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# Case Report

## Intestinal-Type Sinonasal Adenocarcinoma: A Rare Entity with Review of Literature

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# Intestinal-Type Sinonasal Adenocarcinoma: A Rare Entity with Review of Literature

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**Abstract**
Intestinal-type sinonasal adenocarcinoma accounts for about 3% of cancers of the upper aero-digestive tract. It is commonly seen in men in the age group of 55-60 years. We present the case of a 60-year-old woman having extensive disease in the right half of the nasal cavity extending to the dura of the anterior cranial fossa, and the right orbital contents. The patient was given induction chemotherapy to make the condition surgically amenable, but the regression was modest, and hence she has been put on concurrent chemo-radiotherapy.

**Keywords:** Nasal cavity; Sinonasal adenocarcinoma.

## 1. INTRODUCTION

Sinonasal malignancies, a highly heterogeneous group of cancers, account for less than 1% of all cancers. These lesions may originate from any of the histopathologic components of the sinonasal cavities, including Schneiderian mucosa, minor salivary glands, neural tissues and lymphatics (Lango *et al.*, 2010). Adenocarcinomas account for 10-20% of all primary malignant neoplasms of the nasal cavity and paranasal sinuses. Many of these are of salivary gland origin, but others are less familiar and have histologic patterns similar to those of adenocarcinoma of the colon. These latter ones have been named intestinal-type adenocarcinoma (ITAC) and are responsible for less than 4% of the total malignancies of this region (Sklar and Pizarro, 2003). These tumours occur primarily in men aged 55-60 years. They are common in workers in the hardwood and shoe industries. Exposure to wood dust increases the risk of adenocarcinoma by 900 times (Sklar and Pizarro, 2003).

## 2. CASE PRESENTATION

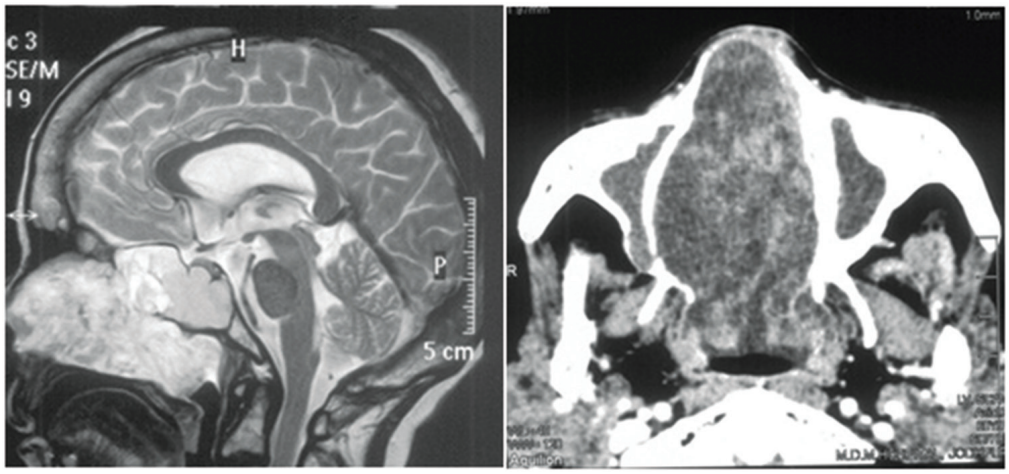
A 60-year-old woman presented with the complaints of rhinorrhoea since last 2 years, along with right-sided unilateral headache, loss in vision and proptosis in right eye since last 2 months. On examination, a mass was seen in the right nostril with deviation of septum toward opposite side. On further evaluation, CT and MRI of the face and neck revealed a large destructive expansile mass in the right nasal cavity measuring 78 × 49 × 46 mm, with extensive osseous erosions and destruction of nasal septum and bilateral turbinates/osteomeatal complexes; remodelling of nasal cavity walls and invasion of the floor of anterior cranial fossa were also seen. Though there was no intracranial extension, the lesion appears to abut the dura (Figure 1).

There was erosion of the medial right orbital wall with a frank intraorbital tumoural extension in the extraconal space, along with the distortion of the optic nerve sheath complex. Posteriorly, the tumour extended to the orbital apex with a breach of the floor of medial cranial fossa, erosion of the sellar floor. Inferiorly, the tumour involved the choanae and posterior pharyngeal wall with obliteration of nasal cavity. There was no lymphadenopathy detected.

To confirm a histopathologic diagnosis of a malignancy, the patient underwent endoscopy-directed biopsy. The histopathology showed multiple tiny polypoidal fragments with lakes and pools of mucinous material around neoplastic glandular elements. There was mild cytologic atypia; numerous inflammatory cells were seen entrapped in mucin, suggestive of intestinal-type sinonasal adenocarcinoma, mucin secreting. The patient was not found to be surgically fit for craniofacial resection because of extensive infiltration of the disease, along with bony involvement.

The patient was put on induction chemotherapy (cisplatin and etoposide) to make the condition surgically amenable. After three cycles of chemotherapy, the patient was examined again for surgical resection; there was minimal reduction in the tumour size, but still the condition was not amenable to surgery. Thereafter, the patient was put on concurrent chemo-radiotherapy, and is still taking the treatment.

**Figure 1: Imaging studies showing the extensive disease.**




## 3. DISCUSSION

Malignant sinonasal tract tumours comprise less than 1% of all neoplasms and about 3% of those of the upper aero-digestive tract. Sinonasal tract malignancies most commonly affect the maxillary sinus (about 60%), followed by the nasal cavity (about 22%), ethmoid sinus (about 15%) and frontal and sphenoid sinuses (3%) (Thompson, 2006).

Adenocarcinomas of the sinonasal tract can originate in the respiratory epithelium or the underlying mucoserous glands. These tumours are divided into two categories: salivary-gland-type and nonsalivary-gland-type adenocarcinomas. The latter are subdivided into two major categories: intestinal-type adenocarcinomas (ITACs) and nonintestinal-type adenocarcinomas. Nonintestinal-type adenocarcinomas are subclassified as low- and high-grade tumours. ITACs are a heterogeneous group of tumours, and they are further classified into a variety of subtypes (papillary, colonic, solid, mucinous and mixed) that are associated with clinically significant differences in outcomes (Thompson, 2010).

The most frequent location of ITACs (85%) is in the ethmoid and the upper part of the nasal fossa (superior and middle conchae and middle meatus), followed by the maxillary sinus (10%), and is exceptional in the rest of the sinual cavities. The mean age on presentation is 50-60 years. The distribution by gender is 4:1 in favour of males (Llorente *et al*., 2008). The site of disease and the age of our patient match with the evidence found in the literature.

Sinonasal adenocarcinomas (SNACs) are related to professional exposure to wood dust. It is estimated that the risk of professionally exposed workers developing SNAC is almost 900 times more than that of the general population. The mean time of exposure to wood dust is generally prolonged, from years to decades. Wood dust particles larger than 5 μm are considered to be the etiological factor (Llorente *et al*., 2008). SNAC grows silently with no symptoms, which leads to a late diagnosis and low survival rates. When symptoms are present, the most common are unilateral obstruction, rhinorrhoea and epistaxis. Nodes and distant metastases are rare at presentation. Patient survival depends on local control, which is extremely difficult because of the anatomical proximity of the orbit and brain (Gatta *et al*., 2009; Thompson, 2010).

Much of the literature on skull base malignancies is characterized by single-institution experiences with small numbers of patients, variable histologies and stages and aggregation of several anatomic sites in patients who have undergone a variety of treatments. Clinical outcomes from studies that include patients with esthesioneuroblastoma, sinonasal undifferentiated carcinoma (SNUC) and sinonasal melanoma are difficult to interpret, given the different clinical behaviours of these lesions. Rapid advances in surgery and radiation treatment further confound analysis because most series reflect decades of clinical experience.

Clinical outcomes for patients with SNUC have historically been dismal. SNUC is characterized by destructive rapid growth, a greater tendency to metastasize than squamous carcinomas and frequent vascular and neural invasion. Some of these symptoms suggest that it may respond more favourably to aggressive chemotherapeutic approaches. Results of a small published series, however, have been inconsistent. Ten patients with SNUCs were treated at the Peter MacCallum Cancer Centre between April 1990 and April 2002. Although one patient had a T1N0 tumour, the rest were classified T4 with intracranial and/or orbital invasion. Of these nine patients, two had initial surgery followed by postoperative radiation to a dose of 54 Gy in 30 fractions, and both developed locoregional recurrence. The other seven patients received induction chemotherapy with cisplatin (or carboplatin, if cisplatin was contraindicated) with infusional 5-FU for three cycles followed by concurrent chemoradiation with cisplatin during the first and last weeks of radiation. One patient progressed while receiving induction chemotherapy but died 5 months after completing

radiation. Six patients had stable disease or partial responses after induction chemotherapy, and four subsequently had complete responses after radiation, remaining free of disease 8-51 months after completing treatment. Two patients with incomplete responses progressed 11 months after completing treatment. Because patients who recurred received 50-54 Gy and those who remained free of disease received 60 Gy, the authors speculate that at least 60 Gy is needed to control this tumour type (Rischin *et al*., 2004). Although generally these results are better than those reported previously, follow-up of 2 of 4 patients rendered disease-free (of the 7 total in the cohort) was less than 1 year (8 and 10 months). In a review of 15 patients with T4 SNUCs treated at the University of Florida between September 1992 and October 2005, patients underwent either postoperative radiation, preoperative radiation or definitive radiation with or without concomitant chemotherapy. All patients received 60-75 Gy of radiation. Concurrent treatment with cisplatin was most commonly employed. Patients treated with surgery, either before radiation or following preoperative radiation, had better local control than patients treated with definitive radiation, despite the use of higher doses of radiation (70-75 Gy) in the nonsurgical group. One of seven patients treated with surgery and postoperative radiation developed a neck recurrence in the nonirradiated side of the neck 4 months after treatment, whereas the others remained free of disease for 12-128 months. Local control was also achieved in the two patients who had preoperative radiation. However, both patients experienced disease recurrence, one in the neck and the other at distant sites. Three of five patients treated with definitive radiation developed recurrences, one local and two locoregional. One of these with a marginal dural recurrence was successfully salvaged. Based on their results, Tanzler *et al*. (2008) conclude that combined surgery and adjuvant radiation likely offer the best chance of cure compared with either modality alone. However, it is important to note that most patients who underwent initial surgery were classified as T4a, whereas those who underwent definitive radiation were staged T4b. On the whole, however, it does seem likely that surgery, either before or after radiation, improves local control. Relapses for SNUCs and other sinonasal malignancies are usually marked by local failure and sometimes locoregional failure. The authors favour a combined approach using surgery followed by radiation to a lower dose for resectable disease, to diminish the risk of radiation-induced optic neuropathy (Bhandare *et al*., 2005).

Our patient was in the same age group as the evidence seen in literature, but with no risk factors identified in the history provided by the patient. Since the patient was found not fit for surgery at the time of presentation, attempt was made to reduce the tumour bulk by induction chemotherapy. Evaluation after three cycles of chemotherapy showed minimal reduction in the tumour size; thereafter, she has been put on concurrent chemo-radiotherapy in a curative attempt, and salvage surgery can be attempted if there will be residual disease.

## 4. CONCLUSIONS

Given the complexity of sinonasal malignancies, a multidisciplinary approach for treatment and rehabilitation is advocated. Survival after treatment for advanced sinonasal carcinomas remains suboptimal. Nevertheless, there is evidence that incremental changes have resulted in a decrease in treatment-related complications. As technical and technological advances are incorporated into the surgical and radiotherapeutic management of sinonasal malignancies, more effective treatments will be offered to a larger variety of patients.

## References

Bhandare N, Monroe AT, Morris CG, *et al*. (2005). Does altered fractionation influence the risk of radiation-induced optic neuropathy. International Journal of Radiation Oncology, Biology, Physics 62(4): 1070-1077.

Gatta G, Bimbi G, Ciccolallo L, *et al.* (2009). Survival for ethmoid sinus adenocarcinoma in European populations. Acta Oncologica, 48(7): 992-998.

Lango MN, Topham NS, Perlis CS, *et al.* (2010). Surgery in the multimodality treatment of sinonasal malignancies. Current Problems in Cancer, 34(5): 304-321. doi:10.1016/j.currproblcancer.2010.07.001

Llorente JL, Núñez F, Rodrigo JP, *et al*. (2008). Sinonasal adenocarcinomas: Our experience. Acta Otorrinolaringológica Española, 59(5): 235-238.

Rischin D, Porceddu S, Peters L, *et al*. (2004). Promising results with chemoradiation in patients with sinonasal undifferentiated carcinoma. Head & Neck, 26(5): 435-441.

Sklar EML, Pizarro JA (2003). Sinonasal intestinal-type adenocarcinoma involvement of the paranasal sinuses. AJNR American Journal of Neuroradiology, 24: 1152-1155.

Tanzler ED, Morris CG, Orlando CA, *et al*. (2008). Management of sinonasal undifferentiated carcinoma. Head & Neck 30(5): 595-599.

Thompson LDR (2006). Sinonasal carcinomas. Current Diagnostic Pathology, 12: 40-53.

Thompson LDR (2010). Intestinal-type sinonasal adenocarcinoma. Ear Nose, & Throat Journal, 89(1): 16-18.

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