

Low grade non-intestinal sinonasal adenocarcinoma

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ABSTRACT

Sinonasal tract malignancies are usually slow growing tumours with non-specific unilateral symptoms (occasionally bilateral) normal to the site of origin. They comprise a wide spectrum of pathological features. We report the case of a low grade non-intestinal sinonasal adenocarcinoma (SNAC) in a 38-year-old man who presented with a six month history of right sided nasal blockage that was associated with occasional purulent rhinorrhoea and a month history of epistaxis.

Keywords: Sinonasal undifferentiated carcinoma, head and neck neoplasms, carcinoma

INTRODUCTION

Primary sinonasal adenocarcinoma (SNACs) are classified by the World Health Organisation as either salivary and non-salivary. The salivary types are well defined myoepithelial neoplasms which closely resemble their salivary counterparts. The non-salivary types are subcategorised into intestinal type and non-intestinal type SNAC, and both have low and high grade categories respectively. The intestinal types often arise in the ethmoid sinus and are typically aggressive, resembling intestinal epithelial neoplasms. The non-intestinal SNAC are believed to originate from seromucous glands and can arise anywhere in the sinonasal tract and have marked morpho-

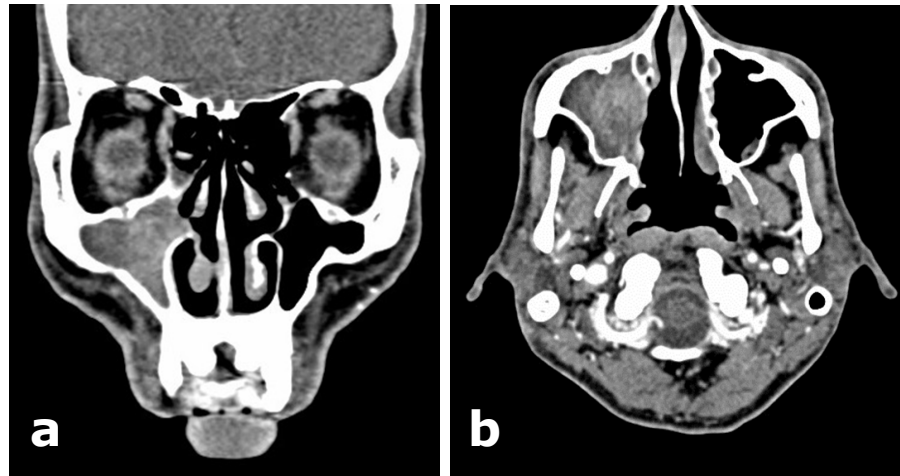
logical heterogeneity. We present the case of low grade non-intestinal SNAC in a 38-year-old man and discuss the difficulty in making the diagnosis.

CASE REPORT

A 38-year-old man presented with a six month history of right sided nasal blockage which was associated with intermittent purulent rhinorrhoea. He also had a month history of recurrent self-limiting epistaxis. He denies experiencing any headache, visual disturbances, pain, loss of appetite or weight loss. His medical and family histories were unremarkable. He was a heavy smoker and drinks on occasions. There was no history of occupational exposure to wood dust or other known carcinogens.

He was admitted during one of the

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Figs. 1: a) Coronal computed tomography (CT) image showing a mass occupying the right maxillary sinus, and b) Axial CT image showing an ill-defined enhancing mass within the right maxillary sinus with erosion of the medial wall of the maxillary sinus.

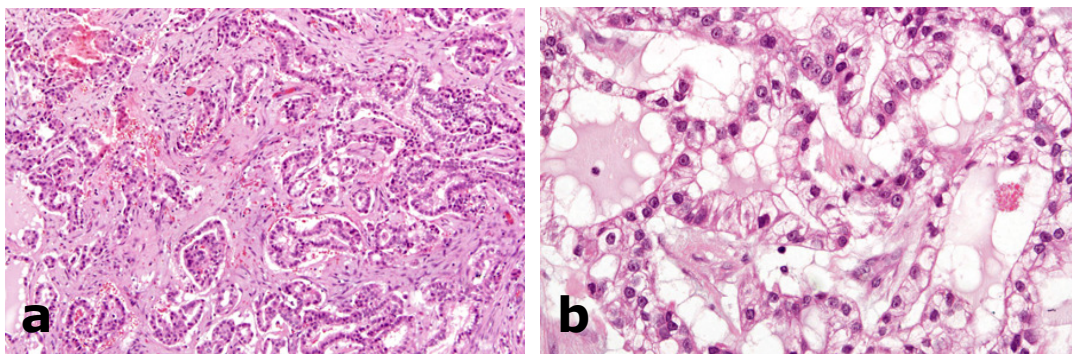
episodes of epistaxis which was profuse and required anterior and posterior nasal packing along with blood transfusion. Three days after admission, he underwent examination under general anaesthesia and a friable mass in the right nasal cavity was seen. There was also a small right sided cervical lymph node that measured 0.5 x 0.5cm.

Computed tomographic imaging (CT scan) of the base of the skull revealed an ill-defined enhancing mass (2.3 x 2.5cm) within the right maxillary sinus (Figure 1a) eroding into the wall of the medial right maxillary

sinus (Figure 1b). There was minimal extension into the right middle turbinate. Shotty submental and cervical lymph nodes were seen. The rest of examination (thorax and abdomen) was normal.

Biopsy from the mass revealed it to be a sinonasal non-intestinal adenocarcinoma, clear cell variant. Immunohistochemistry studies were positive for Cytokeratin (CK) 7 and were negative for CK20 and Carcinoembryonic Antigen (CEA).

The patient underwent a total right



Figs. 2: a) Tumour cells form tubular glands within a desmoplastic stroma (H&E, 100x) and b) a high power image showing that the tumour is composed of low grade malignant glandular cells with clear and eosinophilic cytoplasm (H&E, 400x).

maxillectomy with a modified radical neck dissection (Type 1) and surgical plating of the orbital floor with a titanium mesh. Intraoperatively, the tumour was seen to occupy the right maxillary sinus with erosion into the anterior wall. It extended medially to the middle turbinate and abutting posterior wall. The superior and inferior turbinates were not involved. The resected specimen showed that the tumour arose from the floor of the maxilla and measured 4.5cm in its greatest dimension. Histological examination revealed presence of mucin-producing malignant tubular glands within a desmoplastic stroma (Figure 2a). These malignant glands are lined by columnar cells displaying low grade malignant features with clear and eosinophilic cytoplasm (Figure 2b).

The patient later received radiotherapy, 66 gy in 33 fractions to the paranasal sinuses. There has been no sign of recurrence after five years of follow-up.

DISCUSSION

Based on the WHO histological classification, sinonasal adenocarcinoma is categorised into two types; intestinal-type and non-intestinal-type adenocarcinoma. The latter is not related to professional wood dust exposure. No risk factors, predisposing conditions, or environmental exposures have been described for the non-intestinal SNAC.^{1, 5} The more common type is the intestinal type which possibly arise from intestinal metaplasia of the ciliated respiratory epithelium lining the Schneiderian membrane.² Intestinal SNACs resemble normal, dysplastic, or malignant intestinal epithelium, with varying histologic patterns that may be predominantly papillary, glandular, compact, mucinous, or mixed.²

Despite a high risk of recurrence and subsequent deep local invasion, lymphatic and distant metastases are rare. Intestinal-SNACs are treated by complete surgical resection, followed by radiotherapy.² The non-intestinal SNAC, of presumed seromucous gland origin, are nonsalivary-type that lack intestinal features.^{3, 4} Marked morphologic heterogeneity precludes a precise definition of non-ITACs, often resulting in diagnostic uncertainty and rendering non-intestinal SNAC a diagnostic category of exclusion.⁴ Low and high-grade non-intestinal SNAC are distinguished by the presence or absence of necrosis, mitotic activity and cytologic atypia.¹

Low-grade non-intestinal SNAC can arise from anywhere in the sinonasal tract with the nasal cavity being the most commonly affected site followed by the ethmoid and maxillary sinuses.⁵ There is no gender predilection and more common after the age 50.⁶ Low-grade non-intestinal SNAC vary in both consistency and morphology. The tumours usually demonstrate well-differentiated glandular patterns, which may be exophytic and papillary or infiltrative and tubulocystic. The tumour cells exhibit uniform morphology, with abundant cytoplasm, mild-to-moderate nuclear atypia, inconspicuous nucleoli and few mitoses.^{5, 7} Despite the apparent bland morphology, their complex pattern and locally invasiveness indicate malignancy. The immunostaining profile (Table 1) is helpful to distinguish between low-grade non-intestinal SNAC from intestinal SNAC. However, it does not distinguish non-intestinal SNAC from hamartomas.⁸

Non-intestinal SNAC are treated by complete surgical excision.⁹ Extensive dis-

Table 1: Immunostaining profiles between intestinal and non-intestinal SNAC.

Immunostain	Intestinal Type	Non intestinal
CK 20	+ve	-ve
CK 7	-ve	+ve
CDX 2	+ve	-ve
Villin	+ve	-ve

The prognosis of non-intestinal SNAC is generally determined by the histologic grade. Low-grade tumours are relatively indolent with low risk of local invasion or metastasis, whereas high-grade tumours have higher risk of recurrence and death.⁵ Extensive disease with a higher histologic grade may require adjuvant radiotherapy. As our patient had extensive local disease, post-operative radiotherapy was warranted.

In conclusion, diagnosis of sinonasal malignancies requires differentiating from the squamous cell carcinoma variants, salivary type and non-salivary type tumours. Intestinal and non-intestinal adenocarcinomas can be diagnosed based on the immunohistochemical profiles. However, non-intestinal adenocarcinomas often results in diagnostic uncertainty and rendering diagnosis based on exclusion. Non-intestinal SNAC has a very good prognosis and the 5-year survival rate is very encouraging.

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