

## Case Report

# An extremely rare malignant tumour of nasopharynx: primary nasopharyngeal non salivary type adenocarcinoma

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## ABSTRACT

Adenocarcinoma is commonly found in lung, prostate and gastrointestinal tract. Nasopharyngeal part contributes a very small percentage of adenocarcinoma as the most common histopathology found here is squamous cell carcinoma. We present a case of nasopharyngeal adenocarcinoma which is a very rare clinical presentation. This is 35 years old Iban gentleman, with underlying retroviral disease, hepatitis B on treatment, presented with right neck swelling for 3 months associated with right ear fullness. Examination showed unilateral right level II neck swelling and nasal endoscopy showed mass arising from right fossa of Rosen muller (FOR). The investigation done includes biopsy of right FOR and biopsy of right neck swelling with contrast enhanced computed tomography of neck, thorax and abdomen. The result of right FOR biopsy is non intestinal type adenocarcinoma, and biopsy of right neck swelling is reported as metastatic undifferentiated carcinoma. CECT NTAP showed asymmetry with fullness of right FOR associated with right level II cervical lymphadenopathy. Our centre managed this case with surgical intervention and patient underwent endoscopic nasopharyngectomy, and planned for right modified radical neck dissection (MRND). Patient defaulted our follow up after operation and was not keen for neck dissection surgery. He presented the second time with bilateral neck swelling and underwent chemotherapy and defaulted the follow up thereafter again. In conclusion the approach in treatment for nasopharyngeal carcinoma (NPC) and sino-nasal adenocarcinoma differs whereby the cornerstone management for the latter one is surgical intervention compared to the former which is radiotherapy. Hence early detection and treatment is essential.

**Keywords:** Nasopharyngeal adenocarcinoma, Nasopharyngectomy, Radiotherapy

## INTRODUCTION

Adenocarcinoma arises from mucus secreting glands and the common sites involved are lung, prostate, pancreas, esophageal and colorectal. If occur in head and neck, the commonest site is salivary gland. Hence as demonstrated in this case, nasopharynx is a very rare site for adenocarcinoma to occur which primary nasopharyngeal adenocarcinoma (NPAC) contributes 0.5% of all NPC.<sup>4</sup>

## CASE REPORT

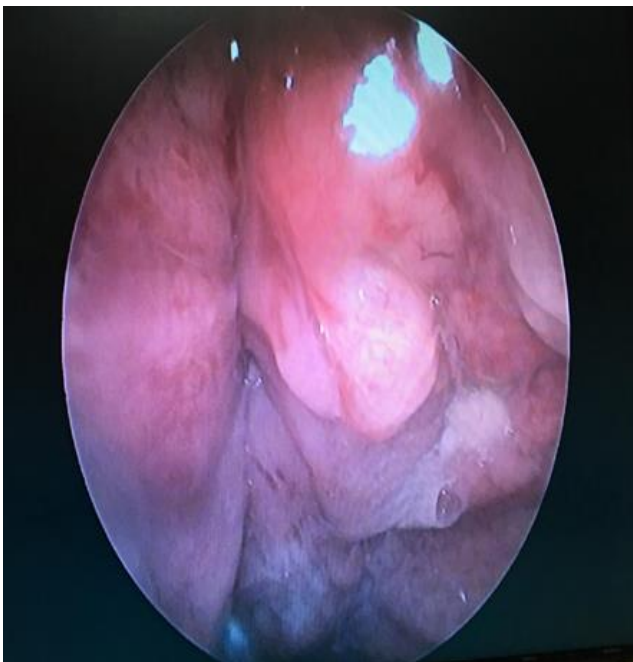
We are reporting 35 years old Iban gentleman, smoker for 30 pack years, ex alcoholic, with underlying retroviral

disease and hepatitis B on treatment. He presented with right upper lateral neck mass for 3 months associated with right ear fullness without significant nasal symptoms. On examination there is single unilateral right level II neck swelling measuring 3x3cm. Hard in consistency and slightly mobile. Nasoendoscopic examination revealed small mass at right FOR. Biopsy taken reported non-intestinal type of adenocarcinoma. A contrast-enhanced computed tomography (CECT) showed asymmetry with fullness of right FOR associated with right level II cervical lymphadenopathy measuring 1.5x3.7x3.4 m. We proceeded with incisional biopsy of neck mass showed metastatic undifferentiated carcinoma. He underwent endoscopic nasopharyngectomy, and

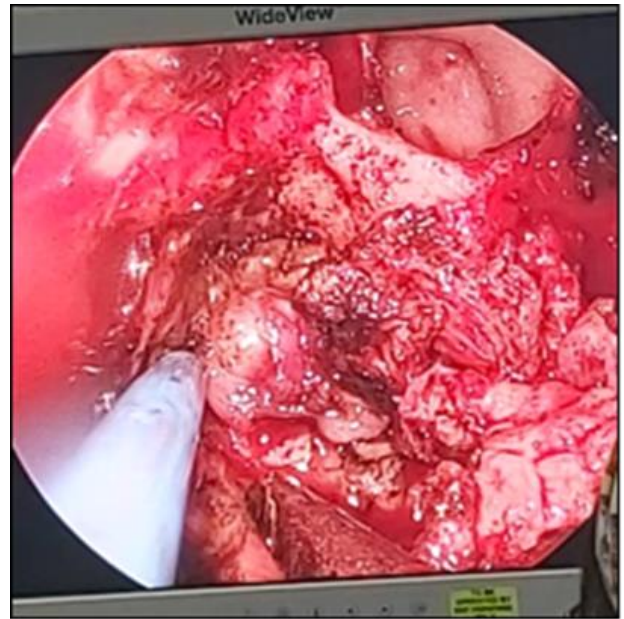
planned for right modified radical neck dissection (MRND). The biopsy of right and left nasopharyngeal mass taken intraoperatively also showed non intestinal type adenocarcinoma similar to neck mass histopathology result. After the operation, unfortunately patient not keen for neck surgery and defaulted follow up. After few months of defaulting follow up, he presented this time around with bilateral neck swelling. He underwent chemotherapy and subsequently defaulted treatment and follow up again.



**Figure 1: CT scan of asymmetry with fullness of right fossa of Rosen muller.**



**Figure 2: Right nasopharyngeal mass arising from superior part of nasopharynx and fullness of right fossa of Rosen muller.**



**Figure 3: Nasopharyngeal mass from endonasal endoscopic nasopharyngectomy procedure.**

## DISCUSSION

The commonest nasopharyngeal tumour is still NPC, followed by rarely salivary gland cancers, lymphoma and very rarely sarcoma. Since the clinical presentation of all types of nasopharyngeal malignancy are almost similar, only histopathology could reveal the type of sinonasal cancer.<sup>1</sup> According to a retrospective database review, 176 cases of sinonasal adenocarcinoma has been reported in which nasal cavity being the commonest site.<sup>1</sup> Patients could be asymptomatic at early stage.<sup>1</sup> In a study done, 78% patient present with nasal symptoms such as epistaxis, nose block and discharge, 73% present with ear symptoms such as deafness, tinnitus, 63% local nodal metastasis.<sup>1</sup>

Distant metastasis comprises about 36% in NPC and 13% in sinonasal adenocarcinoma.<sup>2</sup> Moreover it is reported that nodal metastasis is least likely found in sinonasal adenocarcinoma comprising around 4.1% incidence.<sup>3</sup> However our patient, presented initially with local nodal metastasis, neck swelling with no nasal symptom which opposes the stated incidence. Hence even though the clinical features are similar, the predominant initial presenting symptom differs between NPC and sinonasal adenocarcinoma.

There are different risk factors reported which includes EBV infection, smoking, alcohol intake, male, age >54 years.<sup>4</sup> The EBV may not contribute an essential role in NAC carcinogenesis as it is more associated with NPC.<sup>4</sup> However in a patient with HIV infection the chance of developing EBV induced cancer is increased as this is seen in our patient who has HIV infection as well.<sup>4</sup>

Moreover it is reported that the cancer tends to be more invasive and to have bad prognosis.<sup>4</sup>

The treatment option for NPC vs sinonasal adenocarcinoma differs. The standard treatment option for NPC is radiotherapy mainly.<sup>5</sup> Chemotherapy is also usually given in combination with radiotherapy for NPC.<sup>5</sup> Surgical excision plays a role in NPC when there is cervical nodal involvement or recurrent NPC.<sup>5</sup> The mainstay treatment for sinonasal malignancy is surgical excision of tumour. Moreover, radiotherapy can be offered post operatively in sinonasal non intestinal type adenocarcinoma and also as adjuvant radiotherapy for inoperable cases of sinonasal adenocarcinoma. Chemotherapy can also be given in combination with radiotherapy for nasal cavity and paranasal sinus cancers and the common regime used is cisplatin-based chemotherapy.<sup>5</sup> Our centre successfully performed endoscopic nasopharyngectomy for him. The surgical approach is challenging as its anatomically near to vital neurovascular structures.<sup>6</sup> We invited expertise to co-manage. In a retrospective review it is also stated that endoscopic nasopharyngectomy for both benign and malignant nasopharyngeal tumour revealed good outcome in which no recurrence is noted in benign mass, and recurrence for malignant mass is reported after 5 years of nasopharyngectomy.<sup>6</sup>

The concomitant neck mass must always be investigated and confirmed with histopathology. It could be a coexisting tuberculosis lymphadenitis or other types of synchronous malignancy. This makes the approach of treatment more complicated. Furthermore, lymphoma must be ruled out. In this case, the right cervical mass is metastatic adenocarcinoma from primary nasopharynx. In view of large neck node, it requires MRND. We planned to refer our patient for radiotherapy post-surgery, if his disease shown positive margin, or any perivascular and perineural spread. Sinonasal adenocarcinoma has a good prognosis with surgery and/or postoperative radiotherapy.<sup>6</sup>

## CONCLUSION

Although adenocarcinoma is extremely rare found in nasopharynx, early identification of disease is crucial to prevent late diagnosis and treatment. All suspicious lesion in nasopharynx requires biopsy confirmation until proven otherwise.

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