Low-grade adenocarcinoma of the nasal cavity—an unusual presentation: Case report and review of the literature

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Abstract
Low-grade nasal cavity adenocarcinoma is an unusual neoplasm. When it does occur, it often appears in conjunction with a history of epistaxis and nasal obstruction that spans a period of a few weeks to several months. Most of these tumors arise in patients who are middle-aged or older. We report a case of low-grade nasal cavity adenocarcinoma that was unusual in that it occurred in an adolescent boy and that the presenting symptoms of chronic nasal obstruction and recurrent epistaxis had persisted for 7 years. The occurrence of this uncommon but recognized entity in such a young patient widens the age range of possible patients with this tumor, and a finding of such an extended duration of symptoms should raise clinical suspicion and encourage a thorough investigation in order to make the diagnosis.

Introduction
Sinonasal malignancies are uncommon but well-described entities. Sinonasal glandular neoplasms such as adenocarcinomas occur infrequently, but when they do, they tend to arise in patients who are middle-aged and older.1 We describe a case of low-grade nasal cavity adenocarcinoma that occurred in an unusually young patient who had a long history of symptoms.

Case report
A 16-year-old boy was referred to our otolaryngology clinic with a 7-year history of recurrent left-sided epistaxis. The patient said he had experienced semimonthly to weekly episodes of minor epistaxis for 6.5 years, but during the preceding 6 months, the frequency of the episodes had increased to 3 or 4 per week and the amount of blood lost per episode had increased to 2 to 3 teaspoons. For this reason and because it had become more difficult to control his epistaxis with manual pressure, the patient sought evaluation by his primary care physician, who subsequently referred him to our clinic.

The patient said that he had a history of chronic left nasal obstruction “for as long as I can remember.” His medical and surgical histories were unremarkable, and he was not taking any herbal supplements or over-the-counter or prescription medications. He denied tobacco and alcohol use and any known prolonged exposure to carcinogens such as nickel, leather, formaldehyde, mineral oils, chromium, isopropyl oils, lacquer paint, soldering and welding materials, and radium paint. Notably, he reported a 1.5-year history of exposure to wood dust during classes in high school wood shop.

On examination, a mild right nasal septal deviation was noted. Further examination revealed a large, tan, firm, nonfriable, lobulated mass in the left superior nasal cavity. The mass did not appear to be hypervascular. Its origin could not be ascertained on endoscopic examination because of its large size and its location. Findings on examination of the nasopharynx, eyes, neck, and cranial nerves were normal. In order to better assess the extent and origin of the mass, noncontrast computed tomography (CT) of the paranasal sinuses was obtained. CT demonstrated a left superior nasal cavity mass without intracranial extension or bony destruction (figure 1).

The next day, the patient was taken to the operating room for a thorough nasal endoscopic examination and biopsy under general anesthesia. Intraoperatively, the lesion was found to originate at the junction of the middle turbinate root and anterior skull base (figure 2). Findings on frozen-section analysis of the biopsy were consistent with a glandular tumor (figure 3). No evidence of malignancy was
noted. The remainder of the mass was then removed with a microdebrider. After permanent sectioning, the histopathologic diagnosis remained ambiguous. The specimen was therefore sent to the Armed Forces Institute of Pathology for expert consultation, and it was ultimately diagnosed as a low-grade adenocarcinoma of the nasal cavity.

Only the visible portion of the tumor had been removed during biopsy, so the patient was taken back to the operating room for re-excision of potential microscopic tumor. The tissue removed included the root of the superior middle turbinate and the anterior skull base mucosa adjacent to the superior-most anterior ethmoid cell next to the fovea ethmoidalis. The root of the middle turbinate was found to contain a small focus of residual disease on permanent histopathologic examination. Therefore, the patient was again returned to the operating room for completion middle turbinectomy. This specimen was free of residual adenocarcinoma.

The patient subsequently underwent removal of the remaining portion of the superior turbinate, removal of the adjacent superior septum and superior meatal mucosa, and partial anterior ethmoidectomy. All specimens were negative for malignancy.

Following identification and removal of the residual adenocarcinoma, the patient adhered to follow-up recommendations. Ten months after his most recent surgical intervention, positron-emission tomography (PET) was performed to look for recurrent malignancy, and the findings were inconclusive. At follow-up 20 months after the most recent surgery, the patient exhibited no clinical evidence of recurrent adenocarcinoma.

Discussion

Glandular neoplasms account for 4 to 8% of all primary nasal cavity malignancies.

Low-grade adenocarcinomas of the nasal cavity represent an uncommon but recognized category of glandular neoplasms. Clinically, the appearance of nasal cavity adenocarcinomas is similar to that of benign entities such as nasal polyps. Low-grade nasal adenocarcinomas pose a diagnostic challenge for the pathologist because they can be confused with benign adenomas. Nasal adenocarcinomas occur equally in men and women, and there is no racial predilection. They tend to arise in patients between the ages of 40 and 80 years.

Because these neoplasms are uncommon in younger patients, otolaryngologists should have a high index of suspicion to investigate for this possible diagnosis. Patients with adenocarcinoma limited to the nasal cavity usually present with nasal obstruction, and many present with recurrent or chronic epistaxis. The duration of symptoms varies from a few weeks to several months. Prior to

![Figure 1. Preoperative CT shows the soft-tissue density in the left superior nasal vault without intracranial extension or bony destruction.](image1)

![Figure 2. Intraoperative photograph shows the lobulated mass between the septum (left) and lateral nasal wall (right).](image2)
the case we report here, the longest duration of symptoms prior to adenocarcinoma diagnosis had been 5 years.³

A specific risk factor for primary nasal cavity adenocarcinoma is exposure to wood dust. Furniture workers have a 70-fold increased risk.³ Although our patient’s exposure to wood dust through his shop class may have played a role in the development of his adenocarcinoma, the reasons that some patients go on to develop this cancer while most do not is unknown and points toward a likely multifactorial etiology. The workup of an intranasal mass is guided by the clinical history and physical examination findings. Diagnostic imaging may better define the mass and any local extension into adjacent structures. Biopsy of the mass can be both diagnostic and therapeutic. Fine-needle aspiration biopsy, incisional biopsy, and excisional biopsy may all be performed in the clinic setting.

The decision whether to perform office-based biopsy or biopsy in the operating room depends on patient and physician preference, clinical findings, and potential risks. For example, if a mass is hypervascular and juvenile angiofibroma is high on the list of differential diagnoses, it is advisable to avoid biopsy in the clinic setting, where control of potential hemorrhaging may be difficult. The operating room is also preferable when imaging or clinical examination indicates possible intracranial extension. Other advantages of biopsy in the operating room include the ability to deliver definitive treatment at the time of excisional biopsy when appropriate, and better patient comfort. The advantages of office biopsy include the possible avoidance of general anesthesia, lower costs, and patient convenience. Additionally, making a diagnosis before a patient undergoes general anesthesia allows for more focused preoperative

Figure 3. Histopathology shows a uniform glandular architecture with a crowded appearance (H&E, original magnification ×400).

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Diagnosing adenocarcinoma is clinically challenging, and the diagnosis ultimately rests on histologic evaluation. Low-grade sinonasal adenocarcinomas are grossly papillary or cauliflower-like in appearance, and they are usually soft (although they can be gritty when significant psammomatous calcifications are present). They range in size from 0.3 to 4.0 cm.1 Microscopically, they have papillary and glandular growth patterns, they are unencapsulated, and they tend to infiltrate surrounding tissues. Histologic diagnostic clues include uniform glandular architecture with a crowded appearance and a back-to-back and cribiform pattern.1 Papillae show arborization and hyalinized fibrovascular cores. The cells lining the structures can vary from columnar and pseudostratified to focally cuboidal with eosinophilic cytoplasm. Although mitotic figures are infrequent and nucleoli are inconspicuous, psammoma bodies may be present.2 The cells exhibit a degree of morphologic and cytologic uniformity and round to oval nuclei with vesicular chromatin, and they may be confused with cells seen in adenomas and papillomas. Mild nuclear pleomorphism and a loss of basal polarity in low-grade adenocarcinoma cells help distinguish them from cells found in adenomas and papillomas.

Glandular neoplasms of the sinonasal tract are separated into salivary-type and surface tumors. Low-grade sinonasal adenocarcinoma is believed to originate from the surface epithelium. In the largest series of low-grade sinonasal carcinoma published to date (N = 23), Heffner et al cited the lack of immunohistochemical staining for actin, S-100 protein, and glial fibrillary acid protein and the contiguity with the surface epithelium as evidence of surface origin.1

Despite staining differences with demonstration of myoepithelial cells,2 a surface origin is still favored.

For treatment planning and prognosis, it is important to distinguish low-grade adenocarcinomas from high-grade adenocarcinomas. Histologic findings suggestive of high-grade adenocarcinomas include solid growth patterns with sheets of cells; poorly defined, irregular glandular patterns; hyperchromatism; moderate to prominent nuclear pleomorphism; and a high mitotic rate.3

The diagnostic evaluation of a sinonasal mass may include CT or magnetic resonance imaging (MRI). Either radiologic technique may demonstrate tumor involvement of the nasopharynx, intracranial cavity, paranasal sinuses, orbits, infratemporal fossa, and pterygopalatine fossa. The tumor origin may be difficult to determine on CT if a common wall, such as the medial wall of the maxillary sinus, is involved. This is especially true if an associated obstructive sinusitis is present.3 In such cases, MRI may be a useful adjunct.

In the study by Heffner et al, which was published in 1982, the mortality rate following multiple recurrences was 9%.1 At that time, however, PET scanning and multiplanar reformatted CT imaging (i.e., sagittal and coronal reconstructions) were not available. Perhaps the availability of these imaging methods and techniques will improve the detection of recurrence and subsequently reduce mortality rates. The effectiveness of PET in detecting low-grade malignancy has yet to be reported.

The overall recurrence rate for paranasal sinus adenocarcinoma is 50%; distant metastases are uncommon (9%).1 The prognosis for low-grade adenocarcinoma is very good, although specific recurrence and metastasis rates are not well established. In contrast, patients with high-grade lesions have a poor prognosis.

The primary treatment of sinonasal adenocarcinoma is complete surgical excision. In a review of 13 patients with sinonasal adenocarcinoma, Alessi et al reported that the single most important factor in treatment was adequacy of the surgical margins, although they did not recommend a specific margin size.3 In areas such as the superior nasal vault and skull base, multiple excisional biopsies may be required to obtain clear margins because the surrounding mucosa may appear grossly normal but harbor microscopic disease. Surgery for low-grade adenocarcinoma should be less radical than more aggressive approaches warranted for high-grade lesions.1,5 Adjunctive radiation has been suggested for high-grade lesions, as well as for recurrent low-grade lesions.7 Although radiation therapy in the treatment of low-grade nasal cavity adenocarcinoma has not been well studied, it may play a role when complete surgical excision is not achieved.1

Recurrent epistaxis and nasal obstruction may be the presenting symptoms in an adolescent patient with a sinonasal adenocarcinoma. Our case highlights the importance of maintaining a high clinical index of suspicion for a sinonasal malignancy when a history of epistaxis, even if present for many years, occurs in a young patient. Complete surgical excision and routine clinical surveillance are essential to minimize tumor recurrence and thus decrease potential mortality from the disease.

References