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Adenoid Cystic Carcinoma: A Rare Cause of Epistaxis

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Abstract
Epistaxis is a common complaint and is most often a benign self-limited condition. Therefore primary care providers might deem it unnecessary to evaluate a patient with epistaxis closely if they are not alert to more ominous etiologies. In this case report of an elderly patient with epistaxis, we illustrate this point by describing the presentation of adenoid cystic carcinoma of the sinonasal tract. We further elucidate the natural history of this malignancy as well as the recommended methods of its diagnosis and treatment.

Introduction
Epistaxis, which is thought to occur in about 60% of the general population, is a complaint often encountered in primary care. The causes of epistaxis are often commonplace and benign, among them antiplatelet or anticoagulant use and nasal mucosa irritation due to excessive drying, such as with nasal supplemental oxygen, manipulation or nasal steroid use. However, malignancies of the sinonasal tract also present with epistaxis. It is imperative that the primary care provider keeps this in mind when evaluating a patient with epistaxis.

Case Presentation
At a routine 3-month home visit, an 81-year-old homebound female with moderate mental retardation cared for by her sister was noted to have had persistently recurrent epistaxis. The nosebleeds, from her left nares, had begun several months previously, just after the last visit. She did not have any constitutional symptoms or a sore throat, runny nose, cough, headache or facial pain. She had been known to snore since childhood. She had not been using any antiplatelet or anticoagulant drugs nor did she have any prior history of bleeding problems. Her supplemental oxygen, used at night for many years and delivered via nasal cannula was humidified. Despite the recent additional use of petroleum jelly in the nares, the nosebleeds continued. She had already discontinued her nasal steroid use, prescribed for long-standing allergic rhinitis. Her platelet count was only slightly reduced, a chronic finding. Her coagulation studies were within normal limits. Her blood pressure was well controlled. She had no sinus tenderness, facial asymmetry or other cranial nerve deficits. The rest of her ear nose and throat (ENT) exam was unremarkable except for occlusion of her left nares by a friable mass. Palpation of her neck did not reveal any masses or lymphadenopathy.

She was referred to an otolaryngologist/head and neck surgeon who confirmed the presence of an obstructive mass in the left nares upon nasal endoscopy (Figure 1). He also confirmed the absence of any signs of metastatic disease. Magnetic resonance imaging (MRI) of the brain and sinuses was unsuccessful due to motion artifact, so she underwent computed tomography (CT) scanning, which revealed a large, obstructive mass of the left nasal cavity extending to the maxillary and ethmoid sinuses (Figure 2). Endoscopic biopsy revealed adenoid cystic carcinoma (ACC).

The patient was treated with endoscopic resection followed by postoperative radiation therapy. She recovered fully and is without any evidence of recurrence almost two years after resection. (Figure 3)

Discussion
Presentation
Malignancies of the sinonasal tract are, as a whole, extremely rare, having an incidence of 1:100,000 per year. ACC comprises 10-18% of these malignancies, second in frequency only to squamous cell carcinoma. ACC derives its name from its origin in glandular tissue (hence the term “adenoid”) and from the many fluid-filled vacuoles seen in its typical histological pattern. It predominantly involves the salivary glands but can also occur in the mucoserous glands underlying the sinonasal epithelium, thereby causing epistaxis.

Figure 1. Endoscopic view of patient’s left nares prior to surgery. Courtesy of Ross Germani, M.D.
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As squamous cell carcinoma, which may also present with epistaxis. Biopsy is needed to confirm the diagnosis. Also, in view of ACC’s frequently late presentation and its predilection for spread to the orbit and brain via perineural spread, it is important to perform MRI imaging of the brain and cranial nerves, especially the trigeminal nerve, prior to treatment.

Therapy
The currently accepted treatment for ACC involves radical surgical resection followed by radiation therapy. Even with these treatments, the prognosis is fairly poor. Overall recurrence rates of about 60%, have been cited. A particularly large 2013 systematic review reported rates of local recurrence (most often to the base of skull) and metastatic recurrence (commonly to the lung) of 34.1% and 30.7% respectively, over an average follow-up period of 51 months. Overall 5-year survival rates are approximately 65%, declining to only 37% at 10 years.

However, because ACC’s are slow growing, it is appropriate to treat the cancer aggressively even in an elderly patient with only local disease, such as ours. Such a patient will likely die of another illness before the ACC recurs. Nevertheless, lifelong surveillance is necessary for all survivors even if surgical margins were clear and no recurrence is detected for many years.

Conclusion
This patient reported a very common complaint with a usually benign etiology. In our patient, however, the common complaint had a rare and malignant cause. Her case illustrates the quintessential challenge that the primary care physician faces: to identify more dangerous, rarer conditions from amongst a vast number of more benign ones. This task is rendered all the more difficult (and crucial) when there are commonplace, benign explanations for the problem, such as nasal oxygen or nasal steroid use (as in her case) or when there are no indications of a more ominous cause (as might have been if no nasal mass had been visible on direct inspection). When evaluating a patient for epistaxis, particularly recurrent unilateral epistaxis, it is important to consider ACC or other malignancies as the cause.

References

Diagnostic Methods
Along with direct examination, CT imaging is often initially used to diagnose ACC. However, this technique is unable to distinguish ACC from other more common head, and neck malignancies, such as squamous cell carcinoma, which may also present with epistaxis. Biopsy is needed to confirm the diagnosis. Also, in view of ACC’s frequently late presentation and its predilection for spread to the orbit and brain via perineural spread, it is important to perform MRI imaging of the brain and cranial nerves, especially the trigeminal nerve, prior to treatment.

Additionally, sinonasal tract adenoid cystic carcinomas (STACCs) may present with nasal obstruction, maxillary tenderness, headache, or hoarseness. Because this cancer has a tendency to spread along neural sheaths, it may also be associated with cranial nerve signs, such as facial numbness, dysphagia, hearing changes, tinnitus or diplopia.

While adenoid cystic carcinoma may appear at any age, it is most common in patients between the ages of 50 and 70, and it is observed equally in both sexes. ACC tends to grow insidiously. Therefore, it is often advanced on presentation, in almost 75% of patients in one 2002 review of ACC of the nasopharynx.