome, or decreased visual acuity.^{13,14}

A CT of the sinuses will demonstrate the mass with anatomic detail and will document encroachment on and erosion of any bony structures.¹¹ Magnetic resonance imaging may distinguish tumor from inflammatory soft tissue and is useful in defining intracranial or intraorbital extension and perineural spread.¹⁴ However, even with an adequate history, physical examination, and imaging, tumor tissue biopsy is the essential component of the diagnostic strategy.¹¹

Because the incidence of sinonasal tumors is low, their sites of origin varied, and their histologies multi-form, no large, multicenter, prospective, randomized trials of treatment have been performed. Current treatment recommendations are based on earlier, single institution, retrospective reviews and studies performed

in other head and neck primary sites. Surgery is usually indicated in the diagnosis and treatment of these malignancies, but chemotherapy and radiation are beginning to play more important roles in the treatment regimen.¹⁴

CONCLUSION

This case illustrates the importance of having a high index of suspicion when evaluating patients with refractory nasal congestion. Patients will frequently be referred to the primary care physician for these complaints. One must perform a thorough history and physical examination. Nasal congestion and obstruction unresponsive to medications and accompanied by orbital and cranial nerve complaints and findings should alert one to a more serious diagnosis, such as malignancy. Unfortunately, there is usually a delay in the diagnosis. Diagnosis depends on imaging and tumor tissue biopsy. Surgery is the mainstay of treatment, however, adjunctive chemotherapy and radiation may also play a role.

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For each question, choose the one answer that is most correct.

1. True/False: Vasomotor rhinitis is the most common form of rhinitis and affects 20 to 40 million people in the United States.
2. Which of the following cells would most likely be found in nasal polyps in a child with cystic fibrosis?
 - a. Eosinophils
 - b. Mast cells
 - c. Basophils
 - d. Neutrophils
3. True/False: Nasal polyps are most frequently seen in patients with allergic rhinitis.
4. Patients with a tumor in the nasal passage, paranasal sinuses, or nasopharynx may present with which of the following complaints?
 - a. Refractory nasal congestion
 - b. Diplopia
 - c. Anosmia
 - d. Proptosis
 - e. Patients may present with any of the above complaints.

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