Case report

Primary Nasopharyngeal Adenocarcinoma (NAC) in A Remission Stage of Nasopharyngeal Squamous Carcinoma Patient

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Abstract

Nasopharyngeal adenocarcinoma is an extremely rare entity with the incidence about 0.5% of all nasopharyngeal cancer. We report a case of nasopharyngeal adenocarcinoma (NAC) in a patient who is already on remission of nasopharyngeal squamous carcinoma. He had completed radiotherapy for 35 cycles. However, after remission completed for about 2 years with regular follow up, he was diagnosed with another carcinoma. Due its rarity, treatment modality of nasopharyngeal adenocarcinoma is still debatable as no studies has been shown to be the best modalities. Nasopharyngectomy, neck nodes dissection as well as post-operative radiotherapy are the best treatment of choice for this type of carcinoma.

Keywords: Nasopharyngeal Adenocarcinoma, Nasopharyngeal Carcinoma, Nasopharyngectomy

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Introduction

Nasopharyngeal carcinoma (NPC) is frequently diagnosed in Southeastern Asia, predominantly in southern China¹ NPC is a multifactorial disease. The various causative factors include genetic susceptibility, environmental factors dietary and personal habit. The most common type of NPC cases that has been suggested are keratinizing and nonkeratinizing squamous cell carcinoma². Another types of NPC includes adenocarcinoma, minor salivary gland, lymphoma and sarcoma which contributes <5 % of all NPC cases³. Thus, primary nasopharyngeal adenocarcinoma (NAC) is tremendously rare which are reported to occupy 0.48% of all types of NPC⁴.

Case summary

59-year-old Chinese gentleman was diagnosed in 2016 as squamous cell carcinoma of nasopharynx.

His initial presentation was persistent epistaxis for 3 months duration. Otherwise, there was no other nasal or ear symptoms, no neck swelling and no constitutional symptoms. He completed 35 cycles of radiotherapy in 2016. Patient was well until July 2018 when he presented with complained of intermittent headache. He denied having any episodes of epistaxis, neck swelling, ear symptoms or any cranial neuropathy.

General examinations revealed no significant findings. Nasoendoscopic examination revealed a bulging mass at left fossa of rosenmuller (FOR) with contact bleeding. The mass covered with whitish secretion. Biopsy of the mass was taken with high suspicious of clinical diagnosis at that particular time was to rule out recurrence. However, histopathology report came back as nasopharyngeal adenocarcinoma. Computed

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<u>Correspondence to:</u> Dr. Wan Ahmad Amiruddin bin Wan Hassan, Department of Otorhinolaryngology Sultan Ahmad Shah Medical Centre @IIUM 25200 Kuantan Pahang Malaysia & Department of Otorhinolaryngology, Hospital Tengku Ampuan Afzan, 25100 Kuantan, Pahang, Malaysia E-mail: <u>amiruddin1988@gmail.com</u> tomography staging (neck,thorax abdomen and pelvis) revealed assymetrical of nasopharynx with fullness of left FOR with no evidence of bony erosion and cervical lymphadenopathy (Figure 1).

Second biopsy of the left fossa of rosenmuller was taken after oncological consultation revealed similar findings. Patient went for Positron Emission Tomography (PET) scan and it showed there was a local recurrence at nasopharynx with no evidence of distant metastasis. Nasopharyngectomy with neck dissection was suggested after another discussion with oncologist. However patient was not keen for the operation.



Figure 1 CT scan neck/thyroid showed fullness of left nasopharynx with obliteration of left Fossa of Rosenmuller.

Discussion

Nasopharyngeal carcinoma (NPC) is a malignant tumour of nasopharynx. It usually involved Chinese community particularly in Southeast Asia,

Southern China and North African countries. In Malaysia, NPC is the 4th most common cancer in the population ⁵. The commonest histopathology of nasopharyngeal neoplasm is nonkeratinizing and keratinizing squamous cell carcinomas without glandular differentiation².

Based on Zhong et al⁶, nasopharyngeal adenocarcinoma can be divided into minor salivary glands (salivary gland type) and mucosal surface epithelium (general type). The examples of salivary gland type are adenoid cystic carcinoma and mucoepidermoid while the general type, including papillary adenocarcinoma, acinic cell carcinoma and others.

Our patient was diagnosed to have NPC in 2016 and completed radiotherapy for 35 cycles. However, after 2 years of the initial diagnosis, he was diagnosed to have another cancer which is nasopharyngeal adenocarcinoma. The initial staging of his disease was T1N0M0 in 2016. Two samples from left fossa of rosenmuller were taken within two months' time showed features of adenocarcinoma (Figure 2). Both samples were positive for CKAE1/AE3, CK7, TTF-1 while CK20, Napsin A and thyroglobulin were negative. Following that, he underwent Positron Emission Tomography (PET) scan which showed local recurrence at left nasopharynx with no evidence of metastasis.

The management of the NAC is based on histology grading and clinical staging with no standard treatment exists for this type of tumour. Due to rarity, there is no clear-cut management and prognosis such as long-term survival of patient with NAC⁷. Controversial on treatment

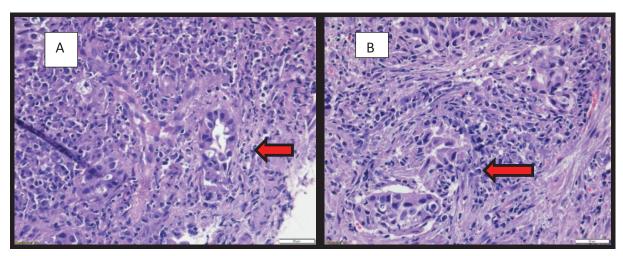


Figure 2 : Sample taken from left fossa of Rosenmuller (A : first sample, B: second sample) showed malignant cells infiltration forming glands (arrow). [H&E, 400x magnification].

modalities either involving radiotherapy and surgical treatment alone, or surgery combined with radiotherapy is the optimal therapeutic approach. There are studies proposed that surgery together with radiotherapy will be the key treatment for resectable and limited NACs, for example adenoid cystic carcinoma, mucoepidermoid carcinoma, and well-differentiated adenocarcinoma8. Treatment of patient with nasopharyngeal adenoid cystic carcinoma with surgery coupled with radiotherapy or surgery alone increase 5-year survival rate compare with compared to radiotherapy, concurrent chemoradiotherapy or chemotherapy alone⁹. However, for the poorly differentiated or unresectable primary NACs, regardless of the general and salivary gland type, it is very tough to achieve the purpose of radical cure with surgery alone. For limited or resectable adenocarcinoma,

combined surgery with radiotherapy may be the appropriate treatment policy. On the other hands radiotherapy or chemoradiotherapy is also suitable for the poorly differentiated NAC or unresectable tumours.

Conclusion

Nasopharyngeal adenocarcinoma is extremely rare. Treatment modalities of nasopharyngeal adenocarcinoma is still debatable as no studies has been shown to be the best modalities. Nasopharygectomy, neck nodes dissection as well as post-operative radiotherapy are currently the best treatment of choice.

Conflict of interest

The authors declare no conflicts of interest regarding the publication of this paper.

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