Case Report

Low-grade sinonasal adenocarcinoma: Report of a rare entity

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ABSTRACT

Sinonasal adenocarcinomas (SNACs) are rare tumors which include two broad categories – the salivary and the nonsalivary type. Nonsalivary type adenocarcinomas arising within the sinonasal tract are composed of intestinal type and nonintestinal type of adenocarcinoma each of which can be either low grade or high grade. The intestinal type is clinically aggressive and generally presents at an advanced stage. Nonintestinal type adenocarcinomas are rare tumors; the majority of these are of a histological low grade and show excellent prognosis. We report a case of this rare entity in a 53-year-old male and discuss the pathologic features of the low-grade SNAC.

Key words: Adenocarcinoma, low grade, sinonasal

INTRODUCTION

Sinonasal adenocarcinomas (SNACs) are rare tumors that account for 0.4% of all human neoplasms[1] and 10–20% of all primary neoplasms of the nasal cavity and paranasal sinuses.[2] They are currently classified by the WHO into the salivary and the nonsalivary type.[3] The salivary type adenocarcinomas are identical to their salivary gland counterparts and constitute the majority of these neoplasms. The nonsalivary type adenocarcinomas arising within the sinonasal tract are uncommon tumors that are often poorly recognized and misdiagnosed.[4] They are classified as intestinal type and nonintestinal type adenocarcinomas and each of these can be either low grade or high grade. The intestinal type SNACs are clinically aggressive and generally present at an advanced stage with an overall mortality of 53%.[4,5] The nonintestinal type adenocarcinomas are rare tumors and the majority of these are of histological low grade and show an excellent prognosis.

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CASE REPORT

A 53-year-old male presented with bleeding from the nose for 1 month. On examination, there was a nasal mass on the right side arising from the nasal septum. It was friable and bled on touch. A clinical diagnosis of pyogenic granuloma was made and the tumor was excised.

Multiple brownish tissue fragments were received. Microscopic examination revealed a neoplasm composed of small acini lined by single layer of cuboidal to columnar cells containing clear or pale eosinophilic cytoplasm and fairly uniform nuclei. The glands were closely packed with minimal intervening fibromuscular stroma containing lymphoplasmacytic infiltrate and congested blood vessels [Figure 1]. Some of the glands showed scanty intraluminal secretions. No mitotic activity or necrosis was noted. Periodic acid-Schiff positive granules were seen in the cytoplasm. The overlying epithelium showed focal ulceration.

Immunohistochemistry revealed that the tumor cells were strongly positive for pancytokeratin, CK 7, and

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BerEp4 and negative for CK 20, chromogranin, and synaptophysin [Figures 2 and 3].

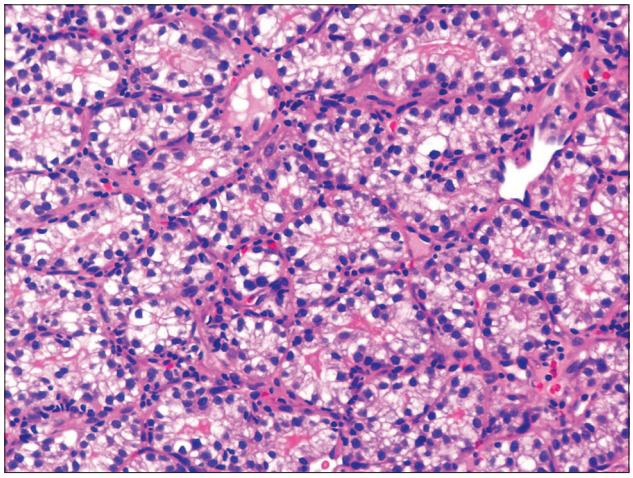

Figure 1: High power view showing acini lined by single layer of cuboidal to columnar cells containing clear to pale eosinophilic cytoplasm and fairly uniform nuclei. Intraluminal secretions are noted in some glands (H and E, ×200)

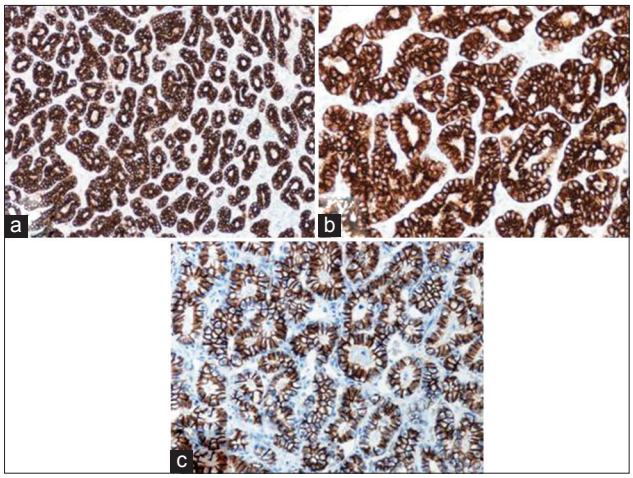

Figure 2: The neoplastic glands are strongly immunoreactive to antibodies to (a) pancytokeratin (×100) (b) CK 7 (×200) (c) BerEp4 (×200)

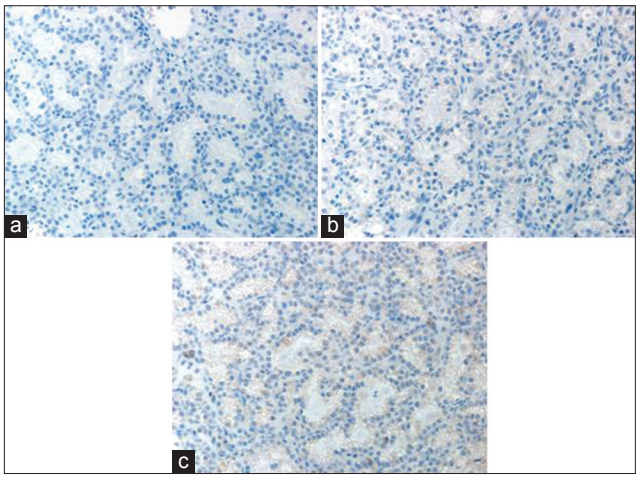

Figure 3: Glands showing negative immunostaining to antibodies to (a) CK 20 (×200) (b) chromogranin (×200) (c) synaptophysin (×200)

At 1 year follow-up, the patient was symptom-free, and nasal examination revealed no evidence of recurrence.

DISCUSSION

Nonsalivary type adenocarcinomas arising within the sinonasal tract are uncommon tumors and represent approximately 10–20% of malignant neoplasms in these locations.[6] They are often poorly recognized and misdiagnosed. The intestinal type adenocarcinomas represent 6–13% of primary neoplasms in this region[7] and are usually indistinguishable from the typical adenocarcinoma of the colon. A slight male preponderance is seen and a strong association with occupational exposure to wood and leather dust has been noted.[3,5] Their immunoprofile is also like intestinal adenocarcinomas, and neoplastic cells are immunoreactive with antibodies to CDX2, MUC2, and CK20 and negative for antibodies to CK 7.[8]

The nonintestinal-type adenocarcinomas are the most diverse of the lot. They are of a presumed seromucous gland origin lacking intestinal features. Their marked morphologic heterogeneity precludes the precise definition often resulting in a diagnostic uncertainty and rendering nonintestinal type adenocarcinomas a diagnostic category of exclusion. They show a variety of growth patterns whose morphologies do not easily fit into salivary and intestinal type adenocarcinomas. For the purpose of prognostication, they are divided into low-grade and high-grade categories based on the architecture, the nuclear features, and mitotic activity.[3,9] The low-grade nonintestinal-type sinonasal adenocarcinoma (non-ITACs) do not show any sex or racial predilection and are more commonly seen at an age beyond 50 years, as was seen in our case. They arise anywhere in the sinonasal tract; the nasal cavity being the most commonly affected, followed by the ethmoid and maxillary sinuses. Unilateral epistaxis or nasal obstruction may be the presenting symptom.[7,10] These tumors may be associated with a preexisting respiratory epithelial adenomatoid hamartoma.[8] Unlike the intestinal type adenocarcinomas, risk factors such as occupational or environmental exposure or predisposing conditions have not been reported for non-ITACs. Microscopically, these tumors are composed of uniform cells arranged in a well-differentiated glandular pattern, with back to back confluent glands, cystic spaces, and papillae showing minimal cellular pleomorphism. Tumor cells maintain tall columnar to cuboidal arrangements without much stratification and have an abundant cytoplasm that is basophilic, granular, mucinous, eosinophilic, and also oncocytic. Some cases may show microcysts filled with mucin. The nuclear atypia is mild to moderate with inconspicuous nucleoli.[10] The hallmark signs of malignancy such as perineural invasion,

lymphovascular invasion, mitotic figures, and necrosis are generally absent.[3,8]

The immunostaining profile is helpful in distinguishing low-grade non-ITAC from ITAC. The low-grade non-ITACs are positive for CK 7, Ber Ep4, and S 100 and negative for CK 20, synaptophysin, and chromogranin, as was seen in our case. Other immunostains which are negative include the CD 10, calponin, p63, MUC2 (Mucin 2), CDX2, and CD57.[8] The absence of high-grade histologic features and the immunostaining profile in our case prompted us to make the diagnosis of low-grade SNAC.

The low-grade SNAC is a relatively indolent tumor with an excellent prognosis. There is a low risk of local invasion, metastasis, and recurrence compared to the more aggressive intestinal and high-grade nonintestinal-type adenocarcinomas.[3,9] High-grade SNACs do not have an intestinal phenotype, and unlike the predominantly papillary or tubular patterns of low-grade lesions, high-grade lesions are often solid with sheets of cells or show trabecular or irregular glandular patterns.[9] They are characterized by marked cellular pleomorphism, hyperchromasia, prominent nucleoli, and a high mitotic activity. Signet-ring cells may be seen, and necrosis is often present.[5]

CONCLUSION

Low-grade SNACs should be considered in the differential diagnosis of nasal mass lesions. The importance of understanding the pathology of this entity and differentiating this neoplasm from other types of SNAC is critical as it has a low risk of local invasion and virtually nonexistent risk of metastasis compared to the more aggressive intestinal and high-grade nonintestinal-type adenocarcinomas.

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Conflicts of interest
There are no conflicts of interest.

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