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CASE REPORT

The Rare Primary Low Grade Papillary Adenocarcinoma of Nasopharynx: A Diagnostic Challenge and Management

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ABSTRAK

Papillary Nasopharyngeal Adenocarcinoma Gred Rendah (LGPAC) adalah sejenis tumor berasal dari mukosa yang jarang berlaku berbanding kanser nasofarinks jenis "keratinized". Secara histologi, terdapat diferensiasi jenis papillari yang juga boleh didapati dalam kes metastatik karsinoma papilari tiroid. Keadaan ini menyebabkan cabaran bagi pihak patologi untuk membezakan antara kes primari atau metastatik. Thyroid Transcription Factor-1 (TTF-1) diekspreskan dalam subset Papillary Nasopharyngeal Adenocarcinoma dan ini merupakan alat diagnostic yang berguna. LGPAC adalah tumour malignan bersifat benign. Diagnosis awal beserta pembedahan membuang keseluruhan tumour melalui cara konvensional atau endoskopik boleh memberikan prognosis bagus dan risiko berlakunya lagi tumour ini adalah rendah.

Kata kunci: adenokarsinoma papilari, nasofarinks, neoplasia tiroid

ABSTRACT

Low Grade Papillary Nasopharyngeal Adenocarcinoma (LGPAC) is a very rare tumour of mucosal origin compared to a higher incidence of well differentiated keratinized/non-keratinized nasopharyngeal carcinoma. It is an epithelial tumour with glandular differentiation. Its papillary figure seen histologically, is also seen in metastatic papillary thyroid carcinoma. This has caused a significant challenge to the Pathologist to differentiate primary papillary nasopharyngeal adenocarcinoma and metastatic tumour. Thyroid Transcription Factor-1 (TTF-1) is also expressed in subsets of papillary nasopharyngeal adenocarcinoma, which is valuable as a diagnostic tool. LGPAC is a benign-like malignant neoplasm. An early diagnosis with a complete tumour removal via conventional excision or endoscopic approach has offered a good prognosis with low risk of recurrence.

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Keywords: adenocarcinoma papillary, nasopharynx, thyroid neoplasms

INTRODUCTION

Primary Low Grade Papillary Nasopharyngeal Adenocarcinoma has been scarcely reported in literature. It is a benign-like malignant neoplasm. The expertise of the Pathologist is required to identify specific histopathological features of this tumour. There are helpful immunohistochemistry markers for e.g. Thyroid Transcription factor-1 (TTF-1) which is expressed in subsets of such tumour, to aid with the diagnosis (Wu et al. 2007). An early diagnosis with a complete tumour removal offers a good prognosis with low risk of recurrence (Fu et al. 2008).

CASE REPORT

A 15-year-old boy presented with 5 months history of right nasal blockage and runny nose. There was no associated epistaxis. Upon examination, there was a mass at the centre of nasopharynx extending anterosuperiorly to the roof of right posterior choana. There was no evidence of cervical lymphadenopathy or anterior neck mass. Biopsy of the tumour showed Low Grade Papillary Nasopharyngeal Adenocarcinoma. Histopathology findings showed neoplastic tissue comprising papillary fronds and crowded glands lined by cuboidal to pseudostraitified columnar epithelium. Immunohistochemical studies revealed that these neoplastic cells were immunopositive for epithelial markers including Epithelial Membrane Antigen, Carcinoembryogenic Antigen and cytokeratin. They were negative for Thyroglobulin and S100.

Paranasal sinuses scan obtained (Figure 1), demonstrated a well circumscribed 3 x 3cm polypoidal hypodense lesion arising from the roof of nasopharynx, obscuring the post nasal space. The lesion did not erode or destruct the base of skull (Figure 2). Subsequently, he underwent sphenodotomy, septectomy and removal of mass. Histopathology findings reconfirmed the result of LGPAC. Postoperative nasal endoscopies showed no evidence of mass in the nasopharynx and sphenoid cavities at 4 years, post surgery. Currently, the patient is on 6-monthly follow-up, and exhibited no clinical evidence of recurrent adenocarcinoma.

DISCUSSION

Nasopharyngeal carcinomas are established tumour classified by WHO into keratinizing or non-keratinizing carcinomas without evidence of glandular differentiation. The incidence of primary adenocarcinoma of nasopharynx is less than 0.3-0.4% of all nasopharyngeal malignancies (Fu et al. 2008). They can be divided into mucosal origin type and salivary type based on their morphological features. It is important to differentiate these two groups as the treatment and prognosis differ. Most of the mucosal type has papillary features and is low grade in nature. Low Grade Papillary

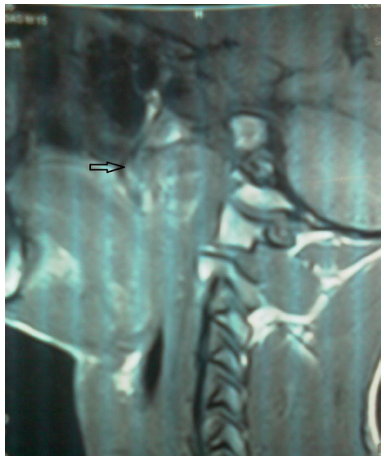

Figure 1: A sagittal view of preoperative CT scan showing a tumour occupying the nasopharyngeal space (arrow)

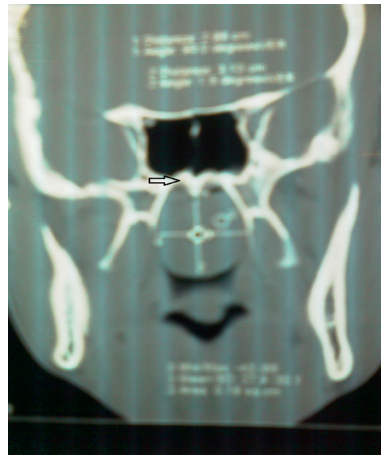

Figure 2 : A coronal view of CT scan showing an intact skull base with no evidence of bony erosion or destruction by the nasopharyngeal tumour (arrow)

Nasopharyngeal Adenocarcinoma (LGPAC) is categorized under mucosal type and to the best of our knowledge and limited reference available is extremely rare. It tends to arise in middle-aged group with female predilection (Ohe et al. 2010). As LGPAC is diagnosed in our patient who is a male teenager, it should be in the list of differential diagnosis in children or teenager with nasopharyngeal tumour.

Papillary nasopharyngeal adenocarcinomas can arise from 3 sources, which are mucosal surface, salivary gland or thyroid gland (as a form of metastasis). It is a known challenge to the Pathologists to propose a proper diagnosis as the histological features of malignancy from these origins are relatively similar to each other. LGPAC commonly involves the roof, lateral, or posterior walls of the nasopharynx and is described as exophytic with a polypoidal or nodular appearance. Majority of patients present with symptoms including nasal obstruction, epistaxis, aural fullness, tinnitus or otitis media.

Histologically, one of the significant findings is to observe a transition of the mucosal surface from normal epithelium to neoplastic surface. There are papillary projections lined by cylindrical to cuboidal stratified cells. There is no evidence of perineural, vascular or lymphatic invasion. It is important to carefully observe the histological findings of this tumour, as a subset of LGPAC can closely resemble metastatic papillary thyroid carcinoma (Wu et al. 2007).

Both have papillary architectures with fibrovascular cores, overlapping nuclei with clear chromatin, and psammoma bodies and this particular subset can express thyroid transcription factor-1 (TTF-1), thus gaining the acronym thyroid-like papillary nasopharyngeal adenocarcinoma. Therefore, when a positive HPE of LGPAC is obtained, a thyroid scan and systemic work-up should be carried out

to rule out metastatic disease. Other helpful features to differentiate primary or metastatic LGPAC with positive TTF-1 include predominant stratified nuclei, absence of Tyroglobulin expression in tumour sections and negative thyroid imaging (Pineda-Daboin et al. 2006). Therefore, TTF-1 immunoreactivity in the absence of thyroid or pulmonary primary can be used as a diagnostic tool for LGPAC.

LGPAC acts relatively like a benign tumour. It has a low percentage of local recurrence and metastasis with no lymphatic spread being reported. The treatment of choice is complete surgical excision using transpalatal conservative techniques (Fu et al. 2008). Endoscopic excision is proposed for small tumour. Patients do not require subsequent radiotherapy as most of the cases are tumour free after a complete surgical excision but is beneficial for cases with incomplete resection to prevent recurrence (Fu et al. 2008).

Even though studies have reported such an excellent outcome with a very small possibility of recurrence, it is advisable to follow-up LGPAC patients regularly. Those with TTF-1 demonstrated, should have regular imaging study of thyroid gland as not to miss delayed presentation of primary thyroid malignancy (Wu et al. 2007).

CONCLUSION

The rare papillary adenocarcinoma of nasopharynx requires experts knowledge in diagnosis, as an early identification may ensure good prognosis. Although, it acts in a benign manner, LGPAC should be of caution as one of a significant malignant tumour of nasopharynx.

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