Case Report

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Low grade nasopharyngeal papillary adenocarcinoma case report from the Middle East

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ABSTRACT

Nasopharyngeal papillary adenocarcinoma variety is extremely rare type of nasopharyngeal carcinoma. Limited number of cases reported in the literature, with most cases being reported from west and south Asia. We report the first case in English literature from the middle east and south Africa. A young gentleman diagnosed with low-grade nasopharyngeal papillary adenocarcinoma with positive cervical lymph node. He received a combined approach with endoscopic resection of the lesion and concomitant Left neck dissection followed by planned chemoradiotherapy. Through this case report, we would like to share our treatment approach and explore the optimum therapeutic strategies for primary nasopharyngeal papillary adenocarcinomas.

Keywords: Papillary adenocarcinoma, Nasopharyngeal, Carcinoma

INTRODUCTION

Nasopharyngeal carcinoma is the most common malignancy of the nasopharynx, with fewer than 100,000 cases in the world every year. Main distribution of cases in southeastern Asia and middle east.¹

Nasopharyngeal carcinoma consists of a group of malignant epithelial tumors. World Health Organization (WHO) categorized these cancers into non-keratinizing, keratinizing, or basaloid type. Non-keratinizing tumors are further sub-divided into undifferentiated or differentiated.¹

Nasopharyngeal papillary adenocarcinoma is a rare primary tumor of the nasopharynx with incidence of less than 1% of nasopharyngeal malignancies. To our knowledge limited cases have been reported in English

literature till date.² Most reported cases are from east and south Asian countries.² We report the first case from the Middle east and south Africa.

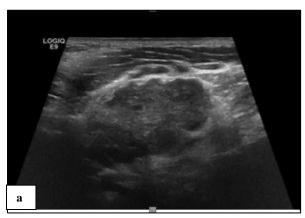
CASE REPORT

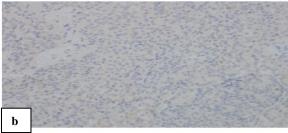
A 28-year-old male Egyptian patient, known smoker (30 pack years) presented with history of left sided neck swelling for about six months with associated fullness and pressure in the left ear. Patient denied any history of nasal obstruction, epistaxis, facial pain, numbness, fever, weight loss or night sweat. Patient consulted in a private clinic, where a magnetic resonance imaging (MRI) revealed nasopharyngeal mass. Patient was subsequently referred to our hospital. Examination revealed, left sided firm, immobile, painless neck mass at level 2, left sided dull

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tympanic membrane and a nasopharyngeal mass obstructing the choana on the left side.

Patient subsequently underwent a nasopharyngeal biopsy with left myringotomy, which revealed a highly vascular nasopharyngeal mass with serous fluid on myringotomy. The biopsy result came out as low grade nasopharyngeal papillary carcinoma (TTF1) figure 1b, cytokeratin 20-CK20 and thyroglobulin negative).





Negative for TTF-1 antibody

Figure 1: (a) Ultrasound of the neck shows metastatic lymph node, (b) immunohistopathology slide:
-Ve TTF1 antibody

The patient was referred to multidisciplinary team (MDT) meeting to determine further course of action. It was decided in the MDT meeting to do an ultrasound of the thyroid with Fine needle aspiration cytology (FNAC) of the neck lesion. The ultrasound ruled out any thyroid lesions, but showed metastatic lymph node (figure 1a), while the FNAC revealed metastatic carcinoma with papillary architecture. Computed tomography (CT) of chest, abdomen and pelvis showed no sign of metastasis.

Patient subsequently underwent transoral nasopharyngeal mass lesion resection with left neck dissection (level 2, 3,4) (Figure 2 a, b).

Postoperatively, pathology report confirmed Low grade nasopharyngeal papillary adenocarcinoma (LGNPPA). Metastasis in one of the 5 lymph nodes from level 2 with lymph vascular invasion and extra nodal extension. The nasopharyngeal mass margins could not be assessed due to the fragmented nature of the specimen. Pathological staging was same as that of clinical staging, T2N1M0.

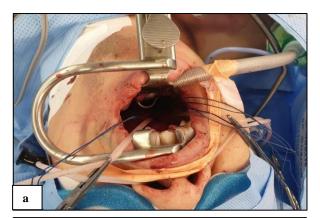




Figure 2: (a) Intraoperative photo: transoral approach to nasopharyngeal mass, (b) Intraoperative photo: Curette of nasopharyngeal mass.

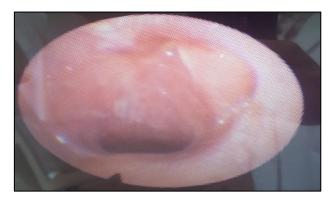


Figure 3: Fibro-optic assessment of nasopharynx after treatment shows normal examination.

Following which the patient received radiation to the tumor, high risk areas. low risk area and electively to a total dose of 70 Gy, 66 Gy, 59.4 Gy and 54 Gy respectively concomitant with cisplatin. Patient reassessed after 3 months of completion of chemoradiotherapy. Patient is asymptomatic with clear fiberoptic assessment (figure 3) ad normal MRI scan.

DISCUSSION

Nasopharyngeal papillary adenocarcinoma 3 originates from nasopharyngeal mucosa and 4 shows a remarkable

papillary architecture. They occur over a wide age range (reported ages: 9-64 years), with equal gender distribution, and can involve any part of the nasopharynx.¹

There are no known etiological agents, for nasopharyngeal papillary adenocarcinoma as there is no Epstein-barr virus or human papillomavirus association.³ Also, there is no association with wood exposure.

Patient with Nasopharyngeal papillary adenocarcinoma (NPPAC) mostly present with nasal obstruction as the common symptom, although otitis media or postnasal drip can be seen in cases where there is involving of posterior or lateral nasopharyngeal walls.³ Rare presentation included secondary to head radiation, and incidental diagnosis during diagnosis of patient with fever of unknown origin also reported.^{2,5}

Primary nasopharyngeal papillary adenocarcinomas present with the similar pathological characteristics as nasopharyngeal metastatic lesions associated with thyroid importance malignancies. Hence the immunohistochemical studies to differentiate.1 NPPAC express EMA, cytokeratin 5/6- CK5/6 and often cytokeratin 7- CK7. A subset of NPPAC cases express cytokeratin 19 (CK19) and Thyroid transcription factor 1 (TTF1) and referred to as "thyroid-like low-grade nasopharyngeal papillary adenocarcinoma."6 However, this subtype does not express thyroglobulin which differentiate it from metastatic papillary thyroid carcinoma.3

In 1988, Wenig et al first described thyroid-like papillary adenocarcinoma of the nasopharynx, and in 2005 Nasopharyngeal papillary adenocarcinoma (NPPAC) was enrolled in the World Health Organization classification system of malignant epithelial tumors of the nasopharynx.^{7,8}

The case we presented was LGNPPA with negative TTF1, CK20 and thyroglobulin. The negative reactivity for both CK20 and thyroglobulin support differentiating metastatic thyroid papillary carcinoma from LGNPPA.

Treatment

Nasopharyngeal papillary adenocarcinoma in an extremely rare malignancy with good prognosis.³ No metastases cases reported to date.⁹ Complete surgical excision is usually curative. Although due to the small anatomical area of the nasopharynx and difficult access complete resection can be a challenge. Partial resection can increase the local risk of recurrence.³ Unfortunately, lowgrade papillary NPACs, have low sensitivity to conventional radiotherapy or chemoradiotherapy, and it became a challenge to deal with partially removed tumors.¹

Photodynamic therapy (PDT) is a new treatment modality for head and neck cancers, with satisfactory treatment responses and minimal complications. 10-11 A recent study by Wang et al, suggested the use of PDT combined with topical 5-aminolevulinic acid as an effective modality of the postoperative adjuvant therapy to successfully eradicate the residual disease, without compromising the quality of life of the patient.¹¹⁻¹³

CONCLUSION

The differential diagnosis of nasopharyngeal masses should include primary NPPACs. Although they are very rare with good prognosis. Inadequate management may result in local recurrence of the disease. Total excision Although challenging can be curative. PDT may be a more suitable alternative method to be the adjuvant therapy for residual tumors. Till date no clear guideline for treatment of Primary nasopharyngeal papillary adenocarcinomas, large multicentric studies or meta-analyses to further evaluate the optimal treatment is needed.

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